Welcome To
Congress of Clinical Rheumatology-West

Fellows’ Poster Session

September 25, 2019
Faculty

FPS Judging Panel Faculty

Janet Pope, M.D., MPH, FRCPC

Professor of Medicine
Western University, London
London, Ontario, Canada
Division Head in Rheumatology
St. Joseph’s Health Centre
London, Ontario, Canada

Yusuf Yazici, M.D.

Associate Professor of Medicine
Director, Seligman Center for Advanced Therapeutics
NYU School of Medicine
New York, NY
<table>
<thead>
<tr>
<th>Last Name</th>
<th>First Name</th>
<th>Associated Program</th>
<th>City, ST</th>
</tr>
</thead>
<tbody>
<tr>
<td>Akram, M.D.</td>
<td>Bushra</td>
<td>Oklahoma University Health Sciences Center</td>
<td>Oklahoma City, OK</td>
</tr>
<tr>
<td>Al Asmar, M.B.B.S.</td>
<td>Rania</td>
<td>Marshall University School of Medicine</td>
<td>Huntington, WV</td>
</tr>
<tr>
<td>Ali, M.D.</td>
<td>Hiba</td>
<td>Ochsner Medical Center &amp; Louisiana State University Health Science Center</td>
<td>New Orleans, LA</td>
</tr>
<tr>
<td>Al-Saiegh, M.D.</td>
<td>Yousif</td>
<td>Pennsylvania Hospital - University of Pennsylvania Health System</td>
<td>Philadelphia, PA</td>
</tr>
<tr>
<td>Aniekwena, M.D.</td>
<td>Judith</td>
<td>Morehouse School of Medicine</td>
<td>Atlanta, GA</td>
</tr>
<tr>
<td>Bourgoyne, M.D.</td>
<td>Kesler</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Bui, M.D.</td>
<td>Brian</td>
<td>Arrowhead Regional Medical Center</td>
<td>Colton, CA</td>
</tr>
<tr>
<td>Chalasani, M.B.B.S.</td>
<td>Swathi</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Chawla, M.D.</td>
<td>Ambreesh</td>
<td>University of Central Florida College of Medicine</td>
<td>Orlando, FL</td>
</tr>
<tr>
<td>Fervaha, M.D.</td>
<td>Harman</td>
<td>Ochsner Medical Center &amp; Louisiana State University Health Science Center Shreveport</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Fuchs, M.D.</td>
<td>Perry</td>
<td>University of Arizona College of Medicine Phoenix</td>
<td>Phoenix, Arizona</td>
</tr>
<tr>
<td>Garlapati, M.B.B.S.</td>
<td>Priyatha</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Goyal, B.S.</td>
<td>Anirudh</td>
<td>Rutgers New Jersey Medical School</td>
<td>Newark, NJ</td>
</tr>
<tr>
<td>Goyal, M.P.H.</td>
<td>Tanvi</td>
<td>University of Toledo College of Medicine</td>
<td>Toledo, OH</td>
</tr>
<tr>
<td>Hackett, B.M., B.S., PhD</td>
<td>Simon</td>
<td>University of Oxford Medical School</td>
<td>Oxford, United Kingdom</td>
</tr>
</tbody>
</table>
# 2019 Fellows’ Poster Session Participants

<table>
<thead>
<tr>
<th>Last Name</th>
<th>First Name</th>
<th>Associated Program</th>
<th>City, ST</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hanif, M.B.B.S.</td>
<td>Soniya</td>
<td>Wayne State University School of Medicine; Henry Ford &amp; Providence Hospitals</td>
<td>Detroit, MI</td>
</tr>
<tr>
<td>Hill, D.O.</td>
<td>Brittany</td>
<td>Brookwood Baptist Health System</td>
<td>Birmingham, AL</td>
</tr>
<tr>
<td>Kachur, M.D., MS</td>
<td>Patricia</td>
<td>Ochsner Medical Center &amp; Louisiana State University Health Science Center</td>
<td>New Orleans, LA</td>
</tr>
<tr>
<td>Kallas, M.D.</td>
<td>Romy</td>
<td>Johns Hopkins University School of Medicine</td>
<td>Baltimore, MD</td>
</tr>
<tr>
<td>Karczewski, M.D.</td>
<td>Jan</td>
<td>Roger Williams Medical Center, Boston University School of Medicine</td>
<td>Providence, RI</td>
</tr>
<tr>
<td>Kang, M.D.</td>
<td>Mandip</td>
<td>University of California San Francisco Fresno</td>
<td>Fresno, CA</td>
</tr>
<tr>
<td>Katz, M.D.</td>
<td>Guy</td>
<td>Thomas Jefferson University Hospital</td>
<td>Philadelphia, PA</td>
</tr>
<tr>
<td>Kazem, M.D., MHA</td>
<td>Mikameh</td>
<td>University of Western Ontario</td>
<td>London, Ontario</td>
</tr>
<tr>
<td>Kollipara, B.S.</td>
<td>Sai</td>
<td>Indiana University of Medicine and Health Sciences</td>
<td>Indianapolis, IN</td>
</tr>
<tr>
<td>Korem, M.B.B.S</td>
<td>Sindhuja</td>
<td>Monmouth Medical Center, Drexel University College of Medicine</td>
<td>Long Branch, NJ</td>
</tr>
<tr>
<td>Koyoda, M.B.B.S</td>
<td>Sai</td>
<td>Monmouth Medical Center, Drexel University College of Medicine</td>
<td>Long Branch, NJ</td>
</tr>
<tr>
<td>Kumar, M.B.B.S</td>
<td>Mukund</td>
<td>CHI Health Creighton University Medical Center</td>
<td>Omaha, NE</td>
</tr>
<tr>
<td>Lora Garcia, M.D.</td>
<td>Luis Gerald</td>
<td>TriHealth’s Good Samaritan Hospital</td>
<td>Cincinnati, OH</td>
</tr>
<tr>
<td>Maheswaranathan, M.D.</td>
<td>Mithu</td>
<td>Duke University Medical Center</td>
<td>Durham, NC</td>
</tr>
<tr>
<td>Malhotra, M.D.</td>
<td>Kanchan</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Last Name</td>
<td>First Name</td>
<td>Associated Program</td>
<td>City, ST</td>
</tr>
<tr>
<td>-----------------</td>
<td>------------</td>
<td>--------------------------------------------------------------------------------------</td>
<td>------------------</td>
</tr>
<tr>
<td>Malus, M.D.</td>
<td>Matthew</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>McConnaughy, M.D.</td>
<td>Taylor</td>
<td>Cottage Health System</td>
<td>Santa Barbara, CA</td>
</tr>
<tr>
<td>Minalyan, M.D.</td>
<td>Artem</td>
<td>Abington Hospital - Jefferson Health</td>
<td>Abington, PA</td>
</tr>
<tr>
<td>Mollaeian, M.D.</td>
<td>Arash</td>
<td>MedStar Union Memorial Hospital, Georgetown University Medical School</td>
<td>Baltimore, MD</td>
</tr>
<tr>
<td>Moore, M.D.</td>
<td>Meriah</td>
<td>University of Michigan Medical Center</td>
<td>Ann Arbor, MI</td>
</tr>
<tr>
<td>Morin, D.O.</td>
<td>Scott</td>
<td>Mount Auburn Hospital, Harvard Medical School</td>
<td>Cambridge, MA</td>
</tr>
<tr>
<td>Movahedian, M.D.</td>
<td>Malahat</td>
<td>MetroHealth Medical Center, Case Western Reserve University</td>
<td>Cleveland, OH</td>
</tr>
<tr>
<td>Muhieddine, M.D.</td>
<td>Leila</td>
<td>MetroHealth Medical Center, Case Western Reserve University</td>
<td>Cleveland, OH</td>
</tr>
<tr>
<td>Myasoedova, MD, PhD, MSc</td>
<td>Elena</td>
<td>Mayo Clinic College of Medicine &amp; Science</td>
<td>Rochester, MI</td>
</tr>
<tr>
<td>Nichols, B.S.</td>
<td>Austin</td>
<td>Marshall University Joan C. Edwards School of Medicine</td>
<td>Huntington, WV</td>
</tr>
<tr>
<td>Onuorah, M.D.</td>
<td>Nneoma</td>
<td>Wright State University Boonshoft School of Medicine</td>
<td>Dayton, Ohio</td>
</tr>
<tr>
<td>Patel, D.O.</td>
<td>Kinal</td>
<td>Ascension Macomb-Oakland Hospital</td>
<td>Warren, MI</td>
</tr>
<tr>
<td>Penumarty, M.D.</td>
<td>Sravani</td>
<td>Rutgers New Jersey Medical School</td>
<td>Newark, NJ</td>
</tr>
<tr>
<td>Piponov, M.D.</td>
<td>Hristo</td>
<td>University of Illinois Hospital &amp; Health Sciences System at Chicago</td>
<td>Chicago, IL</td>
</tr>
<tr>
<td>Riley, D.O.</td>
<td>Mark</td>
<td>Albany Medical Center Hospital &amp; Albany Medical College</td>
<td>Albany, NY</td>
</tr>
<tr>
<td>Last Name</td>
<td>First Name</td>
<td>Associated Program</td>
<td>City, ST</td>
</tr>
<tr>
<td>---------------</td>
<td>------------</td>
<td>--------------------------------------------------------</td>
<td>-----------------</td>
</tr>
<tr>
<td>Sahar, M.B.B.S</td>
<td>Najmus</td>
<td>Wright State University Boonshoft School of Medicine</td>
<td>Dayton, OH</td>
</tr>
<tr>
<td>Sajjadi, M.D.</td>
<td>Faye</td>
<td>Jersey City Medical Center</td>
<td>Jersey City, NJ</td>
</tr>
<tr>
<td>Sen, B.A.</td>
<td>Aslihan</td>
<td>Rutgers New Jersey Medical School</td>
<td>Newark, NJ</td>
</tr>
<tr>
<td>Shih, M.D.</td>
<td>Raymond</td>
<td>St. Mary Medical Center</td>
<td>Langhorne, PA</td>
</tr>
<tr>
<td>Spears, MB, BCH, BAO</td>
<td>Jenna</td>
<td>Hospital of the University of Pennsylvania &amp; Perelman School of Medicine</td>
<td>Philadelphia, PA</td>
</tr>
<tr>
<td>Stacy, MSIV</td>
<td>John</td>
<td>University of North Dakota School of Medicine and Health Sciences</td>
<td>Grand Forks, ND</td>
</tr>
<tr>
<td>Umar, M.D.</td>
<td>Anam</td>
<td>Morehouse School of Medicine</td>
<td>Atlanta, GA</td>
</tr>
<tr>
<td>Vuppala, M.D.</td>
<td>Anusha</td>
<td>Louisiana State University Health Science Center</td>
<td>Shreveport, LA</td>
</tr>
<tr>
<td>Zonoozi, M.B.B.S</td>
<td>Shahrzad</td>
<td>Hospital of the University of Pennsylvania &amp; Perelman School of Medicine</td>
<td>Philadelphia, PA</td>
</tr>
<tr>
<td>CATEGORY</td>
<td>POSTER PRESENTATION TITLE</td>
<td>PRESENTER</td>
<td>POSTER #</td>
</tr>
<tr>
<td>----------</td>
<td>---------------------------</td>
<td>-----------</td>
<td>----------</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>Refractory Adult Onset Still's Disease with Initial Good Response to Canakinumab: A Case Report</td>
<td>Mandip Kang, M.D.</td>
<td>4</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>Atypical Adult Still's Disease-A Diagnosis of Confusion</td>
<td>Judith Aniekwena, M.D.</td>
<td>5</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>An Uncommon Symptom Constellation in Hidradenitis Suppurativa</td>
<td>Anirudh Goyal, B.S.</td>
<td>6</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>A Fatal Case of Adult Onset Still’s Disease (AOSD) Complicated with Hemophagocytic Lymphohistiocytosis (HLH)</td>
<td>Sindhuja Korem, M.B.B.S</td>
<td>7</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>Delirium–Atypical Presentation of Gouty Arthritis</td>
<td>Sai Koyoda, M.B.B.S</td>
<td>8</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>How Sweet of A Flu</td>
<td>Arash Mollaeian, M.D.</td>
<td>9</td>
</tr>
<tr>
<td>CRYSTALS &amp; AUTO INFLAMMATORY DISEASES</td>
<td>Adult-onset Still's disease with Macrophage Activation Syndrome</td>
<td>Raymond Shih, M.D.</td>
<td>10</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Clinical Pearls of Atypical Hemolytic Uremic Syndrome</td>
<td>Kesler Bourgoyne, M.D., MS</td>
<td>14</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>WHEN NOTHING IS SURE, EVERYTHING IS POSSIBLE</td>
<td>Swathi Chalasani, M.D.</td>
<td>15</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Can’t See, Can’t Hear, Can’t Understand</td>
<td>Patricia Kachur, M.D.</td>
<td>16</td>
</tr>
<tr>
<td>CATEGORY</td>
<td>POSTER PRESENTATION TITLE</td>
<td>PRESENTER</td>
<td>POSTER #</td>
</tr>
<tr>
<td>--------------</td>
<td>------------------------------------------------------------------------------------------</td>
<td>----------------------------------</td>
<td>----------</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Acute Monoarthritis and Colitis Secondary to Anti-Cancer Therapy</td>
<td>Jan Karczewski, M.D.</td>
<td>17</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Refractory Valvular Heart Disease Due to ImmunoglobulinG4-Related Disease of the Aortic and Mitral Valves</td>
<td>Guy Katz, M.D.</td>
<td>18</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Massive hemoptysis- A fatal complication in sarcoidosis with airway involvement</td>
<td>Mukund Kumar, M.B.B.S.</td>
<td>19</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Mycosis fungoides: The Great Imitator</td>
<td>Kanchan Malhotra, M.D.</td>
<td>20</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Sarcoidosis or Cryptococcosis: Which Came First? A Puzzling Clinical Association</td>
<td>Matthew Malus, M.D.</td>
<td>21</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>A case of catastrophic antiphospholipid syndrome with severe thrombocytopenia complicated by the development of disseminated intravascular coagulopathy</td>
<td>Artem Minalyan, M.D.</td>
<td>22</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Multidisciplinary Approach to Digital Ischemia and the Role of Rheumatology in Buerger’s Disease</td>
<td>Meriah N. Moore, M.D.</td>
<td>23</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Does Not Vanish</td>
<td>Sravani Penumarty M.D.</td>
<td>24</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>A bumpy Road to Final Diagnosis</td>
<td>Najmus Sahar, M.B.B.S.</td>
<td>25</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>A case of Refractory Neurosarcoidosis</td>
<td>Faye Sajjadi, M.D.</td>
<td>26</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Respiratory Failure Due to Autoimmune Encephalitis Responsive to Cyclophosphamide</td>
<td>Aslihan Sen, B.A.</td>
<td>27</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>Non-Bacterial Thrombotic Endocarditis in a Patient with Primary Antiphospholipid Syndrome</td>
<td>Jenna Spears, M.B., BCh, BAO</td>
<td>28</td>
</tr>
<tr>
<td>CATEGORY</td>
<td>POSTER PRESENTATION TITLE</td>
<td>PRESENTER</td>
<td>POSTER #</td>
</tr>
<tr>
<td>-------------------</td>
<td>-------------------------------------------------------------------------------------------</td>
<td>--------------------------------</td>
<td>----------</td>
</tr>
<tr>
<td>MISCELLANEOUS</td>
<td>An uncommon cause of common presentation—IgG4 related disorder</td>
<td>Anusha Vuppala, M.D.</td>
<td>29</td>
</tr>
<tr>
<td>MUSCLE DISEASES</td>
<td>Idiopathic Inflammatory Myopathy Complicated by Respiratory and Renal Failure</td>
<td>Brian Bui, M.D.</td>
<td>30</td>
</tr>
<tr>
<td>MUSCLE DISEASES</td>
<td>Favorably Unfavorable: Interstitial Lung Disease as the Initial Manifestation of Anti-Mi-2 Positive Dermatomyositis</td>
<td>Ambreesh Chawla, M.D.</td>
<td>31</td>
</tr>
<tr>
<td>MUSCLE DISEASES</td>
<td>The presence of Calciosis cutis universalis in an adult patient with Dermatomyositis</td>
<td>Kanchan Malhotra, M.D.</td>
<td>32</td>
</tr>
<tr>
<td>MUSCLE DISEASES</td>
<td>Can’t walk, Can’t Breathe—A Case report on Anti MDA 5 antibody Positive Dermatomyositis</td>
<td>Anusha Vuppala, M.D.</td>
<td>33</td>
</tr>
<tr>
<td>MUSCLE DISEASES</td>
<td>Statin-Induced Autoimmune Myopathy: A Clinical Case Report</td>
<td>Shahrzad Zonoozi, M.B.B.S</td>
<td>34</td>
</tr>
<tr>
<td>OSTEOPOROSIS</td>
<td>Multiple Ipsilateral Femoral Stress Fractures in a Patient Taking Denosumab for Osteoporosis - A Case Report</td>
<td>Hristo I. Piponov, M.D.</td>
<td>35</td>
</tr>
<tr>
<td>OSTEOPOROSIS</td>
<td>Osteoporosis screening in African American patients with rheumatoid arthritis. Are we doing enough?</td>
<td>A. Umar, M.D.</td>
<td>36</td>
</tr>
<tr>
<td>RA</td>
<td>The Efficacy of Low Dose Prednisone for Remission Induction in Newly Diagnosed Rheumatoid Arthritis Patients</td>
<td>Jack Stacy, MSIV</td>
<td>45</td>
</tr>
<tr>
<td>RA</td>
<td>Rheumatoid Arthritis Patients and Coronary Artery Disease: Are We Considering the Real Risk?</td>
<td>Rania Al Asmar, M.B.B.S.</td>
<td>52</td>
</tr>
<tr>
<td>RA</td>
<td>Influenza Vaccination Rates Among Patients with Rheumatoid Arthritis and Systemic Lupus Erythematosus on Immunosuppressive Therapy</td>
<td>Judith Aniekwena, M.D.</td>
<td>53</td>
</tr>
</tbody>
</table>
## FPS Categories & Poster Numbers

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>POSTER PRESENTATION TITLE</th>
<th>PRESENTER</th>
<th>POSTER #</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>Treat to Target: Do Rheumatologists Adjust Therapy Based on High Disease Activity Score (DAS), and Does This Result in Improved DAS</td>
<td>Patricia Kachur, M.D.</td>
<td>54</td>
</tr>
<tr>
<td>RA</td>
<td>A case of Posterior Reversible Encephalopathy Syndrome in a patient taking Methotrexate and Abatacept</td>
<td>Luis Lora, M.D.</td>
<td>55</td>
</tr>
<tr>
<td>RA</td>
<td>Declining Incidence of Cardiovascular Disease in Patients with Incident Rheumatoid Arthritis In 2000s: a Population-Based Cohort Study</td>
<td>Elena Myasoedova, MD, PhD, MSc</td>
<td>56</td>
</tr>
<tr>
<td>RA</td>
<td>Identifying Prevalence of Pneumococcal Vaccinations Among Rheumatoid Arthritis Patients of the Rural Appalachian Population in an Academic Rheumatology Clinic</td>
<td>Austin Nichols, B.S.</td>
<td>57</td>
</tr>
<tr>
<td>RA</td>
<td>A 60-year-old Male with Felty’s Syndrome Case report and Literature Review</td>
<td>Nneoma Onuorah, M.D.</td>
<td>58</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Treatment of Diffuse Alveolar Hemorrhage with Plasmapheresis in Acute Systemic Lupus Erythematosus</td>
<td>Yousif Al-Saiegh, M.D.</td>
<td>61</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>A Rare Case of ANCA-Negative Pauci-Immune Necrotizing Crescentic Vasculitis in a Systemic Lupus Erythematosus</td>
<td>Hiba Ali, M.D.</td>
<td>62</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Confused and Itchy: An Unusual Presentation of Systemic Lupus Erythematosus in an Elderly Caucasian Male</td>
<td>Ambreesh Chawla, M.D.</td>
<td>63</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Culture Negative Mechanical Valve Endocarditis Complicated with Corticosteroid Induced Mobitz Type II AV Block in a Patient with SLE</td>
<td>Harman Fervaha, M.D.</td>
<td>64</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>A Case Report of Skin Thickening: Scleroderma or Malignancy</td>
<td>Soniya Hanif, M.D.</td>
<td>65</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Prevalence of OSA in lupus patients and effect of intervention with CPAP and its</td>
<td>Soniya Hanif, M.D.</td>
<td>66</td>
</tr>
<tr>
<td>CATEGORY</td>
<td>POSTER PRESENTATION TITLE</td>
<td>PRESENTER</td>
<td>POSTER #</td>
</tr>
<tr>
<td>----------</td>
<td>--------------------------</td>
<td>-----------</td>
<td>----------</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>compliance on lupus disease activity in terms of its flares</td>
<td>Romy Kallas, M.D.</td>
<td>67</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Association of Smoking Status and Total and Individual Damage Index in Systemic Lupus Erythematosus</td>
<td>Romy Kallas, M.D.</td>
<td>67</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Elevated Cardiac Troponin T in Patients with Lupus Myositis</td>
<td>Guy Katz, M.D.</td>
<td>68</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Arthritis in Systemic Sclerosis: A Practical Classification Scheme</td>
<td>Mikameh Kazem, M.D., MHA</td>
<td>69</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Barriers to Medication Adherence among Hospitalized Patients with Systemic Lupus Erythematosus</td>
<td>Mithu Maheswaranathan, M.D.</td>
<td>70</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Autoantibody-Mediated Extreme Alterations in Serum Glucose as the Initial Presentation of a Diagnosis of Systemic Lupus Erythematosus</td>
<td>Taylor A. McConnaughy, M.D.</td>
<td>71</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Eosinophilic Fasciitis in an Elderly Patient with New High Titer Anti-dsDNA</td>
<td>Leila Muhieddine, M.D.</td>
<td>72</td>
</tr>
<tr>
<td>SLE CTD</td>
<td>Opening the Inflammatory Floodgates: A Case of Lupus Presenting as Severe Multi-Systemic Organ Failure</td>
<td>Faye Sajjadi, M.D.</td>
<td>73</td>
</tr>
<tr>
<td>SPA</td>
<td>A Unique Presentation of Aseptic Abscess Syndrome in a Patient with Inflammatory Bowel Disease</td>
<td>Nneoma Onuorah, M.D.</td>
<td>82</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Retinal Vasculitis in a Patient with Systemic Lupus Erythematosus</td>
<td>Hiba Ali, M.D.</td>
<td>86</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>TO BE, OR NOT TO BE</td>
<td>Swathi Chlasani, M.D.</td>
<td>87</td>
</tr>
<tr>
<td>CATEGORY</td>
<td>POSTER PRESENTATION TITLE</td>
<td>PRESENTER</td>
<td>POSTER #</td>
</tr>
<tr>
<td>----------</td>
<td>------------------------------------------------------------------------------------------</td>
<td>----------------------------------</td>
<td>----------</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Pyoderma Gangrenosum, a Rare Dermatologic Complication of IgA Vasculitis</td>
<td>Perry Fuchs, M.D.</td>
<td>88</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Immunosuppression in a case of Staphylococcus associated Henoch Schönlein Purpura</td>
<td>Priyatha Garlapati, M.D.</td>
<td>89</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Rapidly Progressive Glomerulonephritis (RPGN)-An Important Diagnostic Consideration for Patients Presenting with Bilateral Peripheral Neuropath</td>
<td>Tanvi Goyal, MPH</td>
<td>90</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>An unusual case of giant cell arteritis (GCA) in a 75-year-old gentleman</td>
<td>Simon Hackett, B.M., B.S., PhD</td>
<td>91</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Leukocytoclastic Vasculitis Confounding Panton-Valentine Leukocidin Methicillin-Resistant Staphylococcus aureus pneumonia</td>
<td>Brittany Hill, D.O.</td>
<td>92</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Vasculitis Neuropathy Masquerading as Guillain-Barre Syndrome</td>
<td>Jan Karczewski, M.D.</td>
<td>93</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Cutaneous Necrosis: An unexpected trigger</td>
<td>Sai Kollipara, MS-IV</td>
<td>94</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>A Rare Presentation of Concurrent Small and Large Vessel Vasculitis</td>
<td>Sindhuja Korem, M.B.B.S</td>
<td>95</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Pursuit of A Clue</td>
<td>Arash Mollaeian, M.D.</td>
<td>96</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Characteristics Associated with Greater Corticosteroid Requirement in Patients with Giant Cell Arteritis</td>
<td>Scott Morin, D.O.</td>
<td>97</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Antineutrophil Cytoplasmic Antibody-Associated Vasculitis with atypical manifestation of renal involvement</td>
<td>Malahat Movahedian, M.D.</td>
<td>98</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>A Perplexing Case of Granulomatosis with Polyangiitis</td>
<td>Kinal Patel, D.O.</td>
<td>99</td>
</tr>
</tbody>
</table>
### FPS Categories & Poster Numbers

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>POSTER PRESENTATION TITLE</th>
<th>PRESENTER</th>
<th>POSTER #</th>
</tr>
</thead>
<tbody>
<tr>
<td>VASCULITIS</td>
<td>In a Blink of an Eye: Blindness Secondary to Primary Angiitis of CNS Presenting as Recurrent Idiopathic Intracranial Hypertension</td>
<td>Sravani Penumarty, M.D.</td>
<td>100</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Late-Onset Hemorrhagic Cutaneous IgA Vasculitis: A Unique Severe Presentation</td>
<td>Mark L. Riley, D.O.</td>
<td>101</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Adult IgA vasculitis (Henoch–Schönlein purpura) with End-Stage Renal Disease</td>
<td>Raymond Shih, M.D.</td>
<td>102</td>
</tr>
<tr>
<td>VASCULITIS</td>
<td>Apixaban Induced Leukocytoclastic Vasculitis</td>
<td>Jenna Spears, MB, BCh, BAO</td>
<td>103</td>
</tr>
</tbody>
</table>
POSTER PRESENTATIONS

Listed Alphabetically By Presenting Author’s Last Name
Chronic-ies of A Broken Heart. A Case Report of Eosinophilic Myocarditis

Authors: Akram, Bushra (B.A); Peterson. JoElle (J.P); Pakala. Aneesh (A.P); Vaseer, Samera (S.V)

ABSTRACT:
Eosinophilic myocarditis (Loeffler endomyocardial disease) is a rare inflammatory process involving the cardiac musculature. It has been sparsely reported in literature and occurs due to hypersensitivity reactions, asthmatic bronchitis, eosinophilic granulomatous polyangiitis (EGPA), parasite infections and idiopathic hypereosinophilic syndrome. While mimicking acute coronary syndrome, the disease is often missed and untreated. Here we present a case of eosinophilic myocarditis associated with rheumatoid arthritis successfully managed with oral glucocorticoids and rituximab infusion.

CASE REPORT:
A 59-year-old man with recently diagnosed type II diabetes, presented to the Emergency Room (ER) with a six month history of fatigue, chest pain and elevated troponin. Prior ER visits for similar presentation and resulted in Percutaneous Coronary Intervention (PCI) for presumed non ST elevation MI. At the time of admission, patient had mild cognitive impairment, dusky discoloration of toes and bilateral foot drop. Labs showed WBC of 13.9 with 69% eosinophils with a count of 9.45 K/mm3 and troponin of 5.59ng/ml. Echocardiography revealed LVEF 30-40% and biventricular intracavitary thrombi. Eosinophilic myocarditis was suspected. MRI of the brain showed multiple infarcts in the supra and infratentorial region which were thought to be embolic from the thrombus. Cardiac MRI revealed global hypokinesis and diffuse subendocardial late gadolinium enhancement. Cardiac biopsy confirmed the diagnosis and showed myonecrosis with mixed inflammatory infiltrate, predominantly eosinophils, consistent with eosinophilic myocarditis. He was started on prednisone at 1mg/kg daily which improved his symptoms and normalized troponins, improved eosinophilia from 69% to 2% with eosinophil counts from 9.5 to 0.14 K/mm3. He had no history of asthma and workup was negative for parasitic or hematologic causes of eosinophilia but positive Anti CCP and RF titers. He was diagnosed with rheumatoid arthritis, eosinophilic myocarditis and peripheral vasculitis (EGPA) and was started on Rituximab to which he responded well. A follow up cardiac MRI seven months after treatment showed resolution of acute myocardial inflammation but persistent fibrosis. Patient was started on heart failure therapy. He continues to follow up in outpatient rheumatology and cardiology clinics.

DISCUSSION:
Eosinophilic myocarditis is a rare disease associated with increased morbidity and mortality if misdiagnosed or untreated. Thrombosis can be seen in up to 13% cases. Acute myocardial infarction may be caused by thromboemboli, coronary vasculitis, coronary spasm, or regional inflammation. Intra cavity thrombi on echocardiography and fibrosis on cardiac MRI in the setting of eosinophilia are highly suggestive of the disease with endomyocardial biopsy being diagnostic. While treatment depends upon the cause of the disease, in patients with unknown triggering factors, corticosteroids have shown improved outcomes.

DISCLOSURE STATEMENT:
None of the authors have any disclosures to declare pertaining to the above case.
Rheumatoid Arthritis Patients and Coronary Artery Disease: Are We Considering the Real Risk?

Rania Al Asmar, MBBS, alasmar@marshall.edu
Sudha Penumala, MD, sudha.penumala@gmail.com
Emilia Leigh, MD, leighe@marshall.edu
Kanaan Mansoor, MBBS, kanaanm@gmail.com
Ralph Webb, MD, rwebb@marshall.edu
Marshall University School of Medicine, Huntington, WV 25701.

Abstract:

**Background:** Rheumatoid arthritis (RA) is one of the commonest inflammatory conditions linked to premature coronary artery disease (CAD) (1). We live in an area where metabolic syndrome and RA coexist and are very prevalent, as well as cigarette smoking (2), (3). We aim in this study to look at the demographics of our RA patient population, incidence of coexisting metabolic syndrome and smoking, as well as whether primary prevention of CAD using aspirin and statins was done in patients who developed clinically significant CAD on follow up. **Methods:** We conducted an observational, retrospective study based on electronic charts review of patients diagnosed with RA and CAD who are seen in our outpatient Byrd Clinical Center between 2015 and 2019. Charts were collected based on ICD-10 codes of the diagnoses; RA (M05.79, M06.09, M06.9), and CAD (I25.10, I21.9). **Results:** Out of 2275 patients with RA following in one of our clinics, 227 patients had coexisting diagnosis of CAD, an additional 20 patients were excluded due to incomplete charts. Mean BMI of the patients was 29.5 kg/m2. Sixty three percent were smokers, and 65% has family history of CAD. Coexisting comorbidities were prevalent; as there were 38% diabetic, 91% hypertensive, 82% dyslipidemic, and 33% had other autoimmune disease. Only 15% had coexisting diagnosis of cancer (images 1 and 2). Of note, primary prevention of CAD was only recruited in almost 36% of patients by using a statin, and 41% of patients by using low dose aspirin. Death as an outcome from different causes was 16%. **Conclusion:** Rheumatoid arthritis remains one of the important risk factors for premature coronary artery disease, especially in the predisposed patient population. More studies are needed to assess the exact risk of premature CAD in RA patients with metabolic syndrome, and perhaps consider RA as one additional determinant of the Atherosclerotic Cardiovascular Disease (ASCVD) risk.

References:

The Purpose of the Research:
Rheumatoid arthritis (RA) is a common, chronic inflammatory condition that predisposes individuals to premature coronary artery disease (CAD). At this time there is not a unified management plan for patients with rheumatoid arthritis with who have other risk factors for coronary artery disease. We propose an observational retrospective study to review the records of our patients with rheumatoid arthritis to focus on the concomitant use of aspirin and statins and in this population and whether coronary artery disease was apparent before or after the diagnosis of rheumatoid arthritis was established.

The Scientific or Scholarly Rationale:
We will try to calculate the incidence of coronary artery disease in our rheumatoid arthritis patient population, with attention paid to age, gender, body mass index (BMI), and other pertinent risk factors for coronary artery disease, whether patients were on aspirin or statin therapy, and whether risk factors modifications (e.g., smoking cessation therapy, exercise program) were pursued before developing clinically significant coronary artery disease.

We will review records of rheumatoid arthritis patients (ICD-10 codes M05.79, M06.09, M06.9) with coronary artery disease documented on their charts (ICD-10 codes I25.10, I21.9) occurring between 2015 and 2018.

Procedure to be Performed:
Electronic medical records review and data extraction.

The Risks and Potential Benefits of the Research:
No risks identified.

We hope to establish the incidence of coronary artery disease in our rheumatoid arthritis population, with subsets based on gender, BMI, and other CAD risk factors. We plan to assess how many patients were on aspirin and/or statin therapy before the development of clinically significant CAD, and whether use or lack of these agents was statistically significant in regards to the prevalence of CAD in this patient population..

Complete Inclusion/Exclusion Criteria (may be submitted separately if extensive):
Included are all adult patients, 18 years of age or older, seen in outpatient clinics at Byrd Clinical Center between 2015 and 2018, with the diagnoses of rheumatoid arthritis and coronary artery disease.

We excluded patients who were not seen at Byrd clinical center after their initial registration and thus had empty charts.
Graphs 1 and 2: Obesity 1 = BMI 30-34.9 kg/m², Obesity 2 = BMI 35-39.9 kg/m², Obesity 3 = BMI 40 kg/m² or more (WHO).

COID: All the authors, including the presenter, have no conflicts of interest and no disclosures to make.

This study was IRB approved by our institution.
A Rare Case of ANCA-Negative Pauci-Immune Necrotizing Crescentic Vasculitis in a Systemic Lupus Erythematosus

Authors: Hiba Ali MD, William Davis MD, Evangeline Scopelitis MD

1Department of Rheumatology, Ochsner Medical Center, New Orleans, Louisiana

No Financial Disclosures

Background: Glomerulonephritis in SLE is usually characterized by immune complex and complement deposition. Pauci-immune Glomerulonephritis is frequently associated with Anti-nuclear cytoplasmic antibody (ANCA). Overlap of SLE and ACNA associated vasculitis is rare with an estimated prevalence of 2%. We present a rare case of a patient with newly diagnosed SLE who presents with worsening proteinuria. Renal biopsy showed pauci-immune necrotizing crescentic glomerulonephritis in the absence of ANCA.

Case Presentation: A 30 y/o F with a previous diagnosis of psoriasis presents with fatigue, dyspnea, nausea and vomiting for one week. During this illness patient discovered that she was pregnant by home pregnancy test. On arrival to an outside hospital, patient was found to be severely anemic (Hgb 4) and in acute renal failure (Cr. 8) with profound azotemia. She was then transferred for higher level of care. She reported a history of rash to her scalp, hands and feet since 2017, with her left hand becoming contracted over the last year. Further review of systems revealed chest pain for the past one week (substernal intermittent, pressure like). She had no alopecia but a scaly rash on her scalp and behind the ears. She denied oral ulcers but did have history of nasal ulcers. She endorsed Raynauds, dry eyes and dry mouth. She has 4 children with 2 prior miscarriages in first trimester.

Her labs showed lymphopenia (absolute count 500), anemia (post transfusion Hgb 8.3), haptoglobin <10, DAT +. She was found to have +ANA 1:2560, +Sm/RNP, +SSA, SPEP showing polyclonal gammopathy, sed rate 41, CRP 31.4, C3/C4 WNL, negative dsDNA, negative APS panel. Her urine showed 3+ protein, 3+blood, 87 RBC, 20WBC hyaline and granular casts, UPC of 7.14g. Spun urine showing dysmorphic RBCs, WBC casts and 1 RBC casts. ANCA, PR3, MPO and cryoglobulins were negative. Renal biopsy showed changes consistent with concurrent pauci-immune necrotizing crescentic glomerulonephritis and low grade IgA nephropathy. After discussion with nephrology and pathology it was deemed that the IgA nephropathy was insignificant.

During patient’s hospitalization, she had a spontaneous abortion, underwent treatment with IV steroids and cyclophosphamide.

Conclusion: Pauci-Immune glomerulonephritis is the most common rapidly progressive glomerulonephritis, accounting for 80% of cases. ANCAs are negative in about 10% of these. Our case is unique in that it showed a newly diagnosed SLE with ANCA negative pauci-immune necrotizing crescentic glomerulonephritis on initial presentation.
Retinal Vasculitis in a Patient with Systemic Lupus Erythematosus

Authors: Hiba Ali MD1, William Davis MD1

1Department of Rheumatology, Ochsner Medical Center, New Orleans, Louisiana

No Financial Disclosures

Background: About 25% of patients with Systemic Lupus Erythematosus have ocular manifestations. One of the most vision threatening complications is retinal vasculitis. Prompt diagnosis and treatment of eye disease is paramount as these patients often have high levels of systemic inflammation and end organ damage. We present a case of a patient with SLE who presents with acute right eye vision loss.

Case Presentation: A 28 y/o F with a medical history of HTN, SLE (discoid lupus, malar rash, +ANA 1:1280, +SSA, +Sm, +RNP, leukopenia, membranous lupus nephritis) was admitted for one week of blurry vision in her right eye which progressed to complete loss of vision. She initially started seeing black floating dots which progressed to intermittent blurry vision. She described it as looking into a “dirty mirror.” Patient had similar symptoms in 2012 with complete loss of vision in both eyes, that returned after two weeks of hospitalization and treatment with steroids. Of note, she received 6 doses of cyclophosphamide for her renal disease in the past. Current medications included Cellcept 1g BID, PLQ 300mg daily, Benlysta, and prednisone 5mg.

She was evaluated by ophthalmology in the ED. Right eye fundus exam showed temporal pallor, clinically significant macular edema, early perivascular sheating/cuffing, and diffuse cotton wool spots most prominent at superior arcade. Her exam was most consistent with SLE vasculitis based on decreased visual acuity, color and vasculitis changes in posterior pole. MRI brain showed no acute abnormalities. Labs showed Wct of 2.19 ANC 800, Hgb stable at 11.7, plt 174, sed rate of 25, CRP 0.72, dsDNA negative, c3/c4 WNL, GFR> 60, UA with no active sediment and normal UPC. Infectious workup including HIV, syphilis, EBV, HSV and CMV negative. APLS labs and ANCA also negative. Patient had a negative NMO ab but had a positive Ribosomal P. Patient had a severe SLE flare with manifestation of SLE retinal vasculitis, mucosal ulcers, discoid rash, vasculitic nodules on hands, scaring alopecia, leukopenia (SLEDAI of 23). Patient was pulsed with IV solumedrol for 3 days and started on Cytoxan.

Conclusion: Retinal vasculitis is the most serious of ocular manifestations and can lead to progression of CNS lupus especially with a positive Ribosomal P antibody. Our patient was started on Cytoxan and had improvement in her vision and in her Retinal exam. This case emphasizes the importance of ophthalmologic exams in patients with SLE.
Treatment of Diffuse Alveolar Hemorrhage with Plasmapheresis in Acute Systemic Lupus Erythematosus

Yousif Al-Saiegh MD, Philip Williams, Jenna Spears MD

Disclosure: none

**Background:** Systemic Lupus Erythematosus (SLE) is a chronic inflammatory condition in which autoantibodies respond to cytoplasmic and nuclear antigens. It is characterized by an array of clinical manifestations some of which include fatigue, joint pain, rash, hair loss, anemia, nephritis. It primarily affects females (approximately 90%) and is more prevalent in non-Caucasian populations. Patients typically have chronic symptoms requiring immunosuppressive therapy. Acute flares are usually treated with high dose corticosteroids. When acute flares do not respond to pulse dose steroid therapy, or when complications such as diffuse alveolar hemorrhage (DAH) are present, other therapeutic modalities such as plasmapheresis should be considered. The following describes a case of a severe SLE flare successfully managed with five sessions of plasmapheresis.

**Case Presentation:** 25 year old female with past medical history of SLE diagnosed one month prior, managed on mycophenolate and prednisone. She was transferred from outside hospital to our ICU with an acute SLE flare complicated by DAH requiring intubation and worsening Acute Kidney Injury (AKI).

She initially presented to the outside hospital after developing fever, chills, hemoptysis, edema and myalgias. At the outside hospital labs were notable for a creatinine: 1.31, hemoglobin: 9, decreased IgA, IgG, and IgM, C3: 20, C4: 5, and CRP of 8.7. Chest X-ray and Chest Computed Tomography were notable for a large multi-focal pneumonia, predominantly in the left lower lobe. Labs subsequently worsened with a hemoglobin nadir of 5.5, and creatinine of 2.48. The patient developed hemoptysis and bronchoscopy was done. Bronchoscopy findings confirmed DAH.

The patient was treated for the SLE flare complicated by AKI and DAH with mycophenolate, high dose pulsed corticosteroids, two therapeutic plasma exchange sessions and was transferred to our hospital ICU. After transfer, three more sessions of plasma exchange were completed. Hemodialysis was initiated for volume overload and metabolic acidosis, at this time her creatinine was 4.4. Eight days after transfer, the patient no longer required hemodialysis and her creatinine had improved to 2.23. Her hemoglobin was stable at 8.5 and her DAH had improved significantly on repeat chest X-rays. The patient was discharged home on prednisone, mycophenolate, hydroxychloroquine, and atovaquone for PCP prophylaxis.

**Discussion:** DAH is a feared complication of SLE that is associated with a high mortality. The patient had typical presenting features of DAH, including hemoptysis, anemia, pulmonary infiltrates and acute respiratory failure. DAH commonly presents in conjunction with acute nephritis. Bronchoalveolar lavage can be used to confirm the diagnosis. Given the high mortality, treatment should be initiated immediately. First line therapy is intravenous high dose corticosteroids such as methylprednisolone, in combination with Cyclophosphamide or Rituximab. Plasma exchange can be used as an adjunctive treatment, as seen in this case, to reduce the immune complex burden.
Atypical Adult Still's Disease - A Diagnosis of Confusion

Authors
Judith Aniekwena, MD¹; Keerthi Padooru, MD¹; Leslie Anne Cassidy, MD²; Athanasios N Tiliakos, DO²

Author affiliations:
1. Morehouse School of Medicine, Department of Medicine
2. Emory University School of Medicine, Department of Medicine, Division of Rheumatology

Introduction
Adult Still’s disease (ASD) is a rare systemic autoinflammatory disorder of uncertain etiology characterized by the classic triad of high-spiking quotidian fevers, evanescent rash, and arthritis. ASD remains a diagnosis of exclusion, and infection, malignancy, and systemic autoimmune diseases must be ruled-out in all patients with suspected disease. We describe an atypical case of ASD not meeting full classification criteria, leading to a diagnostic odyssey and delay in diagnosis.

Case Description
A 55-year-old Caucasian male with a history of benign prostatic hyperplasia presented to the emergency department with a 10-week history of fevers, night sweats, fatigue, cough, sore throat, and weight loss. On review of systems, he noted mild stiffness in the fingers and toes. His vitals were unremarkable except for a temperature of 39.2°C. On examination, he was pale with bitemporal muscle wasting. Small aphthous ulcerations were noted on the buccal mucosa. The rest of his physical exam was unremarkable. He spiked fevers up to 39.2°C daily. Labs revealed transaminitis, severely elevated inflammatory markers, anemia of chronic disease, monocytosis, hypertriglyceridemia, and an elevated soluble IL-2 receptor.

Computed tomography scan of the chest, abdomen, and pelvis revealed scattered tiny pulmonary nodules but no lymphadenopathy, hepatomegaly, or splenomegaly. Positron emission tomography scan showed diffuse low-level increased metabolism throughout the lungs suggestive of an inflammatory process. A comprehensive evaluation for underlying viral, bacterial, mycobacterial, fungal, and parasitic infection was negative. Autoimmune serologies including anti-nuclear antibody (ANA) and rheumatoid factor were negative. Flow cytometry of peripheral blood, cerebrospinal fluid assay and bronchoscopy with bronchoalveolar lavage were normal. Bone marrow biopsy revealed mildly hypercellular marrow with increased histiocytes with occasional hemophagocytic features, suggestive of a reactive process. Liver biopsy revealed extramedullary hematopoiesis with no evidence of malignancy.
Ultimately, a diagnosis of ASD was made given ongoing fevers, pharyngitis, transaminitis, negative serologic markers and severe systemic inflammation of unknown cause. He was started on IV methylprednisolone with immediate resolution of his fevers. Due to worsening transaminitis, anakinra (interleukin-1 receptor antagonist) was added, which led to improvement of his liver function abnormalities, inflammatory markers, and fatigue.

**Discussion**

The Yamaguchi criteria, which is the most widely used criteria for ASD, requires the fulfillment of 5 or more criteria with the presence of at least 2 major criteria. These criteria were developed to identify patients for inclusion in clinical trials but lack the sensitivity to identify all patients with disease. This was seen in our patient who had clinical findings consistent with ASD alongside notable improvement with standard therapy but however didn’t meet full classification criteria.

**Conclusion**

This case highlights how the lack of specific symptoms and serological markers in ASD can misguide and delay the diagnosis.

**Conflict of Interest and Financial Disclosure**

The authors do not have any disclosures to make.
Influenza Vaccination Rates Among Patients with Rheumatoid Arthritis and Systemic Lupus Erythematosus on Immunosuppressive Therapy

Authors
Judith Aniekwena, MD; Keerthi Padooru, MD; Titilope Olanipekun, MD, MPH

Affiliation
Morehouse School of Medicine, Department of General Internal Medicine, Atlanta, GA

Background
Influenza infection constitutes a significant cause of morbidity and mortality in patients living with Systemic Lupus Erythematosus (SLE) and Rheumatoid Arthritis (RA). The risk of influenza-related morbidity and mortality is higher in these patients due to impaired immune responses and the use of immunosuppressive therapy. Therefore, the advisory committee on vaccination practices recommend annual influenza vaccination to improve clinical outcomes in these patients. There is limited data on Influenza vaccination rate in patients with SLE and RA on immunosuppressive therapy.

Methods
Data was obtained from the electronic medical record system (EMR) of one of the ambulatory arms of Grady Memorial Hospital, Atlanta, GA. Patients with Systemic Lupus Erythematosus and Rheumatoid Arthritis who visited the clinic during the influenza season from October 1, 2018 to May 31, 2019 were included. Patients were asked to complete a questionnaire on demographics, influenza vaccination status, use of immunosuppressive therapy, duration of immunosuppressive therapy use, method of influenza awareness and smoking status. Data was confirmed on the EMR. MS excel and SAS 9.4 software were used for data management and analysis.

Results
A total of 83 patients were included in the study. 62.6% of the population had Rheumatoid arthritis while 37.4% had SLE. All patients were on immunosuppressive medications which included methotrexate, leflunomide, cyclosporine, mycophenolate mofetil and azathioprine. 78.3% were female and mean population age was 53.5 years. The patient population was predominantly African American (91.6%) and the educational level of majority of the participants (41%) was high school. 77% of the participants were non-smokers.

The most common setting for vaccination was during a primary care doctor's visit (66.1%; n=37/56) compared to 32.1% (n=18/56) who received vaccination at their Rheumatology clinic visit.
The overall flu vaccination coverage was 67.5% which is higher than the CDC reported 2018-2019 national coverage rate of 46.3% in patients with high risk conditions. 67.3% of the RA population (n=35/52) received the influenza vaccine while 67.7% of the SLE population (n=21/31) received the influenza vaccine. For the 32.5% of the total population that declined the flu shot, perceived lack of efficacy of the influenza vaccine was the most cited reason (37%), followed by aversion to all vaccines generally.

**Conclusion**
Our study demonstrates that the vaccination coverage rate among RA and SLE patients on immunosuppressive therapy is higher than the CDC reported coverage rate in high risk conditions generally. Given the lower rate of awareness provided by Rheumatologists about the flu shot in comparison to primary providers, we propose more patient education on the part of Rheumatologists, particularly on the perceived lack of inefficacy of the influenza vaccine. More studies should evaluate contributing factors to the relatively high influenza coverage rates seen in RA and SLE patients.

**Conflict of Interests or Financial Disclosures**
The authors do not have any conflict of interests or financial disclosures to make.
Clinical Pearls of Atypical Hemolytic Uremic Syndrome

Kesler Bourgoyne, MD, MS[1], Anusha Jogimahanti, MD[1], Laura Gulotta-Parrish, MSY-2, MS[1], Matthew Malus, MD[1], Richa Dhawan, MD, CCD[2], James Morris, MD[2]. Louisiana State University Health Shreveport – Shreveport, LA.
[1]Department of Internal Medicine, [2]Center for Excellence in Arthritis and Rheumatology

Introduction:
Atypical hemolytic uremic syndrome (aHUS) is a rare disease belonging to a category of diseases known as thrombotic microangiopathies. aHUS is characterized by the triad of microangiopathic hemolytic anemias (MAHA), thrombocytopenia and acute renal failure (ARF). Thrombotic microangiopathies include thrombotic thrombocytopenic purpura (TTP), which is often clinically mistaken for aHUS. The diagnosis of aHUS relies heavily on the recognition of previously listed clinical manifestations along with distinction from other MAHA. This case additionally highlights the importance of early recognition for initiation of Eculizumab.

Case Report:
We present a case of a 61-year-old African American lady with a past medical history significant for cutaneous lupus, type 2 diabetes mellitus and hypertension who presented to the emergency department for left-sided chest pain associated with intractable nausea and vomiting. Inpatient cardiac work-up negative for acute coronary syndrome. Due to concern for possible lupus flare, empiric glucocorticoid therapy was initiated. Labs significant for C3/C4 normal, dsDNA antibody negative, significant thrombocytopenia, elevated creatinine from baseline, decreased hemoglobin, elevated lactate dehydrogenase, decreased haptoglobin and schistocytes visualized on peripheral smear. Concern ITP versus TTP. Due non-response to glucocorticoids, TTP was highest on the differential. Plasma exchanges initiated, and glucocorticoids were tapered. Pt did not tolerate plasma exchanges initially. With two separate 14-day rounds of plasma exchanges, there was an improvement in thrombocytopenia. Unfortunately, patient’s status began rapidly deteriorating. Due this change, plasma exchanges were discontinued. Patient began showing signs of ARF without resolution and thus hemodialysis was initiated by Nephrology. At this point, differential broadened with aHUS being lead. Hematology was consulted and recommended genetic testing for aHUS to be sent to reference lab and initiation of Eculizumab. Genetic testing confirmed homozygous deletion apparent whole gene deletion of the CFHR1 gene, a known genetic association of aHUS. Eculizumab required prior authorization and outpatient initiation for insurance coverage. Patient clinically improved and was discharged to a long-term treatment facility for outpatient initiation of Eculizumab and rehabilitation services. Unfortunately, patient passed prior to initiation of Eculizumab.

Discussion:
This is a typical case of aHUS characterized by the triad of MAHA, thrombocytopenia and ARF. Genetic testing confirmed homozygous deletion apparent whole gene deletion of the CFHR1 gene, a gene associated with regulation of the complement system and thus immune system. Gene deletion of CFHR1 is one of the known genetic association aHUS characterized in
literature and clinically correlated. aHUS has overlapping signs and symptoms with TTP. Prompt clinical differentiation is key for prompt initiation of optimal medical therapy. While plasma exchanges can be used to treat aHUS, Eculizumab has been shown to improve clinical outcomes. This case also highlights the importance of social work and case management in providing adequate prompt treatment of patients.

**Disclosures:**

Authors have no conflicts of interest to disclose.
Idiopathic Inflammatory Myopathy Complicated by Respiratory and Renal Failure

Authors:
Brian Bui, MD
John Daliva, DO

Conflict of Interest Disclosure:
The authors declare that there is no conflict of interest.

Background:
Polymyositis and dermatomyositis are characterized by chronic inflammation and proximal muscle weakness. Diagnosis is supported with clinical, lab and muscle biopsy findings. This is a unique case in which a young patient’s hospital course was complicated by acute hypoxic respiratory and renal failure.

Case Description:
A 23 year old caucasian male with no past medical history initially presented to our hospital with a chief complaint of shortness of breath, subjective fevers, body aches, and polyarthralgia for the past 2 weeks.

On admission the patient was found to be febrile, tachycardic and tachypneic, however was normotensive and saturating well on room air. Chest radiograph demonstrated aspiration pneumonia, thus he was admitted for treatment of sepsis secondary to pneumonia with polyarthritis likely secondary to autoimmune etiology versus reactive arthritis. Initial labs significant for a leukocytosis of 33.5 with hyponatremia of 130. The creatinine on admission was found to be 0.8. On physical exam the patient presented with mildly diminished breath sounds bilaterally and +5/5 strength in all extremities with diffuse joint tenderness.

Two days into his hospitalization the patient was found to be increasingly tachypneic with acute respiratory decompensation requiring intubation and ICU (intensive care unit) admission for acute hypoxic respiratory failure. The arterial blood gas demonstrated PaO2 of 57 at an FiO2 of 45. During his ICU course the patient was found to be anuric with an acute kidney injury of creatine at 3.3. Creatine kinase was found to be significantly elevated at 16,000. Nephrology was consulted and the patient was started on hemodialysis. Lab studies including ANA, Jo-1 antibody, SS-A/SS-B, SM antibody and aldolase returned as positive/elevated suggesting an
autoimmune picture. Muscle biopsy which demonstrated morphologic features consistent with polymyositis. The patient was started on a course of prednisone and mycophenolic acid in addition to methotrexate and folic acid. Minimal improvement in creatine kinase prompted a course of intravenous immunoglobulin at 400mg/kg for five days and rituximab 1g every 14 days. There was interval improvement in the patient’s creatinine of 6.1 to his baseline of 0.6 thus no longer requiring hemodialysis. His creatine kinase levels downtrended down to 1200. He was subsequently extubated and transferred to an acute rehab facility where he continued physical therapy to improve his functional status.

Conclusion:

This case illustrates the course of a young 23 year old patient whose hospital stay was complicated by multiorgan failure requiring a multidisciplinary approach. The patient is outside the age for the diagnosis of juvenile idiopathic inflammatory myopathy yet does not meet the average age for adult onset. Review of the literature demonstrates value in using different autoimmune markers in juveniles versus adults in predicting patient clinical symptoms and outcomes.
TO BE, OR NOT TO BE

Swathi Chlasani, MD; Samina Hayat, MD; Sarwat Umer, MD

LSU Health Sciences, Shreveport, LA

Financial Disclosure: None

Introduction:
Anti-neutrophil cytoplasmic antibody-associated vasculitis is a necrotizing vasculitis that predominantly affects medium to small sized vessels. It includes granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), including renal-limited vasculitis (RLV), and eosinophilic granulomatosis with polyangiitis (EGPA). Initial immunosuppressive therapy in GPA and MPA typically consists of glucocorticoids + Cyclophosphamide or Rituximab.

Case:
56-year-old African-American female presented to emergency room with nausea, vomiting for 2 days. Physical examination was unremarkable. Diagnostic work up was significant for BUN of 19 mg/dl and Creatinine-2.2 mg/dl. Urine analysis showed RBC >100, WBC 61, occasional granular and RBC casts. Urine protein/creatinine was 2.8. Serum Myeloperoxidase antibody was > 8. Renal biopsy showed Pauci-immune necrotizing and crescentic glomerulonephritis. She received Solumedrol 1 gram for 3 consecutive days and sent home on oral Prednisone 60 mg daily. She received IV Rituximab 1000 mg seven days post discharge.

Eight days after IV Rituximab, she was presented to Emergency department again with bruising and mild hemoptysis and blood in urine. She was found to be having platelets of <2 k/ul (262 k/ul during previous admission). Diagnostic work up showed 1-3 schistocytes on peripheral smear, normal ADAMS 13 activity, elevated D dimer but normal fibrinogen levels. Heparin induced platelet antibodies and serotonin assay were negative. Acute hepatitis panel, HIV, CMV, EBV, serum Histoplasma and Blastomyces antibodies and blood cultures were negative. B12, Folate and antiplatelet antibodies were negative. Patient received platelet transfusions twice. Immediately post-transfusion showed appropriate raise in the platelet count but repeat CBC in 2-3 hours post-transfusion showed drop in platelets to undetectable levels.

She was diagnosed with immune mediated thrombocytopenia presumptively Rituximab induced after ruling out other causes of thrombocytopenia. We have had tried pulse dose steroids, plasma exchange and IVIG which didn’t help. Patient got IV Cytoxan 500 mg/m2 with Mesna. Day 2 post Cytoxan, platelet count started to improve and reached 184 k/ul on day 8 post infusion.
Discussion:

Rituximab is a chimeric antibody, composed of both mouse and human portions. It is an immunoglobulin G1 (IgG1) monoclonal antibody (mAb), which targets CD20. Thrombocytopenia is a known adverse drug reaction of rituximab, which was identified during the preregistration trials, but knowledge on incidence, mechanism and risk factors in clinical practice is limited.

Thrombocytopenia due to Rituximab can be due to bone marrow suppression or immune mediated [drug induced Thrombocytopenia (DIT)]. DIT incidence is about 10 cases per million inhabitants per year in the United States and Europe. Although DIT is uncommon, it can have serious and sometimes even fatal consequences. No treatment was ever proposed in literature how to treat Rituximab induced immune mediated thrombocytopenia. We successfully treated our patient with fatal thrombocytopenia from Rituximab with IV Cytoxan.
Introduction:
Relapsing polychondritis is a rare autoimmune disease of unknown etiology causing episodic inflammation of cartilaginous structure and sometimes non-cartilaginous structures. The peak age of onset is between 40 to 50 years, but cases have been observed in children and older adults. It occurs with equal incidence in both sexes and all racial groups. The common manifestations include recurrent inflammation of the auricular, nasal, ocular, laryngo-tracheal, costal and articular cartilage. In this case, we are emphasizing on recurrent auricular chondritis post MOHS excision surgery for Basal cell carcinoma.

Case:
80-year-old Caucasian male with significant past medical history of Coronary artery disease, Diabetes Mellitus type II and recurrent skin cancers status post excision was admitted to hospital with chief complaint of recurrent right auricular chondritis. He had right helical rim nodular basal cell carcinoma status post MOHS excision 5 months ago. He started experiencing recurrent swelling of right ear helix post-surgery. He had 2 hospital admissions and finished several courses of Intravenous and oral antibiotics. He had biopsy of the right auricle done twice in the past which showed benign skin with focal mild/moderated dysplasia, negative of malignant cells. Previous cultures were negative cultures negative for any organisms.

He was started on IV Cefepime and Decadron 8 mg every 8 hours and Rheumatology was consulted for possible relapsing polychondritis. On physical examination, vitals were within acceptable limits. He has right cauliflower ear with erythematos, swollen and tender helix. Right ear lobule was spared. On laboratory investigations, WBC was 14,000, Erythrocyte sedimentation rate was 14, C-reactive protein <0.29. Urine analysis and chest X-ray were unremarkable. Antineutrophil cytoplasmic antibodies, Type II collagen antibodies were normal. Repeat debridement and biopsy was done. Intra operatively a lot of purulence and dead cartilage tissue was noticed. Histo-pathology was consistent with *chondrodermatitis nodularis helicis*.

4 days later, culture came back to be positive for Aspergillus flavus. On further inquire, patient reports that he works in a corn field and sprays Aflo-guard, a bio-control agent that has Aspergillus flavus as active ingredient. We discontinued Cefepime, tapered Steroids and started him on Po Voriconazole 400 mg every 12 hours for 2 doses followed by 200 mg every 12 hours for 30 days. Patients symptoms started improving 2 days after starting the antifungals.

Discussion:
Relapsing polychondritis is a rare autoimmune disease causing recurrent inflammation of cartilaginous structures, commonly affecting ears, nose, eyes, laryngo-tracheal, costal and articular cartilage. When the presentation is typical, the biopsy is not required. But, early disease is mimicked by many conditions like trauma, chemical injury, malignancy and infections (especially subacute and chronic infections) requiring biopsy and culture.
Confused and Itchy: An Unusual Presentation of Systemic Lupus Erythematosus in an Elderly Caucasian Male

Authors: Ambreesh Chawla, MD1: Rheumatology Fellow
Richard Henriquez, MD1: Internal Medicine Resident
Dominique Brouin, MD1: Internal Medicine Resident
Seema Frosh, MD1: Junior Rheumatology Faculty
Sujatha Vuyyuru, MD1: Senior Rheumatology Faculty

1Orlando VA Medical Center, University of Central Florida College of Medicine – Orlando, FL
Contact information: ambreeshchawla@gmail.com; 440-213-6170
No funding; No disclosures.

Systemic Lupus Erythematosus (SLE) is a rheumatic disease characterized by autoantibody response to nuclear and cytoplasmic antigens, leading to damage in various tissues. SLE usually affects women between the ages of 15-45 and its prevalence is 3-4x higher in non-Caucasians. Neuropsychiatric lupus (also known as ‘CNS lupus’) refers to the various neurologic manifestations in SLE. We present a unique case where acute confusional state and concomitant biopsy-proven subacute cutaneous lupus (SCLE) led to a new diagnosis of SLE in an elderly Caucasian male.

A 77-year-old Caucasian man with type II diabetes presented to our ED with a one-month history of altered mental status in the setting of an acute rash. One month before his ED visit, he developed a pruritic rash (raised erythematous lesions with scaling) over his chest & back (sparring his face/oral mucosa). There were no known infections, sick contacts/travel preceding onset of the rash. According to his wife, around the time the rash manifested, her husband became confused with increased forgetfulness & irrational behavior. In the ED, the patient was AAO x 1 and review of systems were negative for fever, oral ulcers, arthralgias, raynaud’s phenomenon, dry eyes/mouth. Labs showed mild leukopenia with normal creatinine, UA and LFTs. Labs also revealed ANA (1:1280, homogenous), DsDNA 671, anti-chromatin >8, hypocomplementemia (C3: 58, C4: 8), elevated ESR/CRP and (+) antiphospholipid antibody titers. All the following labs were unremarkable: SS-A/B, Smith, RNP, JO1, ribosomal P, CPK, aldolase & myositis related autoantibodies. Complete workup of infection/malignancy were negative. MRI-brain depicted diffuse involutional parenchymal changes without any acute intracranial process. MR-A (brain) was unrevealing. Lumbar puncture (LP) showed elevated CSF WBC, protein, and glucose. CSF IgG index was elevated in the presence of oligoclonal bands. All the following CSF tests were negative: paraneoplastic panel, infectious markers, NMDA and ribosomal P. Histopathology from a skin biopsy delineated subepidermal lymphocytic infiltration and increased dermal mucin deposition, consistent with SCLE. The patient received pulse IV methylprednisolone which markedly improved his mentation. He was discharged on prednisone and hydroxychloroquine.

The American College of Rheumatology has recognized 19 syndromes under the umbrella of neuropsychiatric lupus, including ‘acute confusional state’. While several modalities are used to diagnose neurologic disease in SLE, oftentimes such tests are non-specific. In our case, given that serologies were suggestive of active SLE in conjunction with altered mention and elevated CSF IgG index/oligoclonal bands, we made a diagnosis of acute confusional state secondary to SLE. After discharge, his rash cleared, and his mentation continued to improve. Our case demonstrates a unique diagnosis of SLE in an elderly Caucasian male. While SLE is rare in such patients, it should be considered in those with an acute cutaneous eruption and/or abrupt change in mentation regardless of age, race or gender. This may aid in early diagnosis and prompt immunosuppressive therapy to prevent long-term complications.
Favorably Unfavorable: Interstitial Lung Disease as the Initial Manifestation of Anti-Mi-2 Positive Dermatomyositis

Case Report: Clinical
Authors: Ambreesh Chawla, MD; Rheumatology Fellow
Lance Feller, MD; Junior Rheumatology Faculty Attending Physician
Sujatha Vuyyuru, MD; Senior Rheumatology Faculty Attending Physician

1University of Central Florida College of Medicine – Orlando, FL
Contact information: ambreeshchawla@gmail.com; 440-213-6170
No funding; No disclosures.

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by muscle weakness and distinctive skin findings. Interstitial lung disease (ILD) is a well-recognized complication of DM and is seen in at least 10% of cases. The presence of myositis-specific autoantibodies (MSA’s) are felt to predict clinical manifestations (including ILD) and prognosis in DM. Of the MSA’s, anti-Mi-2 (which targets the chromodomain helicase DNA binding protein, CHD4) is classically associated with skin and muscle features of DM, and to be protective for ILD with a favorable prognosis (5-year survival > 90%). We report a unique case of DM (with positive anti-Mi-2) in a young woman initially presenting with dyspnea secondary to ILD.

A 25-year-old Hispanic woman with a one-year history of Raynaud’s phenomenon (RP) presented to our clinic for initial evaluation. Outside of RP, she had no prior medical history and was not on any medications. Review of systems was positive for dyspnea on exertion. On exam, the patient demonstrated severe asymmetric digital blanching with sequential color changes and evidence of an unhealed digital ulceration. Nailfold capillary microscopy revealed dilated loops and avascularity. Auscultation of the lungs demonstrated fine inspiratory velcro-like crackles. Labs revealed ANA 1:640 (dual pattern: homogenous, speckled) with elevated CPK, Aldolase, RF, ESR, CRP and mild transaminitis. All the following serologies were unremarkable: DsDNA, Smith, RNP, Chromatin, SS-A/B, ACPA, antiphospholipid antibody panel. Urine studies were negative for occult blood and protein. Further testing of DM and scleroderma-related autoantibodies were unremarkable with exception of the anti-Mi-2 antibody. PFT’s revealed a decreased FEV1 (71% predicted), FVC (65% predicted), TLC (80% predicted) and markedly diminished DLCO (45% predicted). High resolution CT-Chest revealed bilateral subpleural reticulation and honeycombing, consistent with usual interstitial pneumonia (UIP). During subsequent clinic visits, the patient experienced proximal muscle weakness (UE > LE) and developed a cutaneous eruption over the posterior shoulders and proximal UE. She was maintained on a combination of high dose prednisone & mycophenolate, in addition to pharmacotherapy for RP.

The presence of MSA’s directed against cytoplasmic proteins, ribonucleoproteins and other nuclear antigens can be helpful for both diagnostic and prognostic purposes in DM. The following MSA’s have been well described in the literature for their association with ILD: anti-synthetase antibodies (JO-1, PL-7, PL-12, OJ, EJ, KS, ZO), anti-MDA5 (anti-CADM140) and anti-PM-Scl antibody (polymyositis/scleroderma overlap). Anti-Mi-2 can be seen in both adult and juvenile DM and is usually associated with cutaneous disease, myositis with good response to treatment and a lack of ILD. Our case demonstrates a noteworthy presentation where UIP-ILD coincides with the presence of a ‘favorable’ MSA (anti-Mi-2). While antibody testing is helpful for both diagnostic and prognostic purposes, our case delineates the gravity of screening for less frequently involved organs irrespective of a patient’s MSA profile.
Culture Negative Mechanical Valve Endocarditis Complicated with Corticosteroid Induced Mobitz Type II AV Block in a Patient with SLE

Authors: Harman Fervaha MD, Samina Hayat MD

The authors have no disclosures.

We present a case of a 31 yo F with a PMH of SLE, DVT/PE with protein S deficiency and mitral valve replacement due to culture negative endocarditis who presented to the ED feeling unwell for 1 week. She was admitted 2 weeks prior for fever, rash and elevated troponin. During initial admission, work-up was unrevealing, and patient was discharged. She returned with tachycardia, hypotension, and altered mentation. Troponins were elevated and continued to rise. She was treated as septic shock in the ICU and responded well to treatment. A transesophageal echocardiogram showed vegetations on anterior and posterior suture lines of her mechanical mitral valve. Like her previous endocarditis, her blood cultures were negative. Pathology of the native mitral valves showed myxoid degeneration and fibrinopurulent exudate; culture was negative for bacteria or fungi. This was concerning for Libman-Sacks endocarditis as the patient had no history of IV drug use and never had bacteremia. Infectious disease evaluated the patient and determined the patient had nonbacterial thrombotic endocarditis or culture negative endocarditis and recommended 6 weeks of IV antibiotics. Rheumatology was consulted to evaluate for SLE flare as the cause of her symptoms. Additional workup showed low C3 complements which were normal on previous admission and SLEDAI of 11. She was started on methylprednisolone 1000mg daily IV for 2 days. She developed EKG abnormalities with progressive decreases in heart rate. Her EKG progressed from normal sinus rhythm to sinus bradycardia to Mobitz type I and Mobitz type II AV block. Electrophysiology was consulted and placed a temporary pacemaker. Of note, 30-45 minutes after receiving both doses of IV methylprednisolone, she developed bradycardia and AV block. She was transitioned to oral prednisone and subsequently the episodes of bradycardia resolved, her rhythm normalized, and her pacemaker stopped firing. The pacemaker was removed, and patient was discharged with IV antibiotics, oral prednisone and close follow up.

A comprehensive literature review was conducted to identify publications describing culture negative endocarditis on a mechanical valve as well as the association between arrhythmia and corticosteroid use. Reports suggest culture negative endocarditis in patients with mechanical valves affect the junction of the tissue and prosthesis at the suture site as well as tissue destruction around the prosthesis. Also, insertion of a mechanical valve can lead to formation of a pannus, which then can lead to thrombus and vegetation formation on the pannus. Pathology and culture of these vegetations can show bacteria such as Coxiella, Bartonella, and Chlamydia species but often are negative. There are no reports of Libman-Sacks endocarditis affecting a mechanical valve. High dose corticosteroids have been reported to cause sinus bradycardia in several cases, but none have reported high degree AV block as seen in our patient. It is possible the high dose steroid induced bradycardia and in the setting of endocarditis triggered the AV block. It is important to know the adverse effects of medications as some effects can be more harmful than the condition being treated.

References


Pyoderma Gangrenosum, a Rare Dermatologic Complication of IgA Vasculitis

Perry Fuchs, M.D.\(^{(1)}\), Meenakshi Sridhar, M.D.\(^{(1)}\), Theresa Nieman, M.D.\(^{(2)}\)

\(^{(1)}\)Internal Medicine Residency Program University of Arizona College of Medicine Phoenix, \(^{(2)}\)Phoenix Veterans Affairs Medical Center, Department of Rheumatology

Introduction: IgA Vasculitis (IgAV), also known as Henoch-Schönlein Purpura, is a small vessel vasculitis characterized by IgA deposition with a 90% incidence in the pediatric population. The classic tetrad of IgAV is palpable purpura, abdominal pain, renal dysfunction, and arthralgia. This vasculitis is significantly more rare in the adult population. Morbidity and mortality in adults is typically due to gastrointestinal complications early in the disease and due to renal dysfunction later in the disease process. The following case demonstrates a recalcitrant dermatologic manifestation of IgAV that evolved into a difficult to treat dermatosis that severely impacted the patient’s quality of life.

Case: A 65-year-old male veteran presented with chief complaints of rash and abdominal pain. His physical exam was significant for diffuse, violaceous palpable purpura (Image 1). A comprehensive diagnostic evaluation was completed and most significant for a skin biopsy demonstrating leukocytoclastic vasculitis and direct immunofluorescence with IgA deposition, diagnostic for IgAV. Prednisone therapy was started without further progression of the purpura initially. The considerable surface area and wound care burden of the purpuric rash required transfer to a VA longterm care facility for said wound care and pain management. Over the course of weeks, areas of the purpuric rash on the bilateral lower extremities evolved into deep ulcerations with rolled borders, clinically consistent with a diagnosis of Pyoderma Gangrenosum (PG) (Image 2). Therapy with mycophenylate mofetil (MMF) was started in addition to the prednisone therapy. In the absence of other identifiable etiologies, including a negative malignancy work-up, the PG was attributed to the IgAV. In addition to intensive wound care, the deeply ulcerated lesions required dose escalation of prednisone and MMF before any significant resolution was appreciated. Seven months after diagnosis, the PG wounds responded adequately in order to transition his wound care to home-based healthcare services.

Discussion: This case illustrates a rare dermatologic complication of IgA Vasculitis in an adult, Pyoderma Gangrenosum. There is a dearth of reported cases in the literature of the development and management of Pyoderma Gangrenosum in adult patients with IgA Vasculitis. This case also serves as a reminder of potential severe consequences of IgAV on patient quality of life.

There are no conflicts of interest to disclose.
**Immunosuppression in a case of Staphylococcus associated Henoch Schönlein Purpura.**

Authors: Priyatha Garlapati MD; Sai Krishna Koyoda MD

LSU Health Sciences Center, Shreveport, LA; Monmouth Medical Center, NJ

The authors have no disclosures.

**Background:**

Fifty Four Year old female with Uncontrolled Diabetes Mellitus and Hypertension was admitted for left foot cellulitis. Blood cultures and wound cultures were positive for methicillin resistant staphylococcus aureus. Antibiotic therapy was initiated with vancomycin and cefepime. However, she developed generalized erythematosus macular rash on the upper extremity and trunk that progressed to lower extremities. Antibiotics were switched multiple times from piperacillin-tazobactam to ampicillin-sulbactam to ertapenem as the working diagnosis at this point was a drug induced reaction. Despite these changes, the rash progressively worsened and evolved into a purple non-blanchable palpable rash. She also developed abdominal pain, hematochezia, oliguria and hematuria. Differential diagnoses was considered to be acute interstitial nephritis versus acute glomerulonephritis as either post-infectious glomerulonephritis or IgA nephropathy/ Henoch-Schonlein purpura.

Urinalysis positive for blood and protein but negative for WBC and urine eosinophils, hence acute interstitial nephritis was ruled out. ESR 84 mm/hour, CRP 294mg/L. Her IgA level was elevated at 542mg/dL. Serum complements were normal, ruling out post-infectious glomerulonephritis. Other investigations were unremarkable including hepatitis, cryoglobulinemia and other autoimmune workup.

Due to worsening of kidney function, she was initiated on daily IV methylprednisolone 500mg for 3 days followed by daily oral prednisone after second set of blood cultures were negative. On third day of receiving methylprednisolone her rash, hematuria, diarrhea and kidney function significantly improved. Skin biopsy showed leukocytoclastic vasculitis and Kidney biopsy showed diffuse mesangial and focal segmental endocapillary proliferative glomerulonephritis consistent with IgA nephropathy.

With these findings, a diagnosis of Staphylococcus-associated HSP was made. She developed electrolyte imbalances and volume overload that necessitated hemodialysis. She was later discharged in stable condition with slow tapering prednisone doses and IV cefazolin during dialysis days. Her creatinine significantly improved to baseline.

**Conclusion:**

Emerging terminologies are being established to classify glomerulonephritides. Among the innumerable medical dilemmas, infection-related glomerulonephritis is becoming popular, a reason being the need to determine as to whether antibiotics is to be given or immunosuppressants. Studies that found antibiotics as the main management without steroid would call it as “HSP-like”, or “glomerulonephritis mimicking IgA nephropathy”. The rationale behind this proposition is valid as these studies found that the use of immunosuppressants did not provide improvement and had the risk of overwhelming sepsis. However, studies mention that the role of antibiotics is “undoubtedly essential”, but the controversy lies on the benefit of immunosuppressants. Truthfully told, though there was an improvement seen in our case, this is far from sufficient to establish a guideline of therapy.
An Uncommon Symptom Constellation in Hidradenitis Suppurativa

Anirudh Goyal, BS, Sravani Penumarty, MD, Reena Khianey, MD, Tina Brar, MD, and Eugenio Capitle, MD
Rutgers New Jersey Medical School, Newark, NJ
No disclosures to report
Category: Fellow and Resident Poster Session - Case Report
Author Classification: Medical Student

Background:
Hidradenitis suppurativa (HS) is a chronic inflammatory process of apocrine glands causing inflammatory and suppurative skin lesions such as painful nodules, abscesses and scarring. HS has been linked with extra-cutaneous manifestations like spondyloarthropathies, conjunctivitis, inflammatory bowel disease and osteitis syndrome. We describe a rare presentation of a HS patient complaining of back pain who was found to have multiple extracutaneous manifestations including fevers, hemodynamic instability, osteitis, pleuro-pericarditis, hepatosplenomegaly and splenic lesions.

Case:
A 16-year-old female with history of HS on clindamycin for 4 years presented with progressive sharp mid-lower back pain requiring a cane for ambulation for the past few months. She had no response to NSAIDs or other over the counter medications. She reported an unintentional 50 pound weight loss in the last 8 months and reduced appetite. She denied any fevers, night sweats, trauma or neurologic complaints. Prior imaging before admission was pertinent for back MRI revealing lytic and blastic lesions in her lower thoracic and lumbar spine. Bone marrow biopsy was negative for malignancy. Physical exam revealed purulent draining HS lesions in axillae, breast and groin region with corresponding hyperpigmentation and pustulosis on legs. Back exam revealed limited range of motion with no point tenderness. Hospital workup for autoimmune, infectious and malignant etiologies was unremarkable. ESR and CRP were 117/105 respectively with rest of routine labs within normal limits. T and B flow cytometry ruled out malignancy. CT chest and abdomen revealed pleuro-pericarditis, hepatosplenomegaly and multiple splenic lesions. MRI of the spine showed edema around sacroiliac joints.

Over the next several days in the hospital, the back pain worsened. She became hemodynamically unstable and spiked a fever to 103°F. She was started on broad spectrum antibiotics but subsequent blood and urine cultures were negative for infection. After infection was ruled out, she was started on IVIG for 4 days with a low dose of prednisone to treat a suspected autoimmune process. Soon after this therapy, the patient stabilized and the back pain improved tremendously to the point she no longer required a cane for ambulation. She was sent home on prednisone and methotrexate.

Discussion and Clinical Implications:
Differentials for back pain in HS include osteomyelitis, spondyloarthropathy, osteitis, or SAPHO syndrome. As the patient showed significant symptom resolution on IVIG and steroids, an autoimmune etiology was high on our differential, which was further confirmed by the MRI showing osteitis. She was sent home on methotrexate and prednisone and gradually transitioned to monotherapy with adalimumab with a strong response. The patient no longer required the cane to participate in her daily activities. She did not have a recurrence of symptoms on this treatment.

This presentation of osteitis, pleuropericarditis, hepatosplenomegaly and pustulosis in the setting of HS has not been reported in the past. We should be cognizant of non-classical manifestations which can mask as malignancy or infection.

References:
Rapidly Progressive Glomerulonephritis (RPGN)- An Important Diagnostic Consideration for Patients Presenting with Bilateral Peripheral Neuropathy

Tanvi Goyal, MPH; Maria Alfonso-Jaume, MD; Deepak Malhotra, MD; Lance Dworkin, MD
University of Toledo College of Medicine
No disclosures to report
Category: Fellow and Resident Poster Session - Case Report
Author Classification: Medical Student

Background:
Rapidly progressive glomerulonephritis (RPGN) is a syndrome of renal failure occurring within a few weeks or months with a histological finding of necrotizing crescentic glomerulonephritis. Pauci-immune crescentic PICG is the most common etiology of primary RPGN. This case report features a patient with PICG whose initial presentation consisted of bilateral peripheral lower extremity neuropathy that preceded renal failure by 2 months.

Case:
A 60-year-old woman presented to her neurologist for bilateral dorsolateral numbness and tingling of the feet up to the level of the ankle. X-ray of the spine to investigate dorsolateral numbness was unrevealing. However, an incidental finding of pulmonary nodules prompted a lung biopsy which in turn revealed granulomatous inflammation.

Two months later, the patient presented to the emergency department with acute onset coarse tremors of the upper and lower extremities, was found to have acute kidney injury and was emergently hemodialyzed. The patient is a non-diabetic with no previous history of kidney disease. Medical history is positive for a 45 pack-year history of cigarette smoking and hypertension. The patient denied shortness of breath, cough, hemoptysis or any nasopharyngeal symptoms. Physical exam showed asterixis of upper and lower extremities, dorsolateral numbness and tingling on dorsal foot, and lower extremity strength of 5/5. Lungs were clear to auscultation bilaterally.

Creatinine on initial presentation was high at 9.23 mg/dL and blood urea nitrogen (BUN) was high at 92 mg/dL. Serum sodium was low at 128 meq/L, serum potassium was high at 5.3 meq/L, and serum phosphorus was high at 7.0 mg/dL. GFR on presentation was low at 4 ml/min/1.73m². Hemoglobin was low at 8.7 g/ dL. Urine analysis was significant for hematuria. Cytoplasmic ANCA (c-ANCA) was positive at 1:160 and serine protease 3 (PR3) was high at 787 AU/mL. Kidney biopsy confirmed a diagnosis of crescentic glomerulonephritis suggestive of granulomatous angiitis. The anti-GBM antibody was negative. RPR and B12 tests were unremarkable.

The patient received hemodialysis, IV and oral steroids, cyclophosphamide, rituximab, sulfamethoxazole-trimethoprim prophylaxis, and 5 treatments of plasmapheresis before being discharged.

She was readmitted the same day due to acute shortness of breath. She was diagnosed with Takotsubo stress cardiomyopathy with a 20-25% ejection fraction and was discharged 9 days later with a life-vest and follow-up with nephrology, cardiology, and rheumatology.

Discussion and Clinical Implications:
Clinicians should consider vasculitis in patients presenting with more than one organ involvement, as highlighted in this case where both the nervous and the pulmonary systems were involved. Evaluation of the renal function and the urinalysis early in the course of the patient’s clinical presentation would have prevented the patient from developing ESRD and becoming dialysis dependent. RPGN has a rapid clinical course and an earlier diagnosis would have led to better treatment and improved prognosis, decreasing both patient morbidity and mortality.

References


An unusual case of giant cell arteritis (GCA) in a 75 year old gentleman

Simon Hackett¹, Laura Coates¹

¹Botnar Research Institute, University of Oxford, Oxford, UK

A 75-year-old man presented with an emergent inflammatory arthritis with a background of giant cell arteritis (GCA) and psoriasis. On examination the gentleman had low-grade synovitis in his MCP and PIP joints bilaterally. He was started on subcutaneous methotrexate (MTX) with some therapeutic effect, along with leflunomide. At his initial presentation, his GCA was asymptomatic and was being managed with 5 mg PO prednisolone.

Subsequently, the patient represented with visual disturbance and, upon investigation, was noted to have anterior ischaemic optic neuropathy (AION), with evidence of aortitis and intracranial vasculitis on MRI. On further discussion, the patient also described a history of transient ischaemic attacks (TIAs). An MRI gadolation scan demonstrated persistent left-sided high-grade distal ICA stenosis, secondary to GCA. The patient’s dose of prednisolone was increased to 40 mg PO with appropriate subsequent dose-reduction, along with 20 mg SC methotrexate. His GCA stabilized following treatment, although was complicated by drug-induced hepatitis secondary to leflunomide midsoring which was managed successfully as an inpatient. The patient remains largely asymptomatic in the context of his GCA.

I have no competing interests elsewhere and this abstract has not been submitted elsewhere
I have no conflicts of interest to disclose

A Case Report of Skin Thickening: Scleroderma or Malignancy?
Seniya Hanif, MD; Angelia Mosley-Williams, MD; Anita Bishanci, MD
Department of Internal Medicine, Division of Rheumatology, Henry Ford Hospital, Wayne State University, Detroit, MI USA

Background
- We describe to you a case of diagnostic and therapeutic dilemma with symptoms attributed to both scleroderma and malignancy
- A unique subset of patients with temporal clustering of both entities
- A rare reported case where chemotherapy for malignancy resolves skin manifestations of scleroderma

History of present illness
- 61-year-old veteran presented with lightheadedness and dysphagia
- Associated with odynophagia, inability to swallow saliva, hoarseness, weight loss, progressive dyspnea, arthritis
- Initial symptom one year back at external hospital of Raynaud’s, and skin thickening
- Referred to Dermatology: biopsy showed morphea
- Rheumatology: MTX and prednisone: enrolled in tocilizumab trial for scleroderma

Physical exam
Skin exam: Sclerodactyly in both hands, skin thickening up to elbows, upper chest, both feet and legs, sparing abdomen and lower chest

Laboratory tests
- ANA 1:640 (speckled), SSA negative, SSB positive, Anti-Centromere negative, SCL-70 negative, Anti-dsDNA negative, RNP Polymersome 3 positive
- ESR/CRP in normal range

Hospital Course
- Endoscopic Bronchial Ultrasound and biopsy: Aggressive stage 3 Diffuse Large B Cell Lymphoma, Non-Hodgkin’s type
- Excluded from Tocilizumab Study, transferred to cancer center
- Started on EPOCH regime followed by bendamustine and Rituximab
- Completed four cycles of chemotherapy
  - In five months: dysphagia and dyspnea markedly improved
  - Skin thickening improved (from 2-3 mRSS to 1-2)

CT chest-post-chemotherapy:
- Marked improvement in previously noted mediastinal lymphadenopathy with residual mildly enlarged subcarinal lymph node

Imaging
- Axial left mainstem bronchus appeared narrow in caliber
- Diffuse mediastinal, axillary lymphadenopathy
- Deviation of the trachea to the right with mild narrowing of the distal portion of the left mainstem bronchus
- Hilar lymphadenopathy
- Pericardial effusion
- Biopsy could not be separated from posterior mediastinal lymphadenopathy
- Small amount of right pleural effusion

Exechocardiogram:
- Normal LV size and function EF 60-65%
- Mild LV hypertrophy, Grade 1 Diastolic dysfunction, RVSP 22 mm Hg
- Small circumferential pericardial effusion, no tamponade

Discussion
- Patients with scleroderma have been noted to have increased incidence of cancer compared with general population
- Risk could be secondary to
  - Damage from scleroderma (ILD or Barrett’s)
  - Cytotoxic therapy used for management
  - A Common inciting exposure
  - A genetic predisposition (increased risk of cancer in first degree relatives)
- Conversely, cancer therapies may trigger severe Raynaud’s or lead to exaggerated fibrosis (such as Biologics, Carboplatin, checkpoint inhibitors) and by radiation
- However, a subset has been described with concurrent onset of scleroderma and cancer, showing mechanical association of the two phenomena
- Shat et al noted at John Hopkins (1), a striking temporal clustering of cancer diagnosis with first clinical signs of scleroderma, noting unique molecular expression of RNA polymersome II in their cancer cell not noted in other scleroderma autoantibodies or in normal control
- They found a 5.68 fold increased risk of cancer within two years of scleroderma onset in those with positive RNP polymersome II Ab
- Temporal clustering with short scleroderma-cancer interval observed
- POLR3A is gene locus for RNA Polymersome III
- Mutated RNA polymersome II proteins are hypothesized to be immunogenic that initiate Anti-RNA polymersome III Ab response

Conclusion
- Cancer-induced autoimmunity may occur in a subset of patients with positive RNA polymersome III Ab
- Scleroderma may be paraneoplastic disease
- RNA Polymersome III Ab is a “red flag” that warrants more aggressive cancer screening
- In our patient, we proved that cancer therapy was effective in treating scleroderma
- A rare case report where skin tightening improved after chemotherapy for management of cancer

Outside Records
- CT Thorax without contrast: No ILD/Fibrosis, mediastinal mass that is well-defined with complex fluid internal density. Lesion is favored to represent esophageal duplication cyst. This may have mild mass effect in esophagus
- Skin Biopsy (left upper chest and right arm):
  - Hypertrophic scars with an underlying square-shaped derma, prominent dermal sclerosis, thickening of collagen bundles, loss of perineural adipose tissue. Diagnosis: scleroderma/morphea

References
I have no conflicts of interest to disclose.

Presenting Author: Soniya Hanif

Prevalence of OSA in lupus patients and effect of intervention with CPAP and its compliance on lupus disease activity in terms of its flares

Research proposal

1. Introduction and background

Sleep disturbances have been widely associated with chronic autoimmune conditions including SLE. In his study comprising of women with SLE, De Costa noted moderate to severe sleep impairment reported in 56% of his patients. This trend was seen in study conducted by Chandrasekhara et al who found a prevalence of sleep disturbances in 62% of lupus patients. More recently, Mirbagher et al assessed sleep quality in 77 women with SLE and concluded that poor sleep quality was common in women with SLE and significantly impaired their health related quality of life. Age, disease activity and psychological factors were determinants of sleep quality in their study. Moraleda et al noticed in their study of lupus women of poor sleep quality (both subjective and objective). Obstructive sleep apnea is most widely reported sleep disturbance. Kang et al calculated hazard ratio of developing autoimmune diseases in patients with OSA of 1.91 times greater for patients with OSA than with controls. On the other hand, Chen et al demonstrated hazard ratio of 1.51 in patients with autoimmune diseases to develop OSA in five years.

It is, therefore, evident from above studies of an association of poor sleep quality patients with lupus that could play a modulators role in clinical and psychological manifestations of the disease with effect on their quality of life. There seems to exist a bidirectional relationship where risk of OSA is increased in patients with lupus and lupus would, in turn, increase risk of development of OSA. Complex relationship can also be inferred where disease activity of one entity proportionately affects the other entity. OSA, being the most prevalent sleep disturbance, the management of which has drastically reduced its complications. The prevalence of OSA in lupus patients is currently not well defined. Additionally there have been no studies to to date to determine wether an intervention with CPAP for OSA in lupus patients would affect their sleep quality and sleep architecture, and that would, in turn, improve their quality of life in terms of disease flares and their associated symptoms.

We, therefore, aim for the following:

A. Determine the prevalence of OSA in our tertiary care hospital setting in patients with confirmed lupus

B. Measure the effect of intervention on disease activity: CPAP compliance on disease activity of lupus

2. Study hypothesis / objectives
A. Alternate hypothesis: there is a difference in prevalence of sleep apnea in patients with lupus

B. Alternate Hypothesis: There is a difference in CPAP compliance and number of lupus flares

Design
A. Observational study: cross sectional, prevalence surveys
B. Observational study: prospective cohort

3. Subjects: inclusion and exclusion criteria
A. Inclusion: All patients with dual diagnosis at the Henry Ford Health System of lupus and OSA from July 2015 to June 2019
   Exclusion: Discoid lupus
B. Inclusion: age < 18 years, confirmed diagnosed of SLE by Rheumatologist as per the American College of Rheumatology criteria, Confirmed OSA by sleep physician based on polysomnography, prescribed regular CPAP by Sleep physician for most of the night use, with stable pressures for three months prior to inclusion, stable disease activity as clinically judged by rheumatologist for at least two months before screening, either maintained with no medication or stable treatment regime of low dose < 15 mg of prednisone, antimalarials, NSAIDs, methotrexate, azathioprine, or mycophenolate mofetil. They should have positive ANA, and measurable anti dsDNA, or reduced C3 or C4 levels.
   Exclusion: discoid lupus, age > 75 years of age, other co-existent autoimmune diseases (Sjogren’s, Rheumatoid arthritis, myositis), fibromyalgia, chronic depression. Patients with active lupus nephritis requiring high dose glucocorticoids equivalent > 60 mg of prednisone or cyclophosphamide, or who had received plasmapharesis, IVIG, cyclosporine within six months of screening. Patients with active CNS lupus within six months of screening, history of renal transplant, evidence of clinically significant non SLE related acute or chronic disease of history of any serious infection within four weeks of study entry. Pregnant or nursing mothers, with adequate contraception in all participating patients

Methods
A. All patients meeting the inclusion and exclusion criteria will be analyzed for data gathering. The variables will include age, gender, race, duration of lupus, BMI, renal or extrarenal lupus (dichotomous variable), proteinuria, serologies (ANA, smith, dsDNA, C3,C4), co existing depression or fibromyalgia, OSA severity: AH1 (scores), Epworth sleepiness scores, CPAP use (number of hours, and/or as a discrete variable), immnosuppression use, prednisone dosage (cumulative, average and current), use of opioids/hypnotics or sedatives, disease activity of lupus (SLEDAI/ SELENA SLEDAI/ or as per rheumatologist assessment, number of non routine / urgent visits to Rheumatologist or visit to ER/urgent care for lupus
B. All patients meeting the inclusion and exclusion criteria will be recruited and selected after signing informed consent. Optimum number of patients will be selected as per statistician for maximum power. Baseline visit will include complete assessment by the rheumatologist with physical exam and documentation of SELENA-SLEDAI, SELENA-2K, activity in major organ systems (renal, CNS, cardiopulmonary, vasculitis, hemolytic anemia, GI activity), features of lupus disease activity, current dose of prednisone, immunomodulator medications, HAQ scores,
SRI-4, BILAG, FACIT scores, mean physician global assessment score, mean tender joint counts, mean swollen joint count. Questions regarding OSA (current duration of CPAP use (number of hours each night and number of night in one week), visits to sleep physician, AHI) will be assessed by the nurse and will be masked by the rheumatologist. The study visits will be scheduled in 12 weeks and 24 weeks. Primary and secondary endpoints will be analyzed after completion of the study at 24 weeks. Any additional visits for symptom assessment (urgent care or hospital or clinic) will be recorded. Primary outcome will be SRI-4 response at week 24. Secondary outcomes analyses will be compromised of above variables. CPAP use will be dichotomised to yes: exposed (more than six hours every night) or non-exposed (less than six hours). Disease activity as per the above variable will be statistically analyzed against CPAP use or no CPAP use.
References


Leukocytoclastic Vasculitis Confounding Panton-Valentine Leukocidin Methicillin-Resistant *Staphylococcus aureus* pneumonia

Brittany Hill, D.O.¹; Faheem Shaikh, M.D.²

Baptist Health System Department of Internal Medicine; Birmingham, Alabama

Leukocytoclastic vasculitis, a small vessel vasculitis, has many etiologies including recent infection, drug induced and idiopathic. Rash occurs due to immune complex deposition and complement activity which leads to inflammatory cell migration and resultant vessel damage. This report presents a 74-year-old female without underlying pulmonary pathology who was admitted for acute hypoxic respiratory failure secondary to bilateral pneumonia with significant effusions. The patient had consistent bacteremia despite appropriate antibiotic therapy. On day 13 of admission, she developed extensive non-blanching petechiae to the trunk, abdomen, bilateral lower extremities and eventually the chest and arms. Cutaneous biopsy revealed leukocytoclastic vasculitis. Due to the florid bacteremia, there was difficulty discerning the etiology of the rash. However, it was determined that the recently prescribed lisinopril was the cause and was promptly discontinued. The rash improved over the remainder of the hospital stay. Concern for Panton-Valentine Leukocidin Methicillin-Resistant *Staphylococcus aureus* arose due to persistent bilateral necrotizing pneumonia. Linezolid and Clindamycin were added to the regimen with evidence that these antibiotics decrease the Panton-Valentine Leukocidin toxin. Discerning the cause of leukocytoclastic vasculitis in the setting of sepsis can be difficult due to medication changes and the possibility of the underlying infection as the etiology. In this case, drug eruption was the most likely cause and the rash improved with discontinuation of the medication.

The authors have no conflicts of interest to disclose.
Can’t See, Can’t Hear, Can’t Understand

Author: Patricia Kachur MD, William Davis MD

Ochsner Medical Center, Rheumatology Department

There are no conflicts of interest to disclose.

Susac syndrome (SuS) is an immune-mediated, pauci-inflammatory, ischemic and occlusive microvascular endotheliopathy that mainly affects the brain, retina, and inner ear. A clinical triad of encephalopathy, branch retinal artery occlusion, and low-frequency sensorineural hearing loss characterizes the classical form. MRI shows distinctive “snowball” lesions in the corpus callosum. This is an extremely rare condition with only about 100 reported cases in literature. Today we present the case of a 50 year old female who presented with acute encephalopathy, severe migraines, neuropathy of the LUE, neck, and face in the setting of a history of hypothyroidism. Physical exam showed no focal deficits and stable vitals. Workup with MRI showed numerous foci of abnormal T2 signal throughout the periventricular white matter and cerebellum, cerebro-spinal fluid (CSF) was sterile with an elevated protein. Initial therapy with high dose steroids (4g) resulted transient improvement, with symptom recurrence after a month during which time patient was on a steroid taper.

Despite symptoms repeat MRI brain was now normal, CSF findings were again sterile with elevated protein. A repeat dose of steroids yielded a 2-week remission in symptoms and MRI showed recurrence of abnormal T2 signals with vague, patchy enhancement. Ophthalmology was consulted and found multiple retinal artery branch blockages. Patient was diagnosed with SuS received IVIG, followed by outpatient prednisone 100mg daily, cyclophosphamide (Cytoxan) 15mg/kg IV and IVIG every 2 weeks. Despite these measures, symptoms recurred within 2 months, requiring additional 3g of steroids, and a trial of rituximab 1000mg x 2 doses (14 days apart). Currently the patient has been in remission for 2 months.

Given the rarity of this syndrome, data on treatment is sparse. Given its immunopathogenesis similarity to juvenile dermatomyositis (JDM), some follow treatment strategies for the latter disease. Current expert recommendations are based on case-series and database studies, and suggest using high dose steroid (1mg/kg/day) x 4 weeks follow by taper with IVIG (2gm/kg over 2 days) every 3-4 weeks as first line. Additional therapies are added in the event of severity of CNS involvement. Cyclophosphamide with Mycophenolate mofetil and Rituximab are considered additions to the treatment regiment if needed. Experts had said that the most challenging part of SuS treatment is the long-term management as the disease process is difficult to predict and discern. Relapse is common especially when tapering of medication is too quick. Early detection and aggressive treatment for disease suppression is the main key to prevent permanent damage to the brain, retina, and cochlear.
**Treat to Target: Do Rheumatologists Adjust Therapy Based on High Disease Activity Score (DAS), and Does This Result in Improved DAS?**

Patricia Kachur MD, Gbemisola Olayemi MD, William Davis MD, Robert Quinet MD

Ochsner Medical Center

No financial disclosure

Rheumatoid arthritis (RA) is a systemic inflammatory condition that can significantly impact the quality of life if uncontrolled. The American College of Rheumatology (ACR) guidelines for RA provide recommendations aimed at achieving low activity and/or remission, but in practice non-adherence to guidelines is common. The goal of this study is to determine if adherence to ACR guidelines in RA patients with high DAS28 scores results in more frequent modifications in RA therapy and if these results improved RA disease activity as a result of these decisions. lower DAS28 scores than in patients without guideline-directed treatment (GDT).

We conducted a retrospective chart review of 94 RA patients encompassing 153 index encounters with high DAS28 scores (>5.1) from July 2013 to December 2018. The index visit was used to collect their baseline DAS28 score. The treatment strategy recorded in their chart: GDT or non-GDT. A follow-up DAS28 score was recorded on the follow-up visit and used for analysis. Encounters were excluded if no follow up DAS28 score was present or if the patient had a diagnosis of concurrent fibromyalgia.

89/153 index encounters had sufficient data to include in the study. Mean overall DAS28 for the pre-intervention group was 6.22±0.87. Baseline differences in DAS28 scores between the two groups were not significantly different (GDT 6.33, non-GDT 6.11; p=0.23). In the post-intervention group, the overall mean DAS28 was 5.03±0.87. Mean differences in post-therapy GDT were significantly lower in the GDT. GDT was documented in 46 patients resulting in a mean post-intervention DAS28 of 4.68±1.64. The Non-GDT in the remaining 43 patients resulted in post-intervention DAS28of 5.40±1.38. The mean differences in post-therapy GDT were significantly lower in the GDT group (p = 0.026).

Our study suggests that in our cohort of RA patients with comparable baseline DAS28 scores, adherence to GDT is associated with a significantly lower follow-up DAS28 scores when compared to the non-GDT group. Given well-established evidence that uncontrolled RA reduces the ability to perform daily activities, health-related quality of life, and permanent damage, QI initiatives can focus on increasing GDT adherence to improve DAS28 scores and likely other outpatient outcomes.
Association of Smoking Status and Total and Individual Damage Index in Systemic Lupus Erythematosus

Authors:
Romy Kallas MD, Lupus Fellow
Jessica Li, MPH
Michelle Petri MD MPH, Professor of Medicine

Funding Source:
The Hopkins Lupus Cohort was funded by NIH Grant R01-AR069572

Conflict of interest:
No potential conflict of interest with respect to the research, authorship, and/or publication of this article

Background
Smoking is a risk factor for systemic lupus erythematosus (SLE). It has been associated with increased disease activity and decreased effectiveness of hydroxychloroquine in cutaneous lupus. We looked at the association of both smoking and ethnicity with the individual damage items from the SLICC/ACR Damage Index.

Methods
Poisson regression was used to model the total SLICC/ACR Damage Index score against ever smoking. Cox regression was used to assess the relationship between time to individual damage items and ever smoking. We also looked at this relationship separately for African-American and Caucasian patients. Furthermore, we compared SLICC/ACR Damage Index items among African-American and Caucasian ever smokers.

Results
The study included 2629 patients, 52.6% Caucasian and 39.3% African-American. The prevalence of ever smokers was 35.8%. There was no significant difference in total SLICC/ACR Damage Index score between ever smokers and never smokers after adjustment for ethnicity, gender, age at diagnosis, and years of education. Ever smokers had more atherosclerotic cardiovascular damage (angina, coronary bypass, myocardial infarction and claudication) and skin damage compared to non-smokers. Caucasian SLE patients who ever smoked were more likely to have muscle atrophy and atherosclerosis compared to Caucasian non-smokers. African-American patients who ever smoked were more likely to have skin damage compared to African-American non-smoker. African-Americans who smoked were more likely to have multiple types of organ damage compared to Caucasians who smoked, with only 2 exceptions (gastrointestinal infarction and osteoporotic fracture were more common in Caucasian smokers.)

Conclusion
Smoking is a modifiable factor for organ damage in SLE. Our analysis proved its major effect on cardiovascular and cutaneous damage. Surprisingly, cardiovascular damage items had higher hazard ratios in Caucasian smokers than non-smokers while skin damage items hazard ratios were higher in African-American smokers.
<table>
<thead>
<tr>
<th></th>
<th>All</th>
<th>African American</th>
<th>Caucasian</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td># of events</td>
<td>HR (95% CI)</td>
<td>p-value</td>
</tr>
<tr>
<td>Cranial OR peripheral neuropathy</td>
<td></td>
<td>186</td>
<td>0.85 (0.63,1.16)</td>
</tr>
<tr>
<td>Pulmonary fibrosis</td>
<td></td>
<td>178</td>
<td>0.77 (0.56,1.06)</td>
</tr>
<tr>
<td>Angina OR coronary artery bypass</td>
<td></td>
<td>88</td>
<td>1.55 (1.23,1.99)</td>
</tr>
<tr>
<td>Myocardial infarction ever</td>
<td></td>
<td>99</td>
<td>1.77 (1.17,2.67)</td>
</tr>
<tr>
<td>Claudication x6 months</td>
<td></td>
<td>35</td>
<td>2.87 (1.38,5.97)</td>
</tr>
<tr>
<td>Venous thrombosis with swelling, ulceration, OR venous stasis</td>
<td></td>
<td>66</td>
<td>0.54 (0.31,0.94)</td>
</tr>
<tr>
<td>Pancreatitis</td>
<td></td>
<td>9</td>
<td>5.47 (1.09,27.42)</td>
</tr>
<tr>
<td>Muscle atrophy or weakness</td>
<td></td>
<td>47</td>
<td>1.42 (0.78,2.6)</td>
</tr>
<tr>
<td>Osteoporosis with fracture or vertebral collapse</td>
<td></td>
<td>312</td>
<td>0.9 (0.72,1.14)</td>
</tr>
<tr>
<td>Extensive scarring or panniculum other than scalp and pulp space</td>
<td></td>
<td>37</td>
<td>2.53 (1.26,5.07)</td>
</tr>
<tr>
<td>Skin ulceration (not due to thrombosis) for more than 6 months</td>
<td></td>
<td>25</td>
<td>2.7 (1.15,6.35)</td>
</tr>
<tr>
<td>Premature gonadal failure</td>
<td></td>
<td>67</td>
<td>1.45 (0.87,2.41)</td>
</tr>
</tbody>
</table>

1 Adjusted for sex, race, age at diagnosis, years of education, 2 Adjusted for sex, age at diagnosis, years of education
Table 2: Associations between SLICC/ACR Damage Index items and ethnicity: comparing African Americans and Caucasians ever smokers

<table>
<thead>
<tr>
<th>Condition</th>
<th># of events among smokers</th>
<th>HR(^1) (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any cataract ever</td>
<td>176</td>
<td>1.42 (1.05, 1.93)</td>
<td>0.024</td>
</tr>
<tr>
<td>Estimated or measured GFR &lt; 50%</td>
<td>173</td>
<td>1.82 (1.33, 2.48)</td>
<td>0.000</td>
</tr>
<tr>
<td>Proteinuria 3.5g/24hrs</td>
<td>56</td>
<td>2.97 (1.66, 5.32)</td>
<td>0.000</td>
</tr>
<tr>
<td>End-stage renal disease</td>
<td>37</td>
<td>3.87 (1.88, 8.3)</td>
<td>0.000</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>86</td>
<td>2.25 (1.44, 3.53)</td>
<td>0.000</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>39</td>
<td>2.65 (1.34, 5.21)</td>
<td>0.004</td>
</tr>
<tr>
<td>Infarction or resection of bowel</td>
<td>83</td>
<td>0.56 (0.35, 0.89)</td>
<td>0.014</td>
</tr>
<tr>
<td>Deforming or erosive arthritis</td>
<td>85</td>
<td>1.69 (1.09, 2.63)</td>
<td>0.019</td>
</tr>
<tr>
<td>Osteoporosis with fracture or vertebral collapse</td>
<td>120</td>
<td>0.42 (0.28, 0.63)</td>
<td>0.000</td>
</tr>
<tr>
<td>Avascular necrosis</td>
<td>76</td>
<td>2.50 (1.54, 4.04)</td>
<td>0.000</td>
</tr>
<tr>
<td>Scarring chronic alopecia</td>
<td>33</td>
<td>4.66 (2.01, 10.83)</td>
<td>0.000</td>
</tr>
<tr>
<td>Extensive scarring or panniculum other than scalp and pulp space</td>
<td>23</td>
<td>3.90 (1.44, 10.57)</td>
<td>0.007</td>
</tr>
<tr>
<td>Diabetes</td>
<td>59</td>
<td>3.12 (1.75, 5.54)</td>
<td>0.000</td>
</tr>
</tbody>
</table>

\(^1\) Adjusted for sex, age at diagnosis, and years of education
Vasculitis Neuropathy Masquerading as Guillain-Barre Syndrome

Jan Karczewski MD, Matthew White DO, Stuart Schwartz MD

Mononeuritis multiplex and polyneuropathy may be the presenting manifestations of medium to small vessel vasculitis, making it difficult to distinguish from a primary neurologic process. Non-systemic vasculitic neuropathy is a rare form of vasculitis isolated to the peripheral nervous system which requires neuromuscular biopsy for diagnosis. It is usually clinically distinct from Guillain-Barre syndrome (GBS), which presents most commonly as an ascending symmetric paralysis with depressed reflexes. Non-systemic vasculitic neuropathy may be mistaken for a primary neurologic disorder. We present a case of mononeuritis multiplex rapidly evolving into ascending paralysis masquerading as GBS.

A 57-year-old female with a history of pan-uveitis presented to the emergency department with a two-week history of progressive weakness and numbness in both legs and right wrist, as well as a faint rash on all four extremities. These symptoms were associated with painful paresthesias and frequent falls.

Her vital signs were unremarkable. Physical exam revealed generalized weakness most notable in her lower extremities, with 1/5 strength of ankle dorsiflexion and plantarflexion bilaterally. Reflexes were absent in the patellar and Achilles tendons and remaining reflexes were diminished bilaterally. A faint erythematous macular rash was present throughout all four extremities. Laboratory results were significant only for hyponatremia of 127 mEq. Nerve conduction studies revealed sensorimotor polyneuropathy. Nodular thickening and enhancement of the nerve roots within the cauda equina were present on MRI. These findings were concerning for GBS and treatment with intravenous immunoglobulin was initiated. The patient’s weakness rapidly progressed in a symmetric pattern resulting in flaccid paralysis of all four extremities and rapidly involved the respiratory muscles requiring intubation. Cranial nerve III, V, and VII palsies were also present on exam. Plasmapheresis was initiated due to concern for GBS. Unfortunately, her condition did not improve. Ultimately, a sural nerve biopsy was performed which was consistent with chronic vasculitis involving small perineurial and endoneurial vessels with transmural lymphocytic inflammation. Anti-nuclear antibodies, cryoglobulins, and anti-neutrophil cytoplasmic antibodies (ANCA) were negative. She was started on 60mg of intravenous methylprednisolone daily with a gradual improvement in facial palsies and distal extremity strength.

This case describes a rare presentation of a small vessel vasculitis without the presence of ANCA or cryoglobulins, predominately affecting the peripheral nervous system. The patient’s rapid symmetric ascending paralysis with respiratory failure resembled AIDP and GBS. Treatment failure and a re-evaluation of the diagnosis prompted the sural nerve biopsy which revealed vasculitis. This case illustrates the myriad neurologic manifestations of vasculitis and emphasizes the importance of rethinking the original diagnosis in the face of treatment failures.

The authors have no disclosures.
Acute Monoarthritis and Colitis Secondary to Anti-Cancer Therapy
Jan Karczewski MD, Matthew White DO

The Authors have no disclosures.

Immune checkpoint inhibitors have been revolutionary in the treatment of various cancers over the last decade. Although initially studied in metastatic melanoma, they have now been approved for various types of cancer. They work by inhibiting the natural regulation of T-cells, leading to increased T-cell activation and anti-tumor activity. However, enhanced activation of T-cells can create autoimmunity and contribute to inflammatory side effects.

A 69-year-old male with a history of stage IV urothelial carcinoma, now in remission on atezolizumab, an anti-PDL-1 monoclonal antibody, presented to the emergency department with a one-week history of right knee pain and swelling. He was seen in the emergency department 4 days prior for the same complaint and an arthrocentesis was performed. Since then he developed fevers, chills, as well as new bright red blood per rectum. He reports inability to bear weight on his knee secondary to pain. He denied prior episodes of joint swelling, fever, chills or infection.

His vital signs were notable for a fever of 39.0 C, with normal hemodynamic parameters. Physical exam showed a diffusely tender right knee with large effusion, as well as a small effusion of the left knee. Laboratory investigations revealed a white blood cell count of 10.1 µL with an elevated CRP of 268 mg/L and ESR of 98 mm/hr. He was mildly anemic with hemoglobin of 13.0 g/dL. MRI of the right knee revealed a large joint effusion with enhancing synovitis. Repeat knee aspiration yielded 94 mL of cloudy fluid with 27,000 white blood cells. Synovial fluid cultures were negative, as was testing for Lyme disease and crystal analysis.

His hospital course was complicated by continued gastrointestinal bleeding. Colonoscopy revealed moderate to severe mucosal changes consistent with colitis, which was confirmed on biopsies. Our patient exhibited a systemic inflammatory process affecting symmetric large joints and colon, likely related to anti-PDL-1 therapy. Treatment with 80 mg of prednisone resulted in the resolution of his synovitis and hematochezia.

In summary, this case illustrates a new spectrum of immune-related adverse events secondary to immune checkpoint inhibitors which may be encountered by the rheumatologist in the form of inflammatory arthritis and may have systemic manifestations as in this case. Diagnosis is based on clinical findings, as there is no diagnostic test, and involves ruling out infectious causes.
**Refractory Adult Onset Still's Disease with Initial Good Response to Canakinumab: A Case Report**

Ali H. ALI MD, Mandip Kang MD, Adam Shurbaji MD, Mosaab Mohammeden MD, Candice Yuvienco MD, RhMSUS.

**Purpose:** Treatment-Resistant Adult-Onset Still's Disease (AOSD) has been a treatment challenge. We are reporting an AOSD case refractory to multiple biologics and non-biologics Disease-Modifying Antirheumatics Drugs (DMARDs) but responded very well to Canakinumab.

**Case Description:** 29 years old Caucasian female with AOSD who was diagnosed According to Yamaguchi criteria, as the patient had three major criteria and five minor criteria in the setting of prolonged history of Fever (>39 C) with salmon color rash, arthralgia, sore throat, cervical lymphadenopathy with transaminitis and negative immunological markers (anti-nuclear antibody, double-stranded deoxyribonucleic antibody, complements level, rheumatoid factor, anti-citrullinated peptide). Ferritin was elevated (3724 ng/ml), Computed Tomography of chest, abdomen and pelvis was significant for hepatosplenomegaly and bone marrow biopsy was not remarkable.

Infections and malignancies were ruled out before making the diagnosis. Patient first received adalimumab 40 mg every other week for one year which helped with the joint pain but continued to have fever and rash. Then, Methotrexate 25 mg weekly and prednisone 5 mg daily were added. However, she continued to have skin rash whenever she tried to taper off the prednisone. Consequently, Adalimumab was stopped and Anakinra 100 mg daily was started for 8 months but then discontinued due to wearing off effect with recurrence of fever and joint pain at night. Canakinumab 150 mg every 28 days was started then switched to every 21 days as she continued to have joint pain and fever around the beginning of the fourth week. Patient showed good response for 10 months with canakinumab and methotrexate 12.5 mg weekly but without steroids. However, due to pregnancy planning, methotrexate was stopped, and she had another flare one month later.

**Discussion and conclusion:** AOSD is an uncommon autoimmune disease characterized by high spiking fever with salmon-like rash, arthritis and elevated ferritin. The disease is not homogenous, ranging from mild forms to severe life-threatening complications, such as macrophage activation syndrome. AOSD is a diagnosis of exclusion, that usually made after ruling out infections, malignancies and other autoimmune diseases. Glucocorticoids are usually the initial therapy followed by biologic or non-biologic DMARDs. The selection of DMARDs depends on the clinical presentation of the disease. Methotrexate or TNFα inhibitor are usually recommended for arthritis predominant, while Anakinra, Canakinumab or tocilizumab are preferred for systemic inflammation predominant. Refractory AOSD patients who were treated with various biologic and non-biologic DMARDs without satisfactory response has been an issue due to insufficient data about an alternative regimens. Based on the fact that interleukin-1 family and IL-1β in particular has a vital role in AOSD pathogenesis, a few case reports and series showed a successful treatment of refractory AOSD with Canakinumab, the highly specific IL-1β blocking agent. We suggest that Canakinumab can be considered as an option for refractory AOSD. However, further studies are required to support this approach.

Disclosure Statement: The authors have no conflict of interests to disclose.
Elevated Cardiac Troponin T in Patients with Lupus Myositis

Guy Katz, M.D.; Sharon L Kolasinski, M.D.; Baskaran Sundaram, M.D.; Giorgos Loizidis, M.D.

**Background:** Chest pain is a common complaint in patients with systemic lupus erythematosus (SLE), with causes ranging from cardiac disease, pulmonary disease, esophageal disorders, and musculoskeletal chest wall pain. The biomarkers of choice to detect myocardial injury are cardiac troponin T and I (cTnT, cTnI). In the idiopathic inflammatory myopathies, cTnT is often elevated in the absence of cardiac injury, likely because the protein is released from skeletal muscle in addition to the myocardium. We present two cases of patients with SLE and associated myositis who presented with chest pain and elevated cTnT but whose cardiac magnetic resonance imaging (MRI) showed no myocardial inflammation and cTnI was normal.

**Case 1:** A 21-year-old female with a history of SLE with prior myositis and MRI-confirmed myocarditis who presented with pleuritic chest pain as well as alopecia, Raynaud’s phenomenon, polyarthritis, and proximal muscle weakness. Markers of SLE disease activity and creatine kinase (CK) were elevated, and C3 and C4 complements were low. Cardiac workup revealed an elevated high-sensitivity cTnT, but cTnI, electrocardiogram, and transthoracic echocardiogram (TTE) were normal. Given her prior history of myocarditis, cardiac MRI was performed, and it showed no evidence of recurrent myocarditis. Her lupus flare was treated with prednisone, and her symptoms, including chest pain, resolved.

**Case 2:** A 24-year-old female with a history of SLE with associated myositis presented to the emergency department with episodic but non-exertional chest pain. Other symptoms at that time included a malar rash and Raynaud’s phenomenon. Cardiac workup revealed an elevated cTnT that increased when repeated but normal electrocardiogram, cardiac MRI, and cTnI. Aldolase and CK were elevated, but markers of SLE disease activity were normal. Transthoracic echocardiogram showed mild right ventricular dilation but was otherwise normal, and computed tomography angiography showed no evidence of pulmonary embolism. She was treated with prednisone with improvement in her symptoms.

**Clinical implications:** These are the first two cases reported of patients with SLE and associated myositis with elevated CK who presented with chest pain and were found to have elevated cTnT but normal cTnI. Both patients had a negative electrocardiogram, TTE, and cardiac MRI. Cardiac involvement is common in SLE, and it can be clinically silent. The measurement of the highly sensitive cTnT to uncover silent heart disease in patients with SLE is an area of active investigation. However, clinicians should be aware that the specificity of an elevated cTnT might be lower in active SLE myositis, and cTnI may assist in detecting myocardial injury in these cases.

**Disclosures:** The authors have no financial conflicts of interest to disclose.
Refractory Valvular Heart Disease Due to Immunoglobulin G4-Related Disease of the Aortic and Mitral Valves

Authors: Guy Katz, MD; Brett Victor, MD; Siddharth Bhattacharyya, MD; Adam Binder, MD

Background: Immunoglobulin G4-related disease (IgG4-RD) is a systemic fibroinflammatory disease known to involve nearly every organ system. While aortitis and periaortitis are common components of the disease, cardiac involvement has only rarely been reported. As a result, the clinical presentation of cardiac IgG4-RD remains poorly understood.

Case summary: A 78-year-old male presented to the hospital with dyspnea on exertion, cough, and pleuritic chest pain. Six years prior to this presentation, the patient was found to have aortic stenosis and mitral regurgitation for which he underwent bioprosthetic valve replacements. Two years later, he developed a paravalvular leak and severe stenosis in his bioprosthetic mitral valve, which was repaired percutaneously followed by a valve-in-valve transcatheter mitral valve replacement. At that time, he had imaging showing bilateral mediastinal and hilar lymphadenopathy as well as bibasilar lung opacities; the adenopathy resolved on repeat imaging a few weeks later, and no diagnosis was made at that time. Over the next two years, the patient had a chronic cough and dyspnea on exertion that were treated empirically by various providers with antibiotics, bronchodilators, and short courses of oral corticosteroids with minimal benefit. The patient was then found to have recurrent mitral stenosis and new aortic prosthetic valve regurgitation and stenosis. Shortly thereafter, he presented to the emergency department with recurrent dyspnea on exertion, cough, and pleuritic chest pain. During this presentation, the patient also noted weight loss and mild night sweats over the previous few months, and physical examination revealed bilateral non-tender submandibular gland enlargement. Computed tomography (CT) of the chest demonstrated recurrent mediastinal and hilar adenopathy as well as basilar lung opacities. Extensive workup during that hospitalization revealed elevated total IgG and IgG4 levels. He subsequently had a lung biopsy that showed chronic pneumonitis, interstitial and confluent fibrosis, abundant plasma cells, and greater than 20 IgG4-positive cells in numerous high-powered fields with a high ratio of IgG4+/IgG+ cells. The patient was diagnosed with IgG4-RD and started on prednisone with rapid resolution of his chest pain and cough. After his diagnosis was established, pathology samples from his initial aortic and mitral valve replacement surgery six years prior were obtained and re-examined. Both valves showed chronic inflammation, lymphoplasmacytic infiltration, and an IgG4+/IgG+ cell ratio greater than 40%.

Clinical implication: IgG4-rich plasma cell infiltration has been observed in a small number of cases of aortic and mitral pathology, but this in association with other systemic findings of IgG4-RD has only rarely been reported. This case demonstrates that valvulitis can be a first manifestation of the disease and can precede other findings by years. In addition, it may indicate that IgG4-RD of the heart valves can result in premature, recurrent prosthetic valve failure. Further research is warranted to better characterize the role of IgG4 in valvular heart disease.

Disclosures: The authors have no conflicts of interest to disclose.
Arthritis in Systemic Sclerosis: A Practical Classification Scheme

M. Kazem, J.E. Pope

Inflammatory arthritis associated with Systemic Sclerosis (SSc) is a common finding in patients with the disease. It can present in different patterns and mimic diseases such as rheumatoid arthritis or systemic lupus erythematosus or a present with severely destructive phenotype.

Symptoms in SSc patients can also come from other musculoskeletal manifestation of their disease (tendon friction rubs, hand contractures, calcinosis, tuft resorption) or relate to non-inflammatory causes processes such as crystal arthritis, osteoarthritis, or tendonitis.

Serological risk factors include seropositivity for RF and CCP for significant inflammatory arthritis. Conventional synthetic disease modifying anti-rheumatics have been used successfully before in treatment of inflammatory arthritis associated with SSc. Small studies have also looked at tocilizumab and abatacept for treatment of inflammatory arthritis. TNF-alpha inhibitors are successfully used in erosive polyarthritis associated with SSc.

Given the variable presentation, a comprehensive history along with appropriate investigations can better inform clinicians of their patient’s trajectory as it pertains to inflammatory arthritis. Based on available literature, we propose five phenotypic subtypes of inflammatory arthritis in patients with SSc (Table 1).

Table 1. Arthritis subtype patterns and associated features

<table>
<thead>
<tr>
<th>Arthritis Subtype</th>
<th>Signs &amp; Symptoms on history</th>
<th>Radiologic findings</th>
<th>Treatment modalities</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Erosive polyarticular</strong></td>
<td>Painful swollen joints, could be RA like Or patients with overlap syndromes</td>
<td>Erosive changes (RA like) such as wrists, MCPs</td>
<td>Treat like RA Methotrexate Other conventional synthetic DMARDs Leflunomide TNF inhibitors Etanercept Infliximab Adalimumab Certolizumab pegol Golimumab</td>
</tr>
<tr>
<td>Condition</td>
<td>Description</td>
<td>X-rays/Other Features</td>
<td>Treatment Options</td>
</tr>
<tr>
<td>---------------------------------</td>
<td>-----------------------------------------------------------------------------</td>
<td>------------------------------------------</td>
<td>----------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Erosive arthritis with ankylosis</td>
<td>May look more like seronegative spondyloarthropathy or rheumatoid arthritis but no range of motion where ankylosing occurs</td>
<td>X-rays show joint ankylosis and erosions</td>
<td>Likely treat like polyarticular erosive arthritis</td>
</tr>
<tr>
<td>Non-erosive</td>
<td>Arthralgia, inflammatory arthritis</td>
<td>No erosions on X-rays</td>
<td>NSAIDs</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Low dose glucocorticoids</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Hydroxychloroquine</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Possibly if overlapping with SLE and uncontrolled arthritis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Belimumab</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Rituximab</td>
</tr>
<tr>
<td>Mutilans</td>
<td>Inflammatory arthritis and rapid development of deformities, possibly shortening of the digits</td>
<td>Large erosions and periosteal resorption</td>
<td>Conventional synthetic DMARDs</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Methotrexate</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Leflunomide</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Rapid addition of TNF inhibitor</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>If failing above treatments, follow the RA treatment algorithm</td>
</tr>
<tr>
<td>Non arthritic bone abnormalities</td>
<td>Tuft resorption</td>
<td>Classic tuft resorption of digits on X-ray</td>
<td>Unknown if there is no inflammatory arthritis</td>
</tr>
<tr>
<td>---------------------------------</td>
<td>----------------</td>
<td>-------------------------------------------</td>
<td>---------------------------------------------</td>
</tr>
<tr>
<td><strong>Acro-Osteolysis</strong></td>
<td></td>
<td>May or may not have inflammatory arthritis</td>
<td></td>
</tr>
</tbody>
</table>

Disclosure: I, May Kazem, have no conflicts of interest to disclose.
Cutaneous Necrosis: An unexpected trigger

Sai Kollipara MS-IV, Chandana Kollipara DO

Background: Cutaneous vasculitis of small and medium vessels can lead to purpura, petechiae, or ulcers. Typically diseases that are commonly associated with cutaneous vasculitis include ANCA vasculitides, cryoglobulinemic vasculitis, urticarial vasculitis, polyarteritis nodosa, rheumatoid vasculitis, medication induced. Levamisole-induced vasculitis is rare but an increasingly important phenomenon with similar cutaneous presentation. Levamisole is an anti-helminthic drug which is commonly found in adulterated cocaine. Here we present a novel case of Levamisole-induced vasculitis.

Case: A 53 year old white female with history of bilateral above knee amputation due to necrotic lesions and polysubstance abuse presented with diffuse pain and necrotic ulcers. The patient had fever, chills, tachycardia, decreased oral intake. A recent pseudomonas infection prompted concern for sepsis for which patient was admitted. On physical exam, necrotic pinna was present bilaterally. Integumentary findings showed necrotic ulcerations over torso, bilateral upper and lower extremities as well as dressed stumps with serosanguinous discharge that were tender to touch. Labs showed high erythrocyte sedimentation rate greater than 120, high CRP 18.2mg/dl, positive C-ANCA and P-ANCA titer, myeloperoxidase and Cryoglobulin screens. Patient had negative antinuclear-antibody(ANA), C3, C4, rheumatoid factor, cyclic citrullinated peptide, proteinase 3 and serum protein electrophoresis. Patient’s urine drug screen was positive for cocaine. Skin and soft tissue biopsy shows necrosis, hemorrhage, acute inflammation and fibrin thrombi. Only after prolonged abstinence from cocaine and 15 mg dose prednisone did the lesions improve. For these reasons and the classic retiform purpura, we clinched the diagnosis Levamisole-induced vasculitis. With prednisone and wound care, the patients necrotic ulcerations began to heal with pink granulation tissue forming over the course of 6 weeks.

Discussion: Hallmark features of Levamisole-induced vasculitis (LIV) include tender reticular purpuric rash with or without necrosis, neutropenia, and positive p-ANCA antibody. LIV is unique in that the p-ANCA targets human neutrophil elastase, lactoferrin, cathepsin G rather than myeloperoxidase. It also differs from cocaine induced vasculitis which is typically a c-ANCA driven process. LIV often can present with multiple positive antibodies such as ANA, anti-cardiolipin antibody and MPO ANCA. It is important that physicians are aware of this to minimize unnecessary workup. Primary treatment is cocaine cessation. Patients benefit from supportive measures such as wound care and also low dose steroids. With the increasing usage of cocaine amongst young Americans it is vital for physicians to be aware of this entity. When encountering a cutaneous vasculitis it’s crucial to check urine for cocaine as LIV is a key differential. Early identification and treatment can prevent extensive necrosis and disfigurement.

Disclosure Statement: I have no relationships with any entity producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients.
A Fatal Case of Adult Onset Still’s Disease (AOSD) Complicated With Hemophagocytic Lymphohistiocytosis (HLH)

Sindhuja Korem, Ajay Venkatanarayan, Shilpan Shah - Monmouth Medical Center, NJ.

We present a rare immune-mediated, inflammatory condition with an unfortunate late diagnosis and mortality. AOSD complicated with HLH is a difficult diagnosis due to the non-specific presentation of both the conditions. A missed diagnosis of this condition can be fatal.

A 56 yo lady with a history of SLE, RA, fibromyalgia presented with fevers, myalgias, joint pains, and upper respiratory symptoms. On exam, she was febrile with mild pharyngeal erythema and mucosal ulcerations. Laboratory findings included normal WBC with bandemia, mildly elevated inflammatory markers and LFTs, and negative tests of rapid mono and strep. Her chest xray was unremarkable. The initial diagnosis was a viral infection and the patient was started on Tamiflu and Ampicillin/sulbactam. During her hospital course, her upper respiratory symptoms improved however she remained intermittently febrile. The ongoing workup included a negative respiratory panel, negative blood cultures, unremarkable abdominal US and CT Neck after developing odynophagia. While planning her discharge with oral antibiotics, she suddenly declined with persistent fevers, increasing liver enzymes and new pancytopenia. Her exam was stable with mild abdominal pain. With an unclear diagnosis, a series of auto-immune lab panels were performed all of which returned negative. A CT Torso was performed indicating small bilateral pleural effusions. Viral lab panels returned negative. The hepatic injury and pancytopenia was attributed to her prior use of azathioprine. The patient’s clinical course continued to deteriorate including hypotension, acute kidney injury, and metabolic acidosis. Immediate treatment included critical care support with aggressive hydration, bicarbonate infusion, multiple transfusions for pancytopenia and suspected DIC, and intubation after seizures. Upon close review of her lab values and clinical presentation we began to suspect AOSD. In addition, due to elevated LDH, significantly elevated ferritin and low fibrinogen, her condition was likely compounded with HLH. Intravenous steroids were immediately administered. Concurrently, efforts were made to transfer the patient to a tertiary center. Unfortunately, after arriving at the institution, the patient had a cardiac arrest and expired.

This is a severe presentation of AOSD complicated with HLH, where a delayed diagnosis was fatal. This diagnosis is challenging and often mistaken for septic shock, which can be detrimental. We review the literature and discuss diagnostic criteria and treatments for this critical syndrome.

References:

1. Hemophagocytic lymphohistiocytosis (HLH) and related disorders Alexandra H. Filipovich1 1 Immunodeficiency and Histiocytosis Program, Division of Hematology/Oncology, Cincinnati Children’s Hospital Medical Center, Cincinnati, OH

2. Hemophagocytic Lymphohistiocytosis in a Patient with Adult-Onset Still’s Disease- a Diagnostic Dilemma- Priyank Gupta, Roopesh Pandey, Chetan Unadkat

Disclosures: Author has no disclosures to make
A Rare Presentation of Concurrent Small and Large Vessel Vasculitis

Sindhuja Korem, Mridul Gupta, Nivera Noel, Monmouth Medical Center, NJ.

Vasculitis refers to a group of diseases characterized by inflammation of vessel walls along with reactive damage to mural structures. Vasculitis are classified based on predominant size of vessels involved. Temporal arteritis predominantly involves large vessels and microscopic polyangitis predominantly involves small vessels. We hereby present a rare case of successive large and small vessel vasculitis in an elderly female.

An 82-year-old Caucasian female with a history of a transient ischemic attack presented with sudden onset right sided vision loss. She was treated with pulse steroids and underwent right sided temporal artery biopsy, which was negative for temporal arteritis. She was lost to follow up but presented four months later with 40 lbs weight loss, fatigue, poor appetite and exertional dyspnea. She had a recent urinary infection that was treated with 5 days of trimethoprim-sulfamethoxazole. On presentation, she appeared frail, dehydrated and was hypotensive. She had persistent vision loss in right eye. Investigations were significant for thrombocytosis 808,000 cells/µL, lactic acidosis 3.6 mmol/L, creatinine 6.91 mg/dl, sed rate 130 mm/hr and CRP 98 mg/L. Previous records showed normal renal function 4 months ago. Her urinalysis showed mild proteinuria, positive leukocyte esterase, but was negative for casts. Chest X ray did not show any infiltrates. She was treated for possible sepsis with broad spectrum antibiotics and fluid resuscitation.

Her elevated inflammatory markers, new diagnosis of renal failure with recent episode of monocular vision loss prompted further immunological workup. ANA was weakly positive (1:80), normal C3, C4, negative anti GBM antibodies, negative Hepatitis panel, negative PR3-ANCA and positive MPO-ANCA. She underwent left sided temporal artery biopsy which was suggestive of focal minimum chronic inflammation with changes suggestive of healed arteritis. Previous right temporal artery biopsy was reevaluated at this time and was reported as healed arteritis. Subsequent renal biopsy showed MPO-ANCA related subacute pauci-immune focal crescentic glomerulonephritis and arteritis. Patient was finally diagnosed with healed temporal arteritis with new MPO-ANCA related microscopic polyangitis.

This case report discusses rare occurrence of ANCA associated renal limited vasculitis along with evidence of temporal arteritis. There have been few similar case reports in literature describing this kind of phenomenon. Pathology of this condition is debated in literature, with some authors considering it isolated disease processes. While other papers discuss an overlap phenomenon with large vessel involvement as a part of ANCA related vasculitis. Our patient was treated with Rituximab with improvement in her renal function at 1 year follow up.

REFERENCES


Disclosures: Author has no disclosures to make
Delirium – Atypical Presentation of Gouty Arthritis

Sai Koyoda., Krithika Namasivayam., Margaret Eng., Monmouth Medical Center, NJ.

Patient is a 58-year-old female with PMH of hypertension, hyperlipidemia, alcohol abuse with known Wernicke Korsakoff syndrome brought to the hospital after being found lying beside her dead boyfriend. Patient was found to be very agitated and hence sent to the psychiatry ward after initial evaluation in the ER for psychosis. Later, sent to medical floors to rule out toxic metabolic encephalopathy as a cause of acute delirium. Initial differentials were alcohol withdrawal, acute PTSD or stress disorder, infections leading to acute delirium in an already demented patient.

Patient was non-verbal, depressed, crying intermittently and at times severely agitated. Vitals were stable. Physical examination was limited by patient’s mental status however, erythema and swelling noted over the left ankle. Initial workup was unremarkable except for thrombocytosis and mild renal dysfunction which responded to fluids. Later MRI, EEG, CT chest, abdomen-pelvis, X-ray left ankle and miscellaneous blood work was un-revealing except for diffuse cerebral atrophy. However, multiple drug trials with Mirtazapine, Zyprexa, Valium and Risperidone did not help the patient but make her more sedated hence discontinued. Patient was also started on antibiotics for possible cellulitis. During physical therapy evaluation, patient was noted to not weight-bear on her left leg and was found to have tenderness in multiple small and large joints in both her upper limbs. Her inflammatory markers were high, however, further investigations evaluating polyarticular arthropathy including anti-citrullinated protein, rheumatoid factor, uric acid, ANA, Ds-DNA were with-in normal limits.

Rheumatology was consulted, who performed needle aspiration of the left ankle and right metacarpal joint with very low fluid yield. Uric acid crystals were found on microscopic analysis. Patient was started on colchicine and after three days allopurinol was also initiated. Patient’s mental status drastically improved, appeared more cheerful, was able to communicate with staff and ambulate without support before discharge.

Gout is a crystal arthropathy caused by deposition of monosodium urate crystals in joint spaces. About 40% of acute gout flares can elicit a systemic inflammatory response, the majority of the time manifesting as fever and leukocytosis. The excess cytokine production during systemic inflammatory response syndrome may have neurologic manifestations. Secondary to its effects on various neuro-transmitters, cytokines have shown to cause delirium. Delirium is an unusual presentation of an acute gout flare. Patients initial presentation was a confounding factor delaying her diagnosis. It is very well known that uric acid levels are low during acute flare due to the sudden precipitation in the joint space, hence a definite diagnosis would only be possible through a joint aspiration. It is important to recognize pain and inflammation as a cause of delirium in the at risk population -elderly and demented.

References:


Disclosure:
Author has no disclosures to make.
Massive hemoptysis- a fatal complication in sarcoidosis with airway involvement.

**Authors:** Mukund Kumar, MBBS¹; Nikhil Jagan, MBBS²; Joseph A. Nahas, MD³; Zachary S. Depew, MD²

¹Department of Internal Medicine, Creighton University Medical Center, Omaha, NE; ²Department of Pulmonary, Creighton University Medical Center, Omaha, NE ³Department of Rheumatology, Creighton University Medical Center, Omaha, NE

**Disclosure:** No relevant disclosures for any of the authors

**Introduction:**

Airway involvement in sarcoidosis can span the length from the oral cavity to the terminal bronchiole and is associated with increased morbidity and mortality. Upper airway involvement is rare, isolated and laryngeal sarcoidosis has been reported in only 0.33% - 2.1% of the cases (1-3). Trachea and main bronchi though less affected generally manifest with obstructive symptoms Hemoptysis as a presenting symptom is rare. (3-5) Herein we present a case of a 53-year-old male with fatal hemoptysis and biopsy proven laryngeal and tracheal sarcoidosis.

**Case Report:**

53-year-old male with a long-standing history of sarcoidosis of the larynx and upper airways was referred for evaluation of new onset hemoptysis. He had a tracheostomy in place and was experiencing increasing amounts of blood coming out through the tube. Apart from the tracheostomy done 12 years back he had a history significant for partial epiglottectomy with debulking of supraglottic sarcoid 12 years ago unprovoked Deep Vein thrombosis requiring chronic anticoagulation with warfarin, End Stage Renal Disease and non-ischemic cardiomyopathy.

Flexible fiberoptic bronchoscopy revealed a cobblestone pattern of the mucosa along the trachea and main bronchus with friable endobronchial tissue was visualized with multiple areas of profuse oozing. Multiple tissue biopsies along the midline trachea performed, demonstrating focal poorly-formed non-necrotizing granuloma consistent with sarcoidosis. GMS silver Nitrate and AFB stains were negative for infectious organism and malignant cells were absent on cytology. Sarcoidosis with tracheal involvement was diagnosed based on clinical history, physical exam and corroborative histologic findings. Oral Prednisone 40 mg once daily with a plan to taper was initiated and warfarin discontinued resulting in clinical improvement. Eventually his symptoms of hemoptysis relapsed, and he presented on multiple occasions to the hospital with similar complaints. This led to multiple hospital admissions and bronchoscopy’s requiring aspiration of blood, removal of clot via the cryoprobe Pulse dose of intravenous methylprednisolone followed by oral Prednisone resulted in transient improvement. Infliximab was initiated due to steroid induced cushingoid features and concerns for mucosal thinning and worsening of bleeding Methotrexate and Azathioprine was avoided due to ESRD. After 3 months of similar presentations, he unfortunately had an episode of massive hemoptysis and subsequent Cardiac arrest with unsuccessful resuscitation.
Discussion:
This case report highlights the rare yet potentially fatal manifestation of sarcoidosis when involving the larynx and trachea. It is estimated to occur in 3.5-6% of sarcoid patients. (4) Hemoptysis is usually seen in advanced pulmonary sarcoid with fibrosis and cavitary lesions with development of an aspergilloma. We did not find evidence of underlying fibrosis, cavitary disease, fungal infection or malignancy in our patient. Management with initial corticosteroid therapy, bronchial artery embolization and/or surgery which has been individualized has been reported however there is a lack of evidence and a standardized approach.

Conclusion:
Sarcoidosis of the upper respiratory tract and trachea is exceedingly rare with the need for a high index of suspicion. Bronchoscopy plays significant role diagnostic and therapeutic role. Further studies will aid in the development of comprehensive management strategies to potentially treat this fatal complication.

References


A case of Posterior Reversible Encephalopathy Syndrome in a patient taking Methotrexate and Abatacept

Authors: Luis Lora, MD; Adria Madera, MD.

Background
Posterior reversible encephalopathy syndrome (PRES) is a neurological disorder caused by various etiologies. It is a clinical radiographic diagnosis with characteristic neuroimaging findings of posterior cerebral white matter edema in a patient that presents with a clinical syndrome of headache, altered mental status, visual abnormalities, and seizures. PRES has been associated with multiple immunosuppressive and cytotoxic agents, including methotrexate, but there are no reported cases of its association with abatacept. Here we report a case of a patient with rheumatoid arthritis, treated with methotrexate who was started on abatacept and 4 days later presented with a syndrome of altered mental status and visual hallucinations.

Case presentation
A 51-year-old female with a past medical history of asthma and hypertension was treated for seronegative rheumatoid arthritis (RA) with methotrexate 15 mg/week for 30 years. The patient was started on abatacept due to uncontrolled symptoms associated with RA. Four days after the first infusion, the patient was found to be encephalopathic, with a temperature of 102°F and was admitted to the critical care unit with suspected CNS infection for which she was started on empirical IV antibiotics. Both, methotrexate and abatacept were discontinued. On admission, CT of the head was suggestive of PRES (Figure 1). CSF analysis was consistent with an inflammatory process. It showed mildly elevated white blood cells (76/mcL) with neutrophilic predominance (90%), mildly elevated protein (80 mg/dL) and normal glucose (72 mg/dL). Within 24 to 48 hours, patient had almost complete resolution of altered mental status but complained of blurry vision and visual hallucinations. Further CSF studies were negative for HSV, CMV, varicella zoster, enterovirus and for cryptococcus. After 72 hours of negative CSF cultures and low suspicion for infection, antibiotics were discontinued, and the patient was discharged home. On follow up two weeks later, patient had no visual hallucinations and almost complete resolution of blurry vision. Follow up MRI brain showed near complete resolution of the bilateral occipital lobe process seen on previous CT head (Figure 2).

Conclusion
PRES has been well reported with the use of methotrexate but there are no cases reported in association with abatacept. With more reported cases such as this, clinical suspicion can be raised of CNS manifestations and the use of abatacept in combination with methotrexate.

Funding
None to disclose
Figure 1: Extensive areas of low-attenuation, right greater than left, throughout the posterior aspects of the bilateral occipital lobes.

Figure 2: No confluent T2/FLAIR hyperintensity consistent with near complete resolution of the bilateral lobe process seen on previous CT of the head.
Barriers to Medication Adherence among Hospitalized Patients with Systemic Lupus Erythematosus
Mithu Maheswaranathan¹, Trevor Faith², Holly C. Mitchell², Diane L. Kamen²

¹ Division of Rheumatology, Duke University Medical Center
² Division of Rheumatology, Medical University of South Carolina

The authors have no conflicts of interest to disclose.

Background/Purpose:
Lack of medication adherence is a known risk factor for increased morbidity and mortality among patients with systemic lupus erythematosus (SLE). Low adherence is associated with higher cost acute care utilization and more severe SLE disease activity and damage. In addition to known influences on adherence such as education, socioeconomic status, and polypharmacy, we hypothesized that patient-identified barriers and facilitators to adherence would also impact medication adherence rate and SLE-related outcomes. The purpose of this study was to use survey methodology to characterize medication adherence quantitatively and qualitatively among hospitalized patients with SLE.

Methods:
Hospitalized patients with SLE being seen by Rheumatology consult physicians were invited to participate in an IRB-approved survey, administered in-person by an interviewer not on the patient’s care team. Demographics, medication use, pill counts, information on finances, insurance status, support systems, and other potential factors influencing adherence were elicited. Electronic medical record review was performed to confirm outpatient medications, calculate pill burden and determine length of hospital stay. Pearson’s chi-squared and Fisher’s Exact testing were performed for categorical measures. Two-sample t-tests were performed for continuous measures.

Results:
All 13 of those interviewed were non-Hispanic African American females with SLE, with a mean age of 31.0 ±12.8 years. 69.2% had graduated high school, 46.2% were employed, 69.2% had Medicaid or Medicare and 23% had private insurance.

Eight patients (61.5%) reported medication nonadherence during the 2 weeks prior to hospitalization, including 6 patients who reported forgetting medications and 8 patients who reported being unable to obtain medications. Nonadherent patients were younger (27.6 ± 7.6 vs. 36.4 ±18.2 years, p=NS) and had a longer hospital length of stay (10.1 ±6.8 vs. 8.2 ±3.1 days, p=NS) compared to adherent patients, although not statistically significant. There was no difference in pill burden (mean number of pills prescribed per week) between nonadherent and adherent patients (103.7 ±42.6 vs. 108.7 ±95.2 pills, p=NS). Of patients with a higher-than-median pill burden, 75% reported medication nonadherence. Several patients in both groups
also received infusions, injections and topical medications. Perceived lack of control over complex medication regimens was a universal theme among patients.

Cost was identified as a barrier to adherence, with 23.1% reporting being self-pay with no insurance assistance for medications. Family support was the most frequently reported facilitator to adherence with 92.3% reporting that family is at least somewhat supportive or very supportive in helping manage their SLE. Utilizing a pill organizer to help facilitate medication adherence was reported by 38.5%, but the majority did not use any reminder techniques.

**Conclusion:**
This pilot study identified a high prevalence of medication nonadherence leading up to hospitalization and several modifiable barriers to patients affording and remembering their medications. While nonadherent patients did not have statistically significant longer hospital stay duration or higher pill burden, these findings were limited due to the small sample size. These findings will help inform planned interventions to improve medication adherence among patients with SLE.
Mycosis fungoides: The Great Imitator

Kanchan Malhotra MD, Nestor E Dela Cruz, MD, Sarwat Umer MD

The authors have no disclosures.

Introduction:
Mycosis fungoides (MF) is considered to be the most common cutaneous T-cell lymphoma. The hallmark of this disease is that the lymphocytes are present in the epidermis, a phenomenon termed “epidermotropism.” In MF, there is clonal multiplication of the skin which contains mature T cells, usually CD4. We present a patient who was referred to the Rheumatology clinic for a rash of unknown etiology. Several different diagnosis of her rash were made including lupus, fungal rash, among others and patient was tried on multiple different medications but had no improvement. Eventually, a repeat biopsy came back positive for Mycosis fungoides. This case highlights how Mycosis fungoides can mimic the clinical findings found in other inflammatory skin disorders and the importance of continuing to look for a diagnosis when the patient does not improve with initial treatment.

Case presentation:
Patient is a 54 year old African American female with a PMH of Fibromyalgia and DM2 that presented to the clinic with complaints of a rash on the chest, neck, abdomen and back for about 4 years. She was originally seen and treated by an outside dermatologist for eczema with topical creams but no improvement was noted. Past skin biopsy showed spongiosis and non-specific inflammation but immunofluorescence was unremarkable. On physical exam, erythematous, raised rash in the chest, neck, and abdomen was noted. Pt was started on Methotrexate but the rash worsened. Patient was subsequently switched to Arava as well as Prednisone but no improvement was noted. As the skin rash persisted, the skin biopsy was repeated and showed CD3 inflammatory infiltrate suspicious for Mycosis fungoides without T-cell gene rearrangement positivity on PCR testing. Pt was then referred to Hematology/Oncology to start treatment.

Discussion:
Mycosis fungoides (MF) has been termed a great imitator as it can resemble many other diseases clinically or histopathologically. It has no cure unless the patient presents with a very early stage of the disease. The etiology of MF is not clear, although associations have been made with human T-lymphotrophic virus-1 (HTLV-1), CMV, chromosomal deletions and translocations. Classic MF usually includes erythematous patches or thin plaques and patients later progress to the advanced stage which includes tumors. Other diseases which can also present with lymphocytic infiltrates in the dermis include cutaneous T-cell lymphoma, leprosy, and lupus erythematosus, Hansen’s disease, Lupus, lymphocytoma cutis, lymphocytic infiltration of Jessner, and Lymphoma. The prognosis depends upon the degree of skin involvement. Treatment includes topical steroids, nitrogen mustard, imiquimod, UVB therapy, local radiation therapy, retinoids, interferons, and Methotrexate. MF puts patients at a high risk of developing secondary malignancies including B-cell lymphoproliferative disorders.
The presence of Calcinosis cutis universalis in an adult patient with Dermatomyositis

Kanchan Malhotra MD, Rina Musa MD, Samina Hayat MD

The authors have no disclosures.

Introduction

Calcinosis cutis is a condition in which calcium phosphate deposits are formed in the skin. These insoluble deposits can present as subcutaneous, intracutaneous, or intramuscular. Calcinosis cutis can occur in up to one third of patients suffering from Juvenile Dermatomyositis (JDM), however, it is rare in adults with dermatomyositis. We present the case of a female patient who came to the Rheumatology clinic with a history of Dermatomyositis and weakness in bilateral lower extremities. She also had calcium deposits all over her body and was diagnosed with Calcinosis cutis universalis.

Case presentation

35 y.o. AAF with history of Dermatomyositis who presented to the Rheumatology clinic. Patient was originally diagnosed with Dermatomyositis in her 20s after workup was done for weakness in the bilateral lower extremities. When patient was first seen in clinic, she had bilateral lower extremity weakness and was wheelchair bound. She also had multiple ulcerations over her breast, abdomen and upper arms. The patient was started on Colchicine for the calcifications. Methorexate and steroids were continued with improvement in the muscle weakness. As she had no improvement of her calcifications with the Colchicine, Humira was added. The ulcerations and calcifications improved and have healed completely. She has continued to do well with her strength 5/5 in both upper and lower extremities.

Discussion

Calcinosis cutis is a pathological condition where there is deposition of calcium phosphate crystals in the skin. It was first mentioned in the year 1855 by Virchow. These calcium deposits are insoluble and can be subcutaneous, intracutaneous, or intramuscular. It can be divided into 4 different types: dystrophic, idiopathic, metastatic, and iatrogenic with dystrophic being the most common type. The diagnosis involves a thorough history and physical exam, X-rays of involved organs, and biopsy of the areas. Whole body bone scintigraphy can be used to detect extrasosseous calcification. Once diagnosis has been made clinically, labs must be obtained to rule out other diseases that can cause abnormalities of calcium and phosphorous metabolism, malignant processes, Vitamin D poisoning, and collagen vascular diseases. Calcinosis usually occurs in patients that have long and uncontrolled dermatomyositis but can also occur in severe and aggressive disease. Calcinosis can be found in other conditions including autoimmune connective tissue disorders, cutaneous neoplasms, and hereditary disorders. The treatment is medical or surgical with surgery being the more effective option. Some medications that have been used to treat it include bisphosphonates, aluminum hydroxide, warfarin, IVIG, TNF-α inhibitors, Abatacept, Rituximab, intra-lesional steroids, and colchicine. Recurrence after surgical excision of lesions can occur.
Sarcoidosis or Cryptococcosis: Which Came First? A Puzzling Clinical Association

Harman Fervaha MD, Matthew Malus MD, Puja Nambiar MD, Mamatha Katikaneni MD

The authors have no disclosures.

A 60-year-old African American female with a past medical history of hypertension, diabetes mellitus and hypercholesterolemia presented with a three-month history of fever, fatigue, night sweats, shortness of breath and a twenty-pound unintentional weight loss. Imaging revealed pericardial tamponade requiring an emergent pericardial window. Analysis of the pericardial fluid revealed reactive mesangial cells, with negative bacterial, fungal and AFB culture. An extensive infectious disease work up including syphilis testing with RPR, acute Hepatitis panel, HIV antibody, T spot, Histoplasma serum antigen, blood bacterial, fungal and AFB cultures were all unrevealing. On CT imaging of chest and abdomen, there was hilar, mediastinal, portocaval and hepatic lymphadenopathy along with multifocal hypo vascular lesions in the liver. Liver biopsy showed multiple well-formed granulomas and multiple asteroid bodies concerning for Sarcoidosis. Treatment was initiated with Prednisone 60 mg daily and Hydroxychloroquine 200 mg twice daily. After failing a trial of Methotrexate owing to nausea and diarrhea, she was started on Leflunomide and titrated up to 20 mg daily while tapering Prednisone gradually to 20 mg daily. Minimal clinical improvement was seen with this therapy.

Three months into immunosuppressive therapy, the patient developed fever, bilateral frontal headache, weakness in her legs and staring episodes concerning for seizures. Lumbar puncture resulted with an opening pressure of 30 cm of water, elevated protein (117 mg/dl), elevated glucose (85mg/dl), 90 WBC’s with elevated Neutrophils (27). MRI of the Brain was unremarkable. The CSF viral encephalitis panel, gram stain, bacterial cultures, AFB smear/culture and fungal smear were negative but the fungal culture grew Cryptococcus Neoformans at Day 10. The cryptococcal meningitis was treated with induction therapy with intravenous liposomal amphotericin B 1mg/kg/day along with oral Fluocytosine 25 mg/kg every 4 hours for 2 weeks followed by maintenance Fluconazole 400 mg daily while her Prednisone and Leflunomide were stopped. Two months later, while on antifungal therapy alone, she is asymptomatic and a repeat CT chest and abdomen shows complete resolution of the prior noted adenopathy and liver lesions.

Sarcoidosis is a granulomatous disease of unknown etiology associated with an impaired T-lymphocyte function and cell-mediated immunity thus predisposing to infections due to intracellular pathogens like cryptococci. It is yet to be determined if a dysregulated immune response to the cryptococcal antigen can lead to sarcoidosis or a sarcoid like reaction. With the growing body of evidence of this association, it is prudent to consider cryptococcal infection in patients with Sarcoidosis that is not responding appropriately to steroid therapy and to screen all Sarcoidosis patients with serum cryptococcal antigen to potentially prevent a life threatening complication in these immunosuppressed and at-risk patients.
Autoantibody-Mediated Extreme Alterations in Serum Glucose as the Initial Presentation of a Diagnosis of Systemic Lupus Erythematosus

AUTHOR:
Taylor A. McConnaughy, M.D. Internal Medicine Residency Program

INTRODUCTION:
Autoimmune hypoglycemia is a rare phenomena which is caused by the presence of insulin antibodies as seen in insulin autoimmune syndrome (IAS) or by the presence of anti-insulin receptor antibodies as seen in type B insulin resistance syndrome (TBIRS). TBIRS can cause hypoglycemia, but the majority of patients have insulin resistance. The following is an unusual case of an initial diagnosis of Systemic Lupus Erythematosus (SLE) in the setting of autoantibodies causing both hypoglycemia and extreme insulin-resistant hyperglycemia.

CASE PRESENTATION:
A 42-year-old Hispanic female with diabetes, not previously on insulin, who presented to the hospital from her PCP’s office for six months of hypoglycemic episodes. Physical exam demonstrated normal body habitus and acanthosis nigricans. Hospital laboratory work up included HbA1c of 5.3, normal C peptide and serum insulin levels, pancytopenia, positive ANA, anti-smooth muscle antibody, smith antibodies, RNP antibodies and low C3 and C4. Imaging was negative for pancreatic mass. Furthermore, she had elevated serum creatinine and hematuria, which prompted a renal biopsy and confirmed lupus nephritis class V. During her hospitalization, she was persistently hypoglycemic down to as low as 30mg/dL requiring dextrose drip until she was treated with high dose prednisone for her lupus nephritis. This, however, then resulted in extreme hyperglycemia requiring up to 800 units of insulin per hour to maintain serum glucose below 300mg/dL. A serum insulin level was obtained during this time and resulted 12,061mU/L. Further lab work demonstrated elevated insulin antibodies, with an indeterminate anti-insulin receptor antibody. Her blood glucose levels normalized after cessation of steroids and fluid resuscitation and she did not require any insulin on discharge.

DISCUSSION:
To my knowledge, this patient is the first case of such extreme autoimmune-mediated insulin resistance in an SLE patient. Insulin resistance and hyperglycemia has been seen in TBIRS, however, this patient’s insulin receptor autoantibody was indeterminate. Her insulin antibodies were positive, which is suggestive of IAS, however, resistant hyperglycemia has not been described in the literature with this disease. The high dose prednisone used to treat lupus nephritis likely exacerbated her extreme insulin resistance but potentially may have also treated it as immunosuppression is how TBIRS and IAS are managed. In conclusion, autoantibody mediated hypoglycemia and extreme hyperglycemia should be included in the long list of autoimmune phenomena associated with SLE. Antiinsulin receptor antibodies and insulin antibodies should be looked for in SLE patients with idiopathic hypo- or hyper- glycemia.

DISCLOSURES: None.
A case of catastrophic antiphospholipid syndrome with severe thrombocytopenia complicated by the development of disseminated intravascular coagulopathy.

AUTHORS: Artem Minalyan¹, Babatunde Ogunnaike¹, Ida Micaily¹, Rajeshkumar Patel²

¹- Department of Internal Medicine (Abington Hospital - Jefferson Health), ²- Department of Pulmonology and Critical Care (Abington Hospital - Jefferson Health)

INTRODUCTION: Catastrophic antiphospholipid syndrome (CAPS) is a rare form of antiphospholipid syndrome (APS) characterized by multiorgan failure and generalized small vessel thrombosis.

CASE: A 42-year-old previously healthy Caucasian female presented to our hospital with generalized weakness, right lower quadrant abdominal pain, fever for 2 days. She was found to be febrile (T 101.3F). Her initial blood tests were unremarkable. CT of the abdomen and pelvis (CT A/P) showed generalized lymphadenopathy. On day 2, became hypotensive with worsening of abdominal pain. Repeat CT A/P was significant for abdominal and pelvic ascites with mesenteric edema. Started on broad-spectrum antibiotics. Intubated for increased work of breathing. On day 3, developed multiorgan failure: renal failure, leukocytosis, elevated transaminases, thrombocytopenia, and anemia. On day 4, developed purpura-like rash on upper and lower extremities. HBV, HCV, ANA, anti-dsDNA, MPO, PR3, SSA, SSB, Sm, RNP, Scl-70, RF, CCP, anti-centromere, anti-GBM, Toxoplasma antibodies - all came back normal. Blood culture - no growth. C3, C4 were low. Cytomegalovirus IgM and IgG were significantly elevated. Lupus anticoagulant, cardiolipin IgM were positive. B2-GP antibodies were negative. Started on pulse-dose steroids as well as plasmapheresis for CAPS. Given severe thrombocytopenia (~11K/UL) and laboratory findings consistent with disseminated intravascular coagulopathy (DIC), anticoagulation was not initiated. Serotonin release assay (SRA) was negative. Biopsy from right upper extremity was obtained and showed diffuse subcutaneous vessel thrombosis. On day 6, rash worsened with development of necrotic changes with blistering. Patient was ultimately started on heparin drip, and high-dose steroids were continued. Her mentation improved. She was extubated. Her rash
slightly improved, however, she did develop some irreversible ischemic changes which might need surgical intervention after a comprehensive rehabilitation.

**DISCUSSION:** CAPS is a life-threatening form of APS. In about 50% of patients it can be an initial presentation of APS. Most of the time, there is a precipitating factor (infections and surgery are the most common ones). Given generalized lymphadenopathy and significantly elevated anti-CMV IgM and IgG levels, a viral infection might have triggered CAPS in our patient. DIC is a rare complication of CAPS and may be associated with a worse prognosis.

**CONCLUSION:** Although a rare condition, CAPS should be always considered in a patient with acute multiorgan failure with generalized small vessel thrombosis.

**DISCLOSURE:** The following authors have nothing to disclose: Artem Minalyan, Babatunde Ogunnaike, Ida Micaily, Rajeshkumar Patel
How Sweet of A Flu
Arash Mollaeian MD, Hadi Rudsari MD, Ebrahim Talebi MD
Medstar Health Internal Medicine Residency Program, Baltimore

Introduction:
Acute febrile neutrophilic dermatosis or Sweet’s syndrome is a rare inflammatory condition that is characterized by acute onset of painful papulonodular skin lesion in the setting of prodromal fever, malaise, arthralgia, neutrophilia and pathological findings of neutrophilic infiltration of the upper dermis in the absence of leukocytoclastic vasculitis. It is generally classified into three categories of classical (idiopathic), malignancy-associated and drug-induced Sweet’s syndrome. [1-5]

Case Presentation:
A 41 years old woman with past medical history of insomnia and anxiety presented with fever (as high as 103°F), sore throat and generalized body pain for six days, accompanied by a painful rash involving lower extremities that later progressed to the trunk. During this period she visited emergency department twice and was diagnosed with a flu-like illness and treated conservatively. However her symptoms did not improve and she developed swelling of bilateral elbows, wrists, and MCPs as well as a watery non-bloody diarrhea for two days before admission. Upon her third presentation to the ED she was febrile with a temperature of 39.8°C and appearing ill. She was noted to have symmetrical tender swelling of elbows, wrists and metacarpophalangeals with decreased active and passive range of motion and dark erythematous, tender, nodular rash in bilateral thighs, abdomen, chest and back. Her initial labs were significant for increased ESR to 85 mm/hr and CRP to 131 mg/L without leukocytosis, neutrophilia or bandemia. Blood cultures were drawn, she was started on antibiotics and admitted to general floor. Her infectious lab work up was negative including HIV, monospot, influenza A and B, HCV, HAV, HBV, HCV, chlamydia and gonorrhea. She was taken off antibiotics and got skin biopsy. Symptoms including fever and diarrhea continued and cultures remained negative. Rheumatology was consulted and patient was started on pulse steroid therapy with methylprednisolone 125 mg IV daily on third day. Immunological work up revealed positive ANA of 1:80 (RO/SSA pattern). Her ANCA, C3, C4, anti Ds-DNA, RF, anti-CCP, RNP Ab, ACA IgM /IgG were negative or within normal limits. Her symptoms including the rash improved on day four, she was switched to PO prednisone 40mg daily and discharged on prednisone taper. Later the skin biopsy revealed dermal aggregates of neutrophils and she was diagnosed with classical Sweet’s syndrome in the setting of a viral infection. She was evaluated by oncology and full work up was unremarkable.

Discussion:
Acute febrile neutrophilic dermatosis or Sweet’s syndrome is characterized by acute onset of tender papulonodular skin lesion with histopathologic findings of dense neutrophilic infiltration of the dermis without evidence of vasculitis. [1-5] Currently the syndrome is classified into three major categories of
classical (idiopathic), malignancy-associated and drug-induced Sweet's syndrome. [4-6] The classical Sweet’s syndrome is the most common type, predominantly affects middle-aged women and is usually associated with an infectious process, inflammatory bowel disease and pregnancy. [5-8] Patients most commonly present with fever preceding the skin lesions that may be accompanied by general malaise, arthralgia, headache and other symptoms resembling a flu-like illness. [5-10] The underlying pathogenesis of this syndrome is yet to be clarified, however it is multifactorial and current evidence is suggestive of a hypersensitivity reaction to exogenous antigens. [5-10] Biopsy of skin lesions is pivotal to diagnosis. [5-8] First line treatment consists of corticosteroids as well as potassium iodide and colchicine. Second line treatments are also available such as clofazimine, cyclosporine, dapsone and indomethacin. Novel approaches to treatment have also been reported such as immunoglobulin and Anakinra (IL-1 receptor antagonist). [5-10] Future research potential lies in further investigation of underlying pathogenesis and association, prevalence and prognostic value in other autoimmune diseases such as RA and SLE [5-10]

Disclosure:
Authors declare that they have no conflict of interest.

References:

**Introduction:**

Microscopic polyangiitis (MPA), an autoimmune systemic vasculitis involving small and medium sized vessels, often times affecting kidneys and lungs, is classified as a subset of anti-neutrophil cytoplasmic antibody-associated vasculitis (AAV) [1-3]. Anti-neutrophil cytoplasmic antibody (ANCA) is detected in 80-90% of cases of MPA and its detection has become an essential step in diagnosis, however rare cases of ANCA-negative MPA have been reported [1,5].

**Case description:**

A 23 years-old male presented with dyspnea, pleuritic chest pain and hemoptysis for 2 weeks. He was found to be hypertensive (BP 223/136) and tachycardic. Physical exam was unremarkable except for bilateral wheezing in lungs. CT chest was significant for bilateral centrilobular opacities and hilar/mediastinal lymphadenopathy. He also was found to have acute kidney injury and trace proteinuria. He was initially started on antibiotics and steroids and was worked up for autoimmune diseases. CRP (86) and ESR (67) were elevated. ANA was weakly positive (1:160), however other factors and antibodies including anti-ds-DNA, RF, anti-CCP, p-ANCA, c-ANCA, Scl-70 Auto Ab, GBM Ab, SS-A, SS-B, anti-Sm Ab, C3, C4, anti-histone Ab and other autoimmune related factors were negative or within normal limits. Echocardiogram revealed EF of 40-45% as well as moderate concentric left ventricular hypertrophy. Infectious disease work up was unremarkable. In the light of unrevealing work-up and persistent symptoms, patient eventually had lung biopsy, which revealed extensive intraalveolar hemorrhage with linear PMN collections in alveolar septa, and capillaritis indicative of MPA. He lost follow up and was subsequently readmitted. He had kidney biopsy for worsening proteinuria, revealing focal segmental glomerulosclerosis and changes concerning for MPA as well as tubular atrophy and interstitial fibrosis.

**Discussion:**

MPA classically presents with acute onset of rapidly progressive glomerulonephritis, however the presentation is not often stereotypical and most commonly present with constitutional symptoms and multiple organ involvement in 5th-6th decades of life with a slight male predominance [1-3]. Most common organs involved are kidneys and lungs, followed by skin, gastrointestinal, cardiac and nervous systems [1,4]. ANCA is detected in 80-90% of cases of MPA (70% MPO-ANCA), and plays an important role in diagnosis and prognostication [1-2]. Negative ANCA may delay the diagnosis or lead to misdiagnosis of MPA. ANCA may become positive later in the course of disease. In cases with high clinical suspicion for MPA (pulmonary-renal syndrome) and other ANCA-associated vasculitis (AAV), tissue biopsy is necessary to make a definite diagnosis and initiate appropriate management [1, 2, 5]. Treatment consists of induction phase followed by maintenance phase. Induction usually includes pulse
and/or standing steroids, plasmapheresis, rituximab or cyclophosphamide based on severity of symptoms and organs involved [1-3]. Maintenance generally is achieved by azathioprine, rituximab or methotrexate, depending on individual factors [1-3]

Disclosure:

Authors declare that they have no conflict of interest.

References

Multidisciplinary Approach to Digital Ischemia and the Role of Rheumatology in Buerger’s Disease

Meriah N. Moore, MD and Rory M. Marks, MD
Department of Rheumatology, University of Michigan, Ann Arbor, Michigan

Background
Thrombangiitis obliterans, also known as Buerger’s disease, involves inflammatory thrombotic occlusions of the arteries and veins of the extremities. While tobacco and marijuana are strongly associated with development of the disease, the pathogenesis is not fully understood.

Case
A 31 year old female presented for evaluation of multiple digital ulcerations. One month prior to presentation, she developed pain in her fingers with constant blue color change with no correlation to temperature change. She developed ulceration of the left 3rd digit prompting presentation to the emergency department. Her past medical history included use of tobacco (17 pack-years) and daily marijuana. Exam revealed 0.5 cm ischemic ulcer at the distal left 3rd digit. Over several days, she developed additional ulcerations of the digits. Bilateral ulnar pulses were unable to be palpated. Basic blood counts, chemistry, urine and coagulation studies were normal. Anti-nuclear antibody (ANA) screen was positive to 1:160 (speckled), anti-ribonucleoprotein (anti-RNP) positive to 170, but ANA by immunofluorescence was negative. Antineutrophil cytoplasmic antibodies, IgG4 subtypes, double stranded DNA, antiphospholipid antibodies and complement were negative or normal. Hepatitis and HIV testing was negative. SPEP, UPEP, LDH were normal. CT angiogram demonstrated occluded bilateral distal ulnar arteries and right dorsalis pedis artery occlusion.
Treatment included tobacco cessation, aspirin 81 mg and sildenafil 20 mg TID. Increased size of the ulcerations prompted use of epoprostenol 0.5-2ng/kg/min IV titrated to blood pressure for 5 days.

Conclusion

The differential diagnosis for digital ischemia is broad and includes vasospasm, vasculopathy (Buerger’s disease), vasculitis, atherosclerosis, thromboembolic disease, hypercoagulable states, drugs, vibration injury or frost bite. Rheumatology may be consulted for consideration of associated diseases such as systemic sclerosis, systemic lupus erythematos, myositis, Sjogren’s syndrome, and undifferentiated or mixed connective tissue disease. Rheumatology may be uniquely experienced in the management of vasodilatory medications commonly used for Raynaud’s digital ischemia.

Disclosure Statement

The authors have no actual or potential conflict of interest related to this presentation.

Clinical Implications

Buerger’s Disease should be considered a diagnosis of exclusion. Multidisciplinary input from vascular surgery, hand surgery, and rheumatology guide evaluation and management beyond recommendations for smoking cessation. Rheumatology may be uniquely experienced to manage vasodilatory medications more commonly used in the treatment of Raynaud phenomenon associated digital ischemia.
Characteristics Associated with Greater Corticosteroid Requirement in Patients with Giant Cell Arteritis

Morin, SJ, D.O.1; Reeder, DJ, M.D.2; Danve, A, M.D.3; Baker, JF, M.D., M.S.C.E4; Sehra, ST, M.D.5

1 Mount Auburn Hospital, Department of Medicine
2 University of California Irvine, Department of Allergy and Immunology
3 Yale-New Haven Hospital, Department of Medicine, Division of Rheumatology
4 Corporal Michael J. Crescenz Veteran’s Affairs Medical Center and the University of Pennsylvania.
5 Mount Auburn Hospital, Department of Medicine, Division of Rheumatology

Abstract:

Objective: To determine factors associated with higher dose of steroid use in patients with giant cell arteritis (GCA).

Methods: Retrospective cohort study of newly diagnosed GCA patients during the time period of July 2014 - July 2016. Inclusion criteria: patients treated at Mount Auburn Hospital in Cambridge, Massachusetts, new diagnosis of GCA, follow up of at least six months after initiation of treatment at the hospital or the rheumatology clinic. Exclusion criteria: patients lost to follow up and those who were unable to attend regular follow up for at least 6 months. The outcome was defined as factors associated with higher corticosteroid dose at multiple time-points including at 6, 9, and 12 months after the first dose of corticosteroids.

Results: 17 patients (3 male; 14 female) were included with an average (SD) age of 79 (8.4) years. Baseline factors associated with greater steroid dose requirement at 6, 9, and 12 months included the duration between symptom onset and first dose of prednisone (95% CI: 0.003 to 0.103, p<0.039), jaw claudication (95% CI: 2.83 to 9.54, p<0.001), higher CRP at disease onset (95% CI: 0.001 to 0.032, p<0.032), younger age (95% CI: -0.459 to -0.057, p<0.012, and a positive temporal artery biopsy (95% CI: 3.93 to 8.77, p<0.001). In multivariable analyses, these associations were independent of the initial steroid dose. Jaw claudication (p=0.004) and longer symptom duration (group average 33.4 days) prior to initiation of glucocorticoid therapy (p<0.001) were both found to be statistically significant.

Conclusion: This is among the first studies to identify baseline factors affecting steroid dose during the course of initial GCA treatment. More severe disease features and greater delay from symptom onset were important and highlight the importance of early diagnosis.

Disclosure: The authors of this study have nothing to disclose.
Antineutrophil Cytoplasmic Antibody-Associated Vasculitis with atypical manifestation of renal involvement

Malahat Movahedian, Leila Muhieddine

Background

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) represents a group of systemic, necrotizing vasculitides that involve small-sized blood vessels and includes a wide spectrum of organ manifestations. (1) AAVs are rare, with an estimated worldwide annual incidence ranging from 1.2 to 2 cases per 100,000 individuals. (2)

Case

62-year-old female presented with myalgias, arthralgias, and dyspnea starting 6 months prior. Laboratory results showed negative Anti-Nuclear Antibody, normal kidney function, leukocytosis (21.7 K/μL), thrombocytosis (628 K/μL), positive rheumatoid factor (titer: 85 IU/mL) and negative cyclic citrullinated antibody. CT chest revealed a small (5 mm) peripheral pulmonary nodule. She received 5 days of prednisone 40 mg daily for susceptible bronchitis with improvement of symptoms. At discharge the patient had a positive ANCA with pending results for Proteinase 3 and myeloperoxidase (MPO).

She was readmitted 6 weeks later for difficulty ambulating due to pain in feet. She reported recurrent otitis media episodes resulting in left sided Bell’s palsy. Exam revealed left side facial palsy and tenderness with hyperesthesia in plantar aspects of both feet.

Additional laboratory evaluation showed positive MPO (titer: 122 AI), normal complements, with significantly worsened kidney function (Blood urea nitrogen/Creatinine: 91 mg/dL/6.13 mg/dL). The urinalysis showed 3-5 red blood cells and total urine protein to creatinine ratio was 1,147 mg/g. She was started on pulse dose steroids for a clinical diagnosis of pauci-immune AAV.

The patient received Rituximab 1000 mg IV twice, two weeks apart. Kidney biopsy showed a severe necrotizing vasculitis, with thrombosis of arterioles and muscular arteries, severe tubulointerstitial nephritis with mild-moderate interstitial fibrosis and mild glomerulitis.

Worsening dyspnea prompted evaluation with repeat CT chest and bronchoscopy, which revealed consolidative opacities with mild alveolar hemorrhage. Given persistent worsening of kidney function, hemodialysis was initiated. Due to lack of renal or pulmonary response four weeks after the initial Rituximab dose, the decision was made to start oral cyclophosphamide 50 mg daily. The patient’s symptoms improved gradually, and she was discharged on hemodialysis with plan to reassess renal response to cyclophosphamide as an outpatient.

Conclusion

Typically, AAV manifests in the kidney as severe glomerulitis, usually with crescents. There have been reports of transformation of predominating tubulointerstitial nephritis that progresses to glomerular nephritis on repeat renal biopsy (3). Though glomerulitis is the typical manifestation of renal involvement
in AAV, it is important to recognize atypical presentations including tubulointerstitial involvement, which may represent early disease. In addition, it has been suggested that the degree of tubulointerstitial involvement may act as a prognostic indicator of severity and chronicity of renal impairment. (4,5) That may explain why this patient did not quickly respond to treatment.

Conflicts of interest

The authors have no conflicts of interest to declare.

References:


Eosinophilic Fasciitis in an Elderly Patient With New High Titer Anti-dsDNA

Leila Muhieddine, Malahat Movahedian

Department of Rheumatology, Metrohealth Medical Center/Case Western Reserve University.

Background:

Eosinophilic Fasciitis (EF) is a rare disease involving symmetric erythema and edema of the limbs and trunk followed by collagenous thickening of fascia, sparing the hands and feet. Patients have eosinophilia, elevated inflammatory markers, and hypergammaglobulinemia. Anti-Nuclear antibodies (ANA) can be positive, but Anti-dsDNA are not. The diagnosis requires a full thickness biopsy showing thickened fascia with accumulation of lymphocytes, macrophages, and plasma cells. Eosinophils are not necessarily present (1).

Case:

82-year-old female with 4-month history of intermittent erythema with progressive skin tightening in bilateral upper extremities. She reported dyspnea, sicca and limited range of motion. Exam revealed bilateral upper extremity skin tightening extending from distal deltoid to the proximal wrist with peau du orange appearance and positive “groove sign”. Incisional biopsy showed dermal edema and foci of septal hyaline sclerosis with eosinophils and plasma cells. Laboratory evaluation revealed positive ANA with dsDNA (91 IU/mL), eosinophilia 8%, erythrocyte sedimentation rate 48 mm/hr, C-reactive protein 3.1 mg/dL and normocytic anemia (hemoglobin 10.4 g/dL). Urine studies and complement were normal. Malignancy and infectious work up was negative. She was started on prednisone 40 mg and hydroxychloroquine 200mg daily with improvement and skin softening at 2-month follow up.

Discussion:

Systemic autoimmune disorders should be strongly considered during an evaluation of EF. Though the cause of EF is unknown, suggested triggers include rheumatologic disorders such as Systemic Lupus Erythematosus (SLE) and vasculitis (2). Patients with EF may have inflammatory arthritis, especially in the joints adjacent to the fascia, restrictive lung disease due to trunk fibrosis and myalgias, which may complicate an investigation of systemic disease (1,3). This patient had findings consistent with SLE such as normocytic anemia, sicca, positive ANA and high titer dsDNA. However, some of these findings may have been confounded by the pathologic effects of EF, inarguably the high titer dsDNA requires attention and thus treatment and serial monitoring was recommended. Suggestively, EF is a scleroderma spectrum disorder, however, there are several differentiating factors such the woody texture of skin (peau du orange), “groove sign” (indentation along the course of superficial veins) and lack of visceral organ involvement or Raynaud’s in EF (1).

Authors have no conflict of interest or disclosures to declare
Right Arm

References:

Declining Incidence of Cardiovascular Disease In Patients With Incident Rheumatoid Arthritis In 2000s: a Population-Based Cohort Study

Elena Myasoedova, John M. Davis, III, Veronique L. Roger, Sara J. Achenbach, Cynthia S. Crowson

**Background:** Increased burden of cardiovascular disease (CVD) in patients with rheumatoid arthritis (RA) as compared to the general population is well recognized. Several studies suggested reduced CVD mortality in RA in recent decades. Longitudinal studies on trends in occurrence of CVD events in RA patients over time are lacking. To address this gap in knowledge, we evaluated trends in incidence of CVD in patients with incident RA in 1980-2009.

**Methods:** We studied patients with incident RA (age ≥18 years, 1987 ACR criteria met in 1980-2009) followed until death, migration out of the region, or 12/31/2016. Follow-up of each decade was truncated to make the length of follow-up comparable (i.e., the 1980–89 cohort was truncated at December 31, 1996, the 1990–99 cohort was truncated at December 31, 2006 and the 2000–09 cohort was truncated at December 31, 2016). Incident CVD events included myocardial infarction (MI), stroke (ischemic or hemorrhagic), coronary heart disease death and first occurrence of any of these. Patients with CVD events prior to RA incidence date were excluded. Cox proportional hazards models were used to compare incident CVD events by decade, adjusting for age and sex. Cumulative incidence of CVD events adjusted for death from other causes was calculated.

**Results:** The study included 906 patients with incident RA (mean age 55.9 years; 69% female). There were 201, 299 and 406 patients in 1980-89, 1990-99 and 2000-09, respectively. During median follow-up of 10.6, 10.4 and 10.2 years per decade of RA incidence, CVD events occurred in 31, 38, and 31 patients. Patients with incident RA in 2000–09 had markedly lower cumulative incidence of any CVD events than patients diagnosed in 1990s and 1980s (Figure). Hazard ratios (HR) for any CVD events demonstrated a temporal reduction in CVD events among patients with incident RA in 2000s compared with incident RA in 1980s (HR: 0.52; 95% confidence interval (CI): 0.32-0.86) and a reduction compared with incident RA in 1990s (HR: 0.65; 95% CI: 0.40-1.05).

**Conclusion:** The incidence of major CVD events in RA declined markedly over time. These findings may reflect increased awareness, improved primary CVD prevention and better RA
disease management in recent years. More studies are needed to understand the determinants and implications of these data.

**Clinical Implications:** Improved incidence of cardiovascular disease (CVD) in rheumatoid arthritis (RA) may reflect increased awareness, improved primary CVD prevention and better RA disease management in recent years. More studies are needed to understand the determinants and implications of these data.

**Conflict of Interest/ Disclosure statement:** This work was funded by a grant from the National Institutes of Health, NIAMS (R01 AR46849). Research reported in this publication was supported by the National Institute of Aging of the National Institutes of Health under Award Number R01AG034676. The authors report no conflict of interest relevant to this study.
Identifying Prevalence of Pneumococcal Vaccinations Among Rheumatoid Arthritis Patients of the Rural Appalachian Population in an Academic Rheumatology Clinic

Austin N, Maggie M, Rajesh G, Ralph W

Background:

Patients with rheumatoid arthritis (RA) are often treated with Disease-modifying antirheumatic drugs (DMARD's) alone or in combination with biologic drugs that predispose them to invasive pneumococcal disease that is associated with high mortality. Hence, pneumococcal vaccinations are a critical component of RA management. However, pneumococcal vaccination rates remain suboptimal among RA patients due to several reasons including lack of awareness among the patients and health care providers. The aim of our study is to determine pneumococcal vaccination rates based on the 2015 American College of Rheumatology (ACR) and Centers for Disease Control and Prevention (CDC) recommended guidelines among RA patients in our academic based rheumatology practise and to identify care gaps if present.

Methods:

We performed a retrospective chart review of the electronic health record (EHR) of the adult RA patients seen at the outpatient rheumatology clinic within our Marshall health care system from January 1, 2019 to June 30, 2019. Demographics of the patients such as age, gender, active RA medications and pneumococcal immunization history were included in our study. The immunization data obtained was analyzed to identify if each patient had received the appropriate pneumococcal vaccination series as recommended by the current ACR and CDC guidelines.

Results:

A total of 107 RA patients were identified. Data related to patient demographics, active RA medications and pneumococcal vaccination rates of our study are outlined in table 1. 10 of the 107 patients were identified to be compliant with the current ACR and CDC recommendations for the age appropriate pneumococcal vaccine series. Among those 10 patients, 2 were between ages 19-64 years and 8 were ≥ 65 years of age.

Conclusion:

Our retrospective study showed only 10% of our rheumatoid arthritis patients are in compliance with current ACR and CDC pneumococcal vaccine recommendations. Limitations of our study include vaccinations received outside our clinic that were not documented, incomplete patient records from referring physicians in regards to immunization status, and undocumented patient
refusal of vaccines. Future steps to increase adherence rates include educating health care providers and patients on the importance of adhering to current immunization standards, ensuring adequate supply of vaccines in the clinic and integrating outside medical information into our EHR.

| Number of patients between 19-64 years of age | 70/107 (65.4%) |
| Number of patients ≥ 65 years of age | 37/107 (34.5%) |
| Number of male patients | 40/107 (37.3%) |
| Number of female Patients | 67/107 (62.6%) |
| Mean age of all patients in years | 60.6 |
| Mean duration of RA in years | 6.4 |
| Number of patients on traditional DMARD’s | 102/107 (95.3%) |
| Number of patients on biologic agents | 33/107 (30.8%) |
| Number of patients on long term oral prednisone therapy | 9/107 (8.41%) |
| **Number of patients compliant with age appropriate pneumococcal vaccination series per ACR and CDC guidelines** | 10/107 (9.3%) |
| Number of compliant Patients between ages 19-64 years | 2/107 (1.8%) |
| Number of compliant patients > 65 years of age | 8/107 (7.4%) |

Table 1. Results of patient demographics, active RA treatment and pneumococcal vaccination rates. RA = rheumatoid arthritis, DMARD = Disease-modifying antirheumatic drug, CDC = Centers for Disease Control and Prevention

The corresponding author and the co-authors of the abstract do not have any conflicts of interest to disclose.
A 60 year old Male with Felty Syndrome- Case report and Literature Review

Nneoma Onuorah MD¹, Hrudya Abraham MD², Priyanka Vashisht MD²

1. Department of Internal Medicine, Wright State University
2. Department of Rheumatology, University of Cincinnati Medical Center.

Introduction

The triad of Rheumatoid arthritis (RA), splenomegaly and neutropenia has been rarely encountered over the last nine decades with the incidence declining at an annual rate of 0.5% in the advent of the greatly expanded armamentarium of RA treatment. The challenging diagnostic nature and rarity of this disease may explain why diagnosing this severe condition may be missed leading to significant morbidity and mortality. We discuss this interesting case to shed light on the diagnostic challenges and create awareness of Felty Syndrome (FS).

Case Report

This 60 year old male with a history of alcohol abuse presented with fever and malaise. He also reported bilateral wrist and metacarpal joints pain since 10 years. For the past 6-8 months he was unable to make a fist bilaterally which was worse in the morning. His lab work showed decreased white blood cells 2.2 10E3/ liter with absolute neutrophil count (ANC) of zero, hemoglobin 14.9g/dl, platelets 122 10E3/ul. Other significant lab findings included elevated anti-nucleic antibody of 1:160, homogenous pattern, elevated cyclic citrullinated polypeptide antibody >250 units with positive rheumatoid factor of 93.2 units. He had elevated transaminases. Other serologic studies were negative. Upon further evaluation, serum electrophoresis revealed polyclonal increase in immunoglobulins and bilateral metacarpal and carpal erosions were noted on hand x-ray. Abdominal ultrasound performed revealed splenomegaly. His peripheral blood smear showed lymphopenia, neutropenia and thrombocytopenia with multiple large granulated lymphocytes. He was treated with prednisone and rituximab given his ongoing alcohol abuse and elevated transaminases. The oncology team recommended 2 doses of filgrastim he also received while admitted. So far, two doses of rituximab 1gm has been administered with appreciable symptomatic remission and improvement of ANC which is now 1267 U/L.

Conclusion

FS is a rare subtype of seropositive RA with increased mortality and morbidity. The complete triad of erosive RA, splenomegaly and neutropenia is not an absolute requirement for making the diagnosis. Persistent neutropenia with an ANC less than 2000/mm3 in RA meets criteria for the diagnosis. Despite the decreasing prevalence of this disease, RA patients are still at increased risk and the diagnosis should always be considered in RA presenting with neutropenia to enable timely and appropriate management. MTX and G-CSF has been used to treat neutropenia with successful results and splenectomy can be considered as the last resort if treatment remains refractory.

Disclosures: Authors have no Disclosures
A Unique Presentation of Aseptic Abscess Syndrome in a Patient with Inflammatory Bowel Disease.

Nneoma Onuorah¹, Gregory Mott², Avis Ware²

1. Department of Internal Medicine Wright State University
2. Department of Rheumatology, University of Cincinnati Medical Center

Introduction

Extra-intestinal manifestations (EIM) may increase morbidity in patients with inflammatory bowel disease (IBD). They affect the joints, spleen, skin, eyes and less frequently involve other organs such as liver or pancreas. Aseptic abscesses (AA) syndrome is a rare entity of the auto-inflammatory diseases characterized by the presence of recurrent deep abscesses comprising of accumulated neutrophils, histiocytes and giant cells. AA occur in association with IBD and may overlap with neutrophilic dermatosis features like pyoderma gangrenosum and sweet syndrome.

Case Report

We report a challenging case of an elderly female with a presumed history of seronegative rheumatoid arthritis and recurrent hospitalizations with incision and drainage procedures for sterile abscesses refractory to antimicrobials since one year prior to presentation. She presented with intermittent fever, frequent recurrent painful, sterile, and circumscribed deep collections in both feet and leukocytosis. Further testing showed positive antinuclear antibodies (ANA) in homogenous pattern 1:640, ANCA P3 was elevated at 52.1 U/ml, RF <10, Anti-CCP was 3 U/L. DsDNA, complement, HLA B27 were negative. Laboratory assays for HIV, hepatitis panel, Tuberculosis, fungal and viral infections were negative. Tissue Culture of the right foot and synovial fluid from right wrist showed polymorphonuclear cells with no growth after 72 hours. She was found to have ulcerative colitis confirmed by biopsy despite reporting no prior symptoms of IBD and was treated with Remicade. Only one case report in 2014 describing AA of the right ankle was found. Unlike in our patient, it was reported in a 19 year old female with Crohn’s disease and splenic abscesses.

Conclusion

This case highlights the need for accurately recognizing IBD-associated AA in the absence of diagnostic guidelines. The initial step to early diagnosis is having a clinical suspicion. Clinicians may wrongly forego immunosuppressant therapy in lieu of several antibiotics and unnecessary surgeries which may increase morbidity and mortality. Future studies should be geared towards developing a diagnostic criteria to improve recognition and timely management of this condition.

DISCLOSURES: The authors have no disclosures
A Perplexing Case of Granulomatosis with Polyangiitis

Authors: Kinal Patel, Jared Frisby, Sean Dawes Kyle Antosiek

Pulmonary-renal syndrome is a rare entity presenting as diffuse alveolar hemorrhage and acute renal failure from glomerulonephritis, which can occur simultaneously or at different times. It is usually a manifestation of an underlying autoimmune condition, more commonly a prototype of anti-glomerular basement membrane disease but can also be caused by vasculitides. We present a case of pulmonary-renal syndrome due to granulomatosis with polyangiitis without frank hemoptysis or nasopharyngeal involvement.

Patient is a 76 year old female that presented with nausea, vomiting, and general malaise. Initial vitals were significant for hypoxia requiring BiPAP support. Labs revealed a creatinine of 3.1 which was markedly elevated from a normal baseline of 0.8. Two months prior, she was admitted to the hospital for a month with septic shock and had been in acute renal failure briefly which resolved. As aforementioned, the patient returned to the hospital after 2 months with acute renal failure and significant hypoxia after which she was started on treatment for hospital-acquired pneumonia. Her symptoms failed to improve on antibiotics for hospital-acquired pneumonia. Due to suspicion for pulmonary-renal syndrome, high dose steroids were started with gradual improvement of her symptoms. This was confirmed with positive c-ANCA and PR3 serologies and hallmark findings on renal biopsy. Bronchoscopy revealed the presence of diffuse alveolar hemorrhage not overtly present before. She was tapered off steroids and was discharged home on cyclophosphamide.

This case illustrates the complexities of pulmonary-renal syndrome. Even without overt hemoptysis, pulmonary-renal syndrome should be suspected in patients with worsening and unexplained pulmonary and renal symptoms. Prompt diagnosis is warranted in such cases to potentially avoid hemodialysis and the mortality with ANCA-associated vasculitis.

Disclosure: The authors have no conflict of interest to disclose.
Does Not Vanish

Sravani Penumarty MD, Reena Khianey MD, Debra Chew MD, Tina Brar MD, Eugenio Capitle MD

Rutgers University, Newark NJ

No Disclosures

Author Classification: Fellow Poster Presentation

Introduction

Leishmaniasis caused by protozoa can mimic vasculitis presenting as a midline granulomatous disease. We report a case of a woman presenting with idiopathic nasal granuloma progressive on lowering steroids treated with immunosuppressants that progressed to systemic Leishmaniasis.

Case Report

A 43-year-old female referred by ENT for septal perforation, epistaxis and congestion the past 5 months. Patient denied any drug abuse or prescription medication use. She immigrated from El Salvador 15 years ago with no recent travel or sexual activity. On exam, had nasal septal perforation with erythema and dry crusting. Workup was negative for infection, malignancy and autoimmune serologies including ANCA. Chest X-ray with no consolidation or lymphadenopathy. Two nasal biopsies were done showing non-necrotizing nasal granuloma. She was empirically treated with high dose prednisone with significant response.

Lowering steroids over the next few months, she developed progressive dysphagia with extension of the disease to larynx. Trialed on steroid sparing agents such as methotrexate but with no response. Laryngoscopy showed a large obstructing exophytic growths from posterior pharyngeal wall. CT neck showed a pedunculated mass lesion in oropharynx may represent post granulomatous infection. Biopsy of the mass showed non-necrotizing granuloma and negative for vasculitis and malignancy. Tuberculosis, sarcoidosis and other granulomatous diseases were ruled out. Lymphoma panel, CT abdomen and pelvis and bone marrow biopsy- negative for malignancy

A few years later patient was admitted to the hospital for worsening upper airway symptoms and developed erythematous nodular tender lesions on bilateral upper and lower extremities. Workup showed negative urine and blood cultures, quant gold, EBV, hepatitis, HIV, RPR, histoplasma antigen. All serologies were negative. Skin punch biopsy of the right leg nodule was positive for panniculitis and vasculitis. Trailed on Rituximab as a steroid sparing agent but once again failed to show improvement.
Repeat full thickness skin biopsy showed amastigotes. Molecular and DNA sequencing done by CDC confirmed presence of Leishmania (Viannia) panamensis in skin. Prior biopsies from nasal septum, false vocal cord, and cricoid also sent to CDC and confirmed the diagnosis.

She was treated with IV liposomal amphotericin for 6 months along with tapering off of prednisone. Post treatment developed conjunctival lesions which were biopsy proved leishmaniasis and retreated with IV amphotericin for 6 months. Repeat CT of neck soft tissues post treatment showed resolved pedunculated mass.

**Discussion**

Leishmaniasis has an insidious onset and slow progression and can lead to destruction of the nasal mucosa, cartilage and adjacent tissues before being discovered. Leishmaniasis can mimic systemic vasculitis and skin lesions can be confused with panniculitis and erythema nodosum.

Mucosal leishmaniasis is pauci-bacillary and can be missed on biopsy. Therefore, requires further confirmation through PCR testing. Leishmaniasis should be on the differential for midline granulomatous disease.
In a Blink of an Eye: Blindness Secondary to Primary Angiitis of CNS Presenting as Recurrent Idiopathic Intracranial Hypertension

Sravani Penumarty MD, Catherine Choi MD, Reena Khianey MD, Tina Brar MD, Eugenio Capitle MD
Rutgers University, Newark NJ

No disclosures

Author Classification: Fellow Poster Presentation

Introduction

Primary angiitis of CNS (PACNS) is a vasculitis involving the small and medium vessels isolated to the brain, leptomeninges and spine. We report a case of a young woman initially presenting with idiopathic intracranial hypertension (IIH) and cerebral sinus venous thrombosis (CSVT) which progressed 2 months later to vision loss secondary to PACNS.

Case Report

A 29-year-old woman presented to the ED from ophthalmology clinic for progressive vision changes leading vision loss and bilateral frontal headaches for 5 days.

Review of systems were negative except for vision changes and headaches. Her past history included history of graves disease and an admission 2 months ago for vision changes and headaches diagnosed with IIH. IIH improved with bilateral optic nerve sheath fenestrations and therapeutic lumbar punctures. Also found to have left transverse cerebral venous sinus thrombosis and discharged on warfarin.

Physical exam was significant for bilateral exophthalmos, dilated pupils, sluggish reaction to light and decreased visual acuity. Rest of the physical exam was within normal limits.

Labs including CBC, CMP, UA, ANA, antiphospholipid antibody’s, ANCA, anti MPO, anti-PR3, NMO, infectious workup were negative for any pathology. Lumbar puncture on 1/19/19 was bloody in appearance with 64,128 RBCs, 240 WBC, 79 neutrophils, 19 lymphocytes, 2 monocytes, 99 glucose and 96 protein. Compared to lumbar puncture that was done 1 month prior (12/18/18) was clear in appearance, 4 RBCs, 1 WBC, 75 lymphocytes, 25 monocytes, 97 glucose, 32 protein.

During hospital course, malignancy was ruled out. Day 5 of hospital stay was complicated with generalized tonic clonic seizures and day 7 developed choreiform movements.

MRI brain showed new patchy nodular right occipital parenchymal enhancement with associated FLAIR abnormality. Cerebral angiogram showed diffuse vasculopathy characterized by focal narrowing and irregularity in the MCA and ACA branches.
Brain biopsy right frontal brain and dura biopsy showed CNS parenchyma with minimal chronic inflammatory infiltrate, and unremarkable dura mater. Which were non-specific findings.

EVD which was placed for elevated intracranial pressures was removed after normalization.

PACNS treated with 3 doses of cyclophosphamide and pulse IV steroids followed by prednisone taper. Patient choreiform movements resolved and no further seizure activity noted.

**Discussion**

This is an unusual presentation of PACNS causing vision loss with initially presenting as IIH and cerebral sinus venous thrombosis followed by complete blindness secondary to PACNS. Diagnosis of PACNS is rare and challenging, accounting to 1% of vasculitides. Angiography findings are often very similar to reversible cerebral vasoconstriction syndrome and very important to differentiate, to avoid unnecessary medication exposure. Angiography has 60% sensitivity and 30% specificity. Gold standard is brain and leptomeningeal biopsy. However, 25% biopsies are false negative. PACNS is treated with Cytoxan and steroids. In this patient it helped prevent progression of neurological symptoms.
Multiple Ipsilateral Femoral Stress Fractures in a Patient Taking Denosumab for Osteoporosis - A Case Report

Hristo I. Piponov, MD; Jeffrey M. Goldstein, MD; Gerald M. Eisenberg, MD

There are no conflicts of interest to disclose

Background

Atypical femur fractures (AFF) are well-known complications of antiresorptive therapy. Historically, AFFs have been mostly associated with bisphosphonates but in the past decade multiple reports of AFF in patients receiving denosumab have emerged. Denosumab is a potent antiresorptive agent approved for treatment of osteoporosis. It is a human monoclonal antibody which blocks osteoclast activation, maturation and function by binding to RANKL (receptor activator of NFκB ligand) and thus preventing it from binding to RANK (receptor activator of NFκB), its receptor on the osteoclast surface. Consequently, RANK is not activated on the surface of the osteoclast and its precursors which decreases these cells’ function and survival. As a result, bone resorption decreases and bone mass increases. There are no reports in the literature significant for multiple ipsilateral incomplete AFF linked to denosumab use.

Case Presentation

We present a case of a 74 year-old female who sustained three femoral stress fractures in her left femur. Patient is menopausal since age of 53 and has a long-standing history of osteoporosis and hyperlipidemia. She had been initially treated with bisphosphonates for 5 years and calcitonin for 2 years prior to denosumab initiation. In 2013, due to active ongoing bone loss and multiple risk factors for worsening osteoporosis she was prescribed denosumab 60mg/ml every 6 months. Patient is 98 pounds, 5’1” tall and has a body mass index (BMI) of 18.5, she is a former smoker who quit more than 15 years ago. Her total hip T-score was -2.7 at therapy initiation. She was also prescribed Vitamin D 2500IU and 1200mg of elemental calcium daily. Patient denies having any thigh or leg pain while on bisphosphonate therapy. She tolerated denosumab well without any osteoporotic fractures while maintaining excellent level of activity. Her bone density improved in her lumbar spine and left hip as measured by Dual Energy X-ray Absorptiometry (DEXA) scans in 2016 and 2018. Her total hip T-scores were -2.3 and -2.2 in 2016 and 2018, respectively.

In May of 2019, patient started complaining of pain in the left hip and thigh area aggravated by weightbearing activity. Radiographs of the left femur were taken in the office and 3 areas of proximal femur lateral cortical hypertrophy were noted. A Magnetic Resonance Imaging (MRI) study was performed which confirmed 3 stress fractures with reactive marrow and periosteal soft tissue edema in the most proximal one. Patient was referred to an orthopaedic surgery specialist for prophylactic left femur nailing and denosumab was discontinued. She sustained long intramedullary nailing fixation of the femur.
without complications and was seen at 3 weeks postoperatively weightbearing as tolerated with mild pain and slight limp.

Summary

There is emerging evidence for AFF in patients using denosumab for osteoporosis. Our case is unique because it is significant for 3 ipsilateral incomplete AFFs involving the lateral femoral cortex 6 years after discontinuation of bisphosphonate and start of denosumab therapy. In our patient, the AFFs were symptomatic and were diagnosed prior to complete displaced fracture of the femur. Complete displaced fractures are associated with much higher morbidity and are more difficult to treat and heal. High index of suspicion is needed in any patient with osteoporosis on denosumab complaining of thigh or groin pain.

Image 1. AP X-Ray view of the femur at presentation.
Image 2. Arrows pointing to lateral femoral cortical thickening and bone marrow edema. MRI images at presentation.

Image 3. 3 weeks postoperative AP pelvis X-Ray.
Late-Onset Hemorrhagic Cutaneous IgA Vasculitis: A Unique Severe Presentation

Authors:
Mark L. Riley DO*, Ana Maheshwari MD**, Ruben Peredo-Wende MD**

*Albany Medical Center Department of Medicine
**Albany Medical Center Department of Medicine, Division of Rheumatology
47 New Scotland Avenue, Albany, NY 12208

Introduction:
IgA vasculitis (IgAV) is a small vessel vasculitis that primarily affects the skin, gastrointestinal tract, joints, and kidneys. Although it is the most common systemic vasculitis in children, presentations in adults do occur, and can be more severe and catastrophic. We present a case of diffuse cutaneous hemorrhagic IgAV with primary cutaneous, neuropathic, and renal involvement.

Case Report:
A 67-year-old female with no prior rheumatological history presented with 3 months of painful, ulcerating skin lesions with malodorous discharge on her lower extremities, arthralgias, and paresthesia of the hands and feet. She has a history of chronic sinusitis, nasal polyps, asthma, and allergic rhinitis. She denied any upper respiratory, gastrointestinal, or urinary symptoms as well as any preceding illnesses. As an outpatient, she was initially treated with multiple courses of trimethoprim-sulfamethoxazole for a presumed infection with no response. Subsequently, hydroxychloroquine 200 mg twice daily and prednisone 60 mg daily were started, but the lesions continued to progress. On examination, multiple large hemorrhagic ulcerated lesions of variable size on both lower extremities, palpable purpura on the dorsum of both feet, and petechiae on the upper extremities, chest and back were noted. Labs were significant for 2.3 grams of proteinuria and hematuria with red blood cell casts on urine microscopy and positive ANA 1:160 in a homogenous pattern. Complete blood count, renal function and inflammatory markers were normal. Wound cultures were positive for Pseudomonas aeruginosa, which was treated with a 10-day course of cefepime. Extensive computed tomography imaging showed no evidence of malignancy or metastatic disease. Histological examination of a skin biopsy was consistent with leukocytoclastic vasculitis with perivascular deposition of IgA and C3. The patient was diagnosed with IgA vasculitis and treated with rituximab 880 mg for four doses and prednisone 60 mg daily. At two-month follow-up, the patient’s skin lesions and arthralgias had markedly improved, but with evidence of skin hyperpigmentation and persistent neuropathic symptoms.

Conclusion:
Although IgAV is predominantly seen in the pediatric population, this must be considered in the differential of elderly patients with severe skin rashes and other systemic manifestations. The presentation is typically more severe, may not respond to standard therapy with corticosteroids, and require the use of immunosuppressive therapy.

Disclosure
The authors of this abstract attest that we have no financial or other conflicts of interest to disclosure
A bumpy Road to Final Diagnosis

Author/Affiliation: Najmus Sahar MBBS., Asghar Ali MBBS. Wright State University, Boonshoft School of Medicine

Introduction: This unique patient case illustrates diagnostic dilemmas related to syndromes of diffuse lymphadenopathy and elevated IgG4 levels, along with variable organ dysfunction and other systemic manifestations. The more widely appreciated version is idiopathic Multicentric Castleman Disease (iMCD); however, an increasingly recognized autoimmune disorder of poorly understood pathogenesis, termed IgG4-related disease (IgG4RD), requires consideration.

Case Presentation
A 60-year-old man with history of pancreatic insufficiency presented with one-year duration of progressive diffuse lymphadenopathy. There was associated anemia, renal dysfunction, hypoalbuminemia, elevated ESR/CRP, and transient eosinophilia. Extensive work-up for infectious and rheumatological etiologies was unrevealing. PET CT scan was remarkable for increased uptake throughout the lymphoid tissues. Lymph node biopsy showed histopathologic changes concerning for iMCD; HHV-8 serology was negative. The IgG4: IgG ratio was >80% in the context of hypergammaglobulinemia involving all IgG subclasses, but a markedly elevated IgG4 subclass, raising concern for IgG4RD. The patient responded well to weekly rituximab and steroid therapy.

Discussion
Formerly called “multifocal fibrosclerosis”, now renamed as “IgG4-Related disease” is immune mediated chronic fibrotic inflammation with potential for multiple organ involvement. Swelling of organs, a history of atopy and peripheral eosinophilia are common. Characteristic histopathologic features and elevated IgG4+ plasma cells within tissue are diagnostic though neither IgG4 level nor tissues IgG4+ plasma cells are specific for diagnosis. Isolated IgG4-related lymphadenopathy has been reported in 41% cases in some studies. Based on history, laboratory findings, and histopathology, type I-Multicentric Castleman disease like IgG4-RD Lymphadenopathy was diagnosed in this case. Glucocorticoids and/or B-cell depletion induces prompt clinical response.

Conclusion
This case will increase clinician awareness of alternate diagnoses for lymphadenopathy with variable systemic syndromes such IgG4-RD and variants, in the setting of suggestive histopathology and IgG subclass patterns.

Statement of Disclosure: I have no disclosures of any kind.
A case of Refractory Neurosarcoidosis

Faye Sajjadi MD, Valetin Marian MD

Background:

Sarcoidosis is a systemic inflammatory disease of unknown etiology, and although it has an affinity for the lungs, it may affect any organ, allowing it to masquerade as other diseases. Neurosarcoidosis, a rare subtype of sarcoidosis, refers to involvement of any area of the nervous system, presenting in up to 5-10% of patients with sarcoidosis\(^1\). We demonstrate a rare case of isolated neurosarcoidosis in which appropriate treatment was later deemed difficult in light of symptom recurrence and treatment intolerance.

Case description:

A 50 year old African American male with no past medical history was brought to the hospital for altered mental status and falls. Upon admission CSF analysis revealed elevated white count with lymphocytic predominance, elevated protein, and low glucose. Gram staining, cytology, and cultures were all negative. Magnetic Resonance imaging (MRI) brain demonstrated diffuse leptomeningeal enhancement and computed tomography (CT) chest showed diffuse pulmonic tree in bud nodularity with hilar adenopathy and necrosis. Mediastinal node biopsy revealed non-necrotizing granulomas with marked hyalinization. Biopsy and imaging results in conjunction with elevated angiotensin converting enzyme (ACE) and low 25 OH Vitamin D, confirmed the diagnosis of neurosarcoidosis. He was started on dexamethasone 10 mg IV for 3 doses followed by oral prednisone. After one month, patient returned to baseline and was discharged on prednisone.

A month later, the patient was re-admitted for bizarre behavior after he had recently stopped his prednisone. On admission he was started on steroids and methotrexate (MTX) with improvement in symptoms. Infectious workup was negative and repeat imaging suggested sequelae of neurosarcoidosis. In the setting of possible steroid induced psychosis, steroids were tapered down, methotrexate was discontinued given transaminitis, and hydroxychloroquine was started. A decision to administer infliximab, a monoclonal antibody against tumor necrosis factor-\(\alpha\) was made taking into account its successful off-label use for neurosarcoidosis in conjunction with the patients failed treatment options. Infusions at a dose of 5mg/kg was initiated, continued every 2 weeks for the first 3 doses, followed by maintenance infusions every 8 weeks. In the following week, his mental status continued improving with the combination of infliximab infusion, steroid taper, and hydroxychloroquine. He was discharged with follow up for his infusions without exhibiting any further psychotic symptoms.

Summary:

Neurosarcoidosis poses as a diagnostic challenge given the atypical site of disease; especially in cases where patients present with isolated neurological symptoms without signs of an inflammatory process. Neuroimaging and LP are currently the most utilized diagnostic
modalities, as serum ACE levels are elevated in only about 25% of patients, while tissue biopsy remains the gold standard of diagnosis. CSF analysis and imaging mirror that of our patient and are reported in 20 – 40% of patients, respectively. Our case was not only a diagnostic dilemma but once established, was deemed difficult to manage, confirming the importance of both disease and management awareness amongst Rheumatologists.

Disclosure Statement: I have no conflicts of interest to disclose.
Opening the Inflammatory Floodgates: A Case of Lupus Presenting as Severe Multi-Systemic Organ Failure.

Faye Sajjadi MD, Valetin Marian MD

Background:

Systemic Lupus Erythematosus (SLE) is a chronic multi-systemic autoimmune disorder of unknown etiology. Lupus Cerebritis, a manifestation of Neuropsychiatric systemic lupus erythematosus (NPSLE) is a life threatening complication of SLE. NPSLE can present with either focal symptoms; such as cerebrovascular accident, or diffuse non-focal symptoms characterized by inflammation of brain vasculature, as seen in Lupus Cerebritis.

Despite the broad range of symptoms categorized as NPSLE and the variable incidence, there are very few cases reported to date. We present a young female with no prior diagnosis of SLE who presented for altered mental status and rapidly progressed into severe multi-organ failure.

Case Description:

A 26 year old female with no medical history was brought to the hospital for altered mentation. She experienced a seizure in route and upon arrival she was combative and hypoxic, requiring intubation. CSF analysis revealed elevated protein and pleocytosis with negative microbiology studies. Magnetic Resonance Imaging (MRI) of the brain showed significant temporal hyperdense lesions and scattered leptomeningeal enhancement. Patient was started on pulse steroids for suspected Lupus cerebritis, confirmed with elevated dsDNA antibodies and low complement levels.

She was diagnosed with severe multi-systemic lupus including cerebritis, myopericarditis, pneumonitis, pancreatitis, pancytopenia, and nephritis. Transthoracic echocardiogram (TTE) was significant for severely reduced LV function with apical ballooning suspicious for Takotsubo Cardiomyopathy. Patient required intra-aortic balloon pump (IABP) and a short course of pressors for hemodynamic support. On second day, immunosuppression with Cytoxan 1300mg was added on to the pulse methylprednisolone. Over the next two days she deteriorated and decision was made to proceed with plasmapheresis and hemodialysis. Following these interventions, patient began to show improvement including decreased oxygen requirements, resolution of cytopenia, and increased LV function. Unfortunately, on the fifth day, when she was found to have a sluggish left pupil, aphasia, and hemiplegia. Imaging revealed a malignant left middle cerebral artery (MCA) ischemic stroke with 8mm midline shift, requiring urgent hemi-craniotomy. Patient was eventually hemodynamically and neurologically and discharged on hydroxychloroquine, mycophenolate mofetil, aspirin and prednisone taper.

Summary:

Our case was incredibly challenging, considering rapidly progressive multiple systemic failure, likely due to exuberant immune complex production and deposition. She had evidence of severe inflammation in most of the vital organs, such as brain, heart, lung, kidney, reticuloendothelial system, and pancreas. Immeasurably high dsDNA autoantibodies, depleted complement, and improvement with plasmaphoresis
support an immunepathophysiologic theory. Given resolution of systemic organ failure, control of inflammation and normalization of serology; we can confirm that aggressive management with pulse steroids and cyclophosphamide, followed by plasmapheresis are mainstay treatments indicated in aggressive lupus.

Image 1, 2: T2 weighted MRI with FLAIR demonstrating areas of leptomeningeal signal throughout multiple pyriform areas.

Image 3: Non-contrast CT head showing large ischemic stroke involving the left MCA distribution

Disclosure Statement: I have no conflicts of interest to disclose.
Respiratory Failure Due to Autoimmune Encephalitis Responsive to Cyclophosphamide

Aslıhan Sen BA, Ada Baisre MD, Eugenio Capitle MD, Departments of Internal Medicine and Pathology, Immunology, and Laboratory Medicine, Rutgers New Jersey Medical School

Introduction

Autoimmune encephalitis (AIE) is antibody-mediated inflammation of the CNS, which can have various presentations, such as weakness, altered mental status, ataxia, seizures, and sensory/vision deficits. We present a case of a young patient with autoimmune encephalitis causing respiratory failure which was responsive to cyclophosphamide.

Case Report

A 29-year-old African-American man with history of recurrent pharyngitis presented with severe headaches, left hand and foot paresthesias progressing to gradual left-sided paresis, double vision, bowel and bladder incontinence, slurred speech and confusion. Treated with steroids for CNS inflammation seen on MRI, he gradually improved. Four months later, he presented with recurrence of symptoms, but with progressive right-sided weakness for two weeks. He also reported intermittent episodes of jerking movements during the interval months.

Physical Exam: Sensory deficits of the right hand and foot, right-sided weakness, and nystagmus. Labs: Significant only for mild anemia. Autoimmune: Negative ANA, Smith, dsDNA, SSB, CCP, GQ1b, and NMO antibodies. Mildly elevated SSA and RNP antibodies. LP: Consistent with inflammatory process without evidence of infection on gram stain and culture: clear, colorless CSF, glucose 68, protein 107(H), lactic acid 3.2(H), 151 WBC(H) (94% lymphocytes, 4% monocytes, 1% PMNs), 70 RBC(H), elevated IgG synthesis rate, elevated myelin basic protein, no oligoclonal bands. Additional infectious workup: Negative. Imaging: MRI showed mirroring changes consistent with inflammation in the cervical spinal cord, corticospinal tracts, and left thalamus. Interestingly, new MRI images showed near resolution of prior right-sided brain lesions. EEG: Diffuse slowing with no seizure activity. Pathology: Biopsy of the right thalamus at initial presentation had posed a diagnostic challenge. Pathology showed non-specific inflammatory infiltrate and was not consistent with infection. The pathology slides were sent to the CDC, which also confirmed absence of parasites and protozoa.

Hospital Course: Empiric antibiotics and antivirals were ineffective, and the patient's condition deteriorated, becoming too weak to walk, then lethargic and dysarthric. He then developed progressive respiratory distress and respiratory failure, requiring intubation on day 6. IVIG and pulse steroids were started due to suspicion for possible atypical Guillain-Barre. By day 8, his mental status improved, and he was extubated, but with minimal improvement by day 15. Cyclo-phosphamide was then started due to concern for severe presentation of an autoimmune process. After 3 cycles, the patient started azathioprine as maintenance therapy. He experienced improvement of weakness and sensory deficits over the next 2 years. The patient is now able to ambulate with a cane and has no difficulty with speech.

Discussion

Once considered to be a very rare paraneoplastic condition, AIE more recently has become recognized as its own entity, often with no underlying malignancy. Many antibodies associated with AIE are already recognized, the most well-known being anti-NMDA receptor antibodies, and more are being rapidly discovered. When it is recognized and immunosuppressive treatment is started early, AIE is potentially reversible, but in younger patients who relapse or remain with residual deficits after treatment, the socio-economic burden is substantial. There are generally better functional outcomes and fewer relapses when intervention is early and aggressive. This case raises awareness of a rare presentation of AIE as respiratory failure. It also illustrates that more aggressive immunosuppression with cyclo-phosphamide on second presentation of AIE produced better outcomes with no relapse after 2 years, whereas the less aggressive approach on initial presentation allowed severe relapse in just 4 months. More research needs to be conducted to elucidate rare presentations of AIE and identify tailored therapies.

Figure 1 MRI showing signal indicative of inflammation in thalamus (A) and cervical spinal cord (B). Figure 2 A) H&E shows several intra and extracellular basophilic, round/oval structures. B) Brain parenchyma infiltrated by CD68(+) macrophages and C) CD3(+) T-cells. D) Van Kossa stain for calcium highlights the round/oval structures. Iron stain, to a lesser extent, and GMS and PAS (not shown) were also positive.
Disclosures: The authors of this poster have no financial or other conflicts of interest to disclose.

References


Disclosures: The authors of this poster have no financial or other conflicts of interest to disclose.
**Adult IgA vasculitis (Henoch–Schönlein purpura) with End-Stage Renal Disease**

Raymond Shih, MD, Manisha Naik, DO  
Department of Internal Medicine,  
St. Mary Medical Center,  
Langhorne, PA

IgA vasculitis, also known as Henoch-Schönlein purpura (HSP), is a systemic vasculitis most commonly seen in pediatric populations. However, ten percent of cases are in adults. Patients with HSP may present with palpable purpura, abdominal pain, arthritis, and/or renal involvement. In adults, renal disease may be severe, including end-stage renal disease.

A 65-year-old male observed bilateral pedal petechial/purpuric rash, which diffusely extended to his thighs and arms, including his palms and soles. Within days, he also noticed decreased urine outputs and cola-colored urine. No complaints of abdominal or joint pain were noted. With the suspicion of vasculitis, he was started on Prednisone 60mg daily. Subsequently, skin biopsy of the rash revealed leukoclastic vasculitis. Outpatient urinalysis revealed hematuria and 2+ proteinuria. His blood work showed a creatinine of 3 mg/dL and hyperkalemia. Creatinine, over the next several days rose to 5.4 mg/dL. He was, then, admitted to the hospital for pulse dose corticosteroids and management of acute kidney injury. The patient received hemodialysis. A kidney biopsy was performed and revealed “mild proliferative glomerulonephritis with IgA predominant deposits.” Combined with petechial/purpuric rash on presentation, the diagnosis of IgA vasculitis was reached. Hyperkalemia was resolved with dialysis, however creatinine level remained to be elevated.

This case demonstrates that HSP may be seen in adults. It can result in end-stage renal disease.

Clinicians should be mindful when assessing patients with petechial/purpuric rash to not overlook HSP solely based on demographic alone and assess kidney function promptly.

**STATEMENT OF DISCLOSURE**

Drs. Raymond Shih, MD, and Manisha Naik, DO have no conflicts of interest or financial ties to disclose.
Adult-onset Still’s disease with Macrophage Activation Syndrome

Raymond Shih, MD, Manisha Naik, DO

Department of Internal Medicine
St. Mary Medical Center
Langhorne, PA

Adult-onset Still’s disease (ASD) is an inflammatory disorder characterized by arthritis, daily fevers, and evanescent rash. Age of onset is over the age of 16. ASD can be associated with the Macrophage activation syndrome (MAS), which can be a fatal syndrome of excessive immune activation. It is Hemophagocytic lymphohistiocytosis in the setting of rheumatic disease and should be treated as a life-threatening emergency.

A 52-year-old, who had a history of inflammatory arthritis being treated with methotrexate and prednisone, presented to ED for a six-month history of nightly fevers of 38.3 degrees Celsius, chills, night sweats, and general malaise. He progressively worsened with pleuritic chest pain associated with dry cough and shortness of breath. He was admitted and initiated on antibiotic therapy for suspected sepsis secondary to pneumonia. During the admission, the patient developed new hoarse voice and pain in his toes and knees. Infectious work up, including TB, HIV, and Malaria were negative. Serum ferritin was found to be 18,581ng/mL. His fevers waxed and waned. They continued at the time of discharge. Arthralgias in his toes had improved but never resolved.

He returned to the ER five months later with progressively worsening night sweats, chills, weight loss, body aches, lymphadenopathy, and weakness. He developed fever with rigor. His blood work revealed Hemoglobin of 5.4, creatinine of 1.7, lactic acid level of 2.6, as well as significant leukocytosis of >68K, mainly neutrophils and bands. LFTS His CXR showed bilateral opacities. He, subsequently, developed respiratory failure, requiring intubation and was transferred to ICU. It was observed that he had waxing and waning of salmon-colored rash that coincides with occurrence of fever. In the setting of arthralgia, spiking fevers, and evanescent rash, a diagnosis of Adult onset Still’s disease was made. Peripheral smear, then, showed phagocytosis of neutrophils. Repeated serum ferritin was >40,000ng/mL. He was then diagnosed with macrophage activation syndrome. Due to being critically ill, he was transferred to a tertiary care center for further care. There, he was found to have significant EBV viremia via PCR. Unfortunately, the patient expired two days after transfer.

This case illustrates the difficulty in diagnosing MAS due to its infrequency. The length of time to diagnosis is often protracted secondary to lack of physician awareness. Moreover, the nature of adult onset still’s disease is primarily a diagnosis of exclusion. Improved recognition of the combination of symptoms can help to minimize morbidity and mortality in patients with this disease.

STATEMENT OF DISCLOSURE

Drs. Raymond Shih, MD, and Manisha Naik, DO have no conflicts of interest or financial ties to disclose.
Apixaban Induced Leukocytoclastic Vasculitis

Jenna Spears MB BCh BAO, Yousif Al-Saiegh MD

Jenna Spears MB BCh BAO, Department of Medicine, Pennsylvania Hospital, University of Pennsylvania Health System (UPHS), Philadelphia, PA

Yousif Al-Saiegh MD, Department of Medicine, Pennsylvania Hospital, University of Pennsylvania Health System (UPHS), Philadelphia, PA

Classification:
Jenna Spears: Internal Medicine Resident

Funding
No funding to report

Disclosures
Jenna Spears: None
Yousif Al-Saiegh: None

Background: Apixaban is a rare cause of leukocytoclastic vasculitis within the first six months of initiation. Leukocytoclastic vasculitis is the most common form of vasculitis in the skin, and is the result of deposition of immune complexes at the vessel wall. Almost 30% of all cases of leukocytoclastic vasculitis have been found to be drug induced, however there are only several cases of cases due to Apixaban in the literature.

Case: 95 year old male with history of congestive heart failure, prior cerebrovascular accident and non-valvular atrial fibrillation presents with a one day history of a diffuse palpable purpura of his lower extremities (Figure 1). The rash was mildly tender, but not pruritic. The patient had recently been transitioned from Warfarin to Apixaban twelve days prior, due to the burden of associated monitoring on the former. Differential of this new purpuric rash included infectious, hematological or autoimmune causes. Apixaban was discontinued on admission, as it was hypothesized to be a contributor to the vasculitis. During the work-up significant lab values included: Erythrocyte Sedimentation Rate (ESR): 33, C3: 23, C4: <8. Inpatient infectious and autoimmune laboratory tests were otherwise negative. Biopsy of the lesion showed extensive purpura with superficial perivascular neutrophilic infiltrate and leukocytoclasia. Given the concern that an inflammatory process was contributing, the patient was
initiated on oral Prednisone. The purpuric rash rapidly improved with withdrawal of Apixaban and a course of Prednisone. At time of discharge he was placed back on Warfarin.

Summary: Apixaban is a rare but important cause of leukocytoclastic vasculitis. In cases isolated to the skin, treatment is mostly supportive, and consists of withdrawal of the offending medication. The patient should be transitioned to a different direct oral anticoagulant or warfarin in the absence of contraindications. However, it should be noted that other direct oral anticoagulants are more commonly associated with leukocytoclastic vasculitis than Apixaban. In more complicated cases, involving skin necrosis or severe systemic vasculitis there is a role for immunosuppressants and steroids.

Figure 1:
Non-Bacterial Thrombotic Endocarditis in a Patient with Primary Antiphospholipid Syndrome

Jenna Spears MB BCh BAO, Anand Gopal MD, Saloni A. Shah BS, Yousif Al-Saiegh MD

Jenna Spears MB BCh BAO, Department of Medicine, Pennsylvania Hospital, University of Pennsylvania Health System (UPHS), Philadelphia, PA

Anand Gopal M.D., Wills Eye Institute, Thomas Jefferson University Hospital, Philadelphia, PA

Saloni A. Shah B.S. Sidney Kimmel Medical College at Thomas Jefferson University, Philadelphia, PA

Yousif Al-Saiegh M.D., Department of Medicine, Pennsylvania Hospital, University of Pennsylvania Health System (UPHS), Philadelphia, PA

Classification:
Jenna Spears: Internal Medicine Resident

Funding
No funding to report

Disclosures
None

Background: Non-Bacterial Thrombotic Endocarditis is often clinically silent with autopsy series showing an estimated prevalence of 0.9-1.6%. It is most often seen in patients with advanced malignancy, but can also be seen in autoimmune disorders such as systemic lupus erythematosus (SLE), antiphospholipid syndrome (APS) and rheumatoid arthritis (RA). It is often a diagnosis of exclusion, that requires a high degree of clinical suspicion and an extensive workup to diagnose.

Case: 61 year-old male with past history of hypertension and heart failure with preserved ejection fraction, antiphospholipid syndrome, complicated by prior pulmonary embolism (30 years prior), and cerebellar stroke (one year prior), on chronic Rivaroxaban and Aspirin. He presented with a ten day history of acute dyspnea, shortness of breath, dry cough and anorexia. Approximately one week later, he developed an acutely painful, erythematous and progressively edematous right hand. The progressive swelling resulted in loss of function and he became unable to flex or extend his right digits. Significant lab values were: Lactate: 3.5, CRP: 10.6, proBNP: 11432, WBC: 11300. Coagulation levels were normal. Inflammatory
work-up was significant for Beta-2-Glycoprotein IgG>100 and IgM>100, Anticardiolipin IgG>100, and positive Lupus Anticoagulant. Chest X-Ray showed a large right pleural effusion with associated atelectasis, and bilateral interstitial prominence suggestive of pulmonary edema. Right Hand X-Ray showed marked diffuse soft tissue swelling without soft tissue gas. Right Upper Extremity Doppler showed no evidence of venous thrombosis. Skin biopsy resulted as extensive neutrophilic inflammation with fibrinous exudate and thrombosed small vessels extending deep into the dermis. Patient was started empirically on broad-spectrum antibiotics for possible cellulitis. Transthoracic Echo (TTE) was done to evaluate for a potential source of septic emboli while cultures were pending. TTE showed normal ejection fraction, severe mitral regurgitation, severely dilated left atrium. A large, irregular, mobile echodensity was seen on the posterior mitral valve leaflets measuring two cm in diameter. Patient’s condition was self-limited. He was bridged with heparin to warfarin. Blood, tissue and pleural fluid cultures eventually returned negative.

Summary: Non-Bacterial Thrombotic Endocarditis (NBTE) is a rare cause of acute onset heart failure, but is an important differential, as a diagnosis of exclusion. Patients should undergo workup for hypercoagulable states, autoimmune conditions and infectious causes. This patient presented with left heart failure and embolic sequela, however his recent stroke on Rivaroxaban may also have been related to underlying NBTE, not detected on previous TTE. TTE can fail to detect NBTE as the vegetations are often small, easily friable and frequently embolize, leaving only small remnants. APS is managed with lifelong anticoagulation. Direct oral anticoagulants are less effective than warfarin in preventing recurrent thrombosis in individuals with APS, especially with a history of arterial thrombosis.
**The Efficacy of Low Dose Prednisone for Remission Induction in Newly Diagnosed Rheumatoid Arthritis Patients**

**Jacob Greenmyer, MSIV and Jack Stacy, MSIV (Co-principal investigators)**  
**James Beal, Ph.D and Abe Sahmoun, Ph.D.**  
**Erdal Diri, M.D.**

**Background/Purpose:**

Glucocorticoids (GCs) are commonly used in RA patients as remission induction monotherapy or as bridging therapy when starting DMARD/biologic therapy. Despite the ubiquity of GC use in RA treatment, agreed upon standard dosages, particularly for remission induction in newly diagnosed patients, remain elusive. Past studies indicated that the well-known long term adverse effects of GCs are directly related to cumulative dose. Therefore, identification of the lowest GC dose that reproducibly induces remission is imperative. The purpose of our study was to assess the response of newly diagnosed RA patients to low dose prednisone monotherapy (defined as less than or equal to 10 mg/day).

**Methods:**

We conducted a chart review for new diagnoses of RA (ICD-9 714) from January 1, 2005 to September 1, 2018 at Trinity Health Group in Minot, ND. Patients treated with \(<10\) mg prednisone daily for at least six weeks were included. Those previously treated with a DMARD or already started on a GC upon referral were excluded. Disease severity was calculated using the Disease Activity Score (DAS28-ESR). Response to treatment was determined based on the change in the DAS28-ESR score before and after treatment. The European League Against Rheumatism (EULAR) response criteria was used to categorize response to therapy as good, moderate, or no response.

**Results:**

A total of 1386 patients were screened and 201 of them met inclusion and exclusion criteria. The average dose of prednisone was 8 mg daily, ranging between 5 and 10 mg, for an average of 42.2 days. Average age at presentation was 55.1. Majority of them were female (65.7%) and white (91.5%). The average DAS28-ESR score among our entire cohort dropped from 5.1 ± 1.1 at presentation to 2.7 ± 1.3 after 6 weeks of treatment with low dose prednisone (\(p<0.001\)). The average DAS28-ESR of our seropositive patients (n=134) dropped from 5.2 ± 1.1 to 2.7 ± 1.3 (\(p<0.001\)), and that of our seronegative patients (n=67) dropped from 4.9 ± 1.2 to 2.6 ± 1.1 (\(p<0.001\)). As defined by the EULAR response criteria, 69.7% of patients showed a good response to treatment, 20.4% showed a moderate response, and only 10% showed no response. At presentation, 50.2% of the total cohort qualified as having either severe disease according to DAS28-ESR score. After treatment, only 5% qualified for severe disease, and 54.2% had reached remission.

**Conclusion:**

Low dose prednisone monotherapy leads to statistically significant improvement in clinical severity of RA in newly diagnosed patients.

**Disclosures:** No conflicts of interest to disclose.
Osteoporosis screening in African American patients with rheumatoid arthritis. Are we doing enough?

Presenting Author: A. Umar, M.D.

Introduction

Osteoporosis is a well-known extra-articular complication in rheumatoid arthritis (RA) patients. It is more common in patients with RA (50%) than in the general population, due to active systemic inflammation, the use of corticosteroids, and lack of mobility. The International Society for Clinical Densitometry (ISCD) and National Osteoporosis Foundation (NOF) has recommended dual-energy x-ray absorptiometry (DEXA) testing for all adult RA patients due to their high-risk status. However, a significant percentage of RA patients do not undergo DEXA scan despite these recommendations. Our aim was to assess osteoporosis screening rates in African American patients with RA.

Method:

Patients with a diagnosis of RA who visited a primary care clinic of Grady Memorial Hospital between July 1, 2017, and June 30th, 2018 were included (n=132). Data was extracted from the electronic medical record (EMR) system. We obtained data regarding the diagnosis of low bone marrow density in terms of osteoporosis and osteopenia through DEXA scan of the hip and lumbar spine. Co-morbidities included in the analyses were congestive heart failure, chronic kidney disease, HIV, hypertension, diabetes mellitus, vitamin D deficiency, hyperthyroidism, hyperparathyroidism, and immunosuppressive medication use (steroids, methotrexate, leflunomide, azathioprine, hydroxychloroquine, adalimumab and any other disease modifying anti-rheumatic drugs). STATA software was used and two-sided P-value < 0.05 was considered statistically significant.

Results

Out of 132 patients (74% females, 98% African American, median age 55), only 60 patients had a DEXA scan on file. Of these 60 patients, 50% had low bone marrow density. Osteoporosis and osteopenia prevalence were 40% (24/60) and 10% (n= 6/60) respectively. 43% (N =57 people) were on steroids at the time of data collection and only 30% had a DEXA scan on file. Out of 24 patients, who had osteoporosis, 50% (n=12/24) were on bisphosphonates and 33% of them had repeat DEXA scan in 2 years. The patients who were on steroids were more likely to have screening done as compared to patients who were not on steroids. (OR=2.29 CI 1.1-4., p=0.0234). The patients' age 50-60 were less likely to have DEXA scan on file compared to patients age > 60 (OR=0.29 CI 0.1-0.7 P = 0.01). There was no statistically significant difference in DEXA screening rates between patients with multiple comorbidities versus patients with RA only or patients with low vitamin D versus normal vitamin D.

Discussion:

Our study provides information regarding osteoporosis screening rates in our predominantly African American RA. Our study shows the suboptimal implementation of guidelines in our high-risk patient population which is consistent with previous studies. The results of this audit will make us more vigilant to identify those patients who need DEXA scanning to ensure that treatment is efficacious. Future goals are to set up a resident-driven intervention to not only educate providers about the increased risk of osteoporosis in RA patients but to also increase guideline compliance rates.

Disclosure: None
An uncommon cause of common presentation – IgG4 related disorder

Anusha Vuppala, MD, Kristen Sandoz, MD, Adijat Olanrewaju, MD, Juan Mercado, MD, Sarwat Umer, MD

Introduction

Neck swelling is commonly seen in pediatric clinical practice. The differential is broad which includes infectious (bacterial, viral), congenital, drugs, connective tissue disorders, malignancy. It is seen in all age groups ranging from infancy to adulthood. We present a case of a patient with neck swelling which was unresponsive to antibiotics and subsequently diagnosed as IgG4 related disorder on biopsy.

Case Presentation

Our Patient is a 3 yr old African American Female who presented with 5 day history of left sided neck swelling and subjective fever. It was preceded by clear rhinorrhea for a few days. Patient was started on amoxicillin by PCP with no improvement with presentation to our facility for further management. Patient was noted to have 4 x 4 cm non mobile, non erythematous, non indurated, non tender mass lateral to midline of trachea on left side. Labs notable for elevated sed rate of 46 (normal – 0 – 20), wbc – 8.53 (normal – 6 – 17). Neck ultrasound showed large heterogenous hypoechoic mass in left neck, posterior to left thyroid gland measuring 4.2 x 2.4 cm with no internal flow. CT neck with contrast showed 3 x 3 x 4 cm solid mass in left neck between thyroid lobe and great vessels with extension to retropharyngeal and parapharyngeal space. Patient was initiated on broad spectrum antibiotics with no improvement. Flow cytometry was done with no evidence of leukemia or non hodgkin lymphoma. Subsequently biopsy of the mass was done which showed acute and chronic inflammation, mixed inflammatory infiltrate with lymphocytes, plasma cells, macrophages, neutrophils, with obliteration of vessels and fibrosis. CD 138 highlights foci of plasma cells. IgG4 immunochemistry showed positive plasma cells. Serum IgG levels elevated at 1003 (453 - 916), IgG4 – 85 (1 – 78). Initiated steroids with dramatic improvement in the size of the lesion. (Biopsy pictures available on request).

Discussion

IgG4 related disorders are not commonly encountered in Pediatric practice. There are 25 reported cases of IgG4 thus far in pediatric population. Median age at presentation is around 13 years. Presentation of IgG4 at such an young age of 2 years is extremely rare. Patient’s biopsy result along with elevated IgG4 in serum pushed towards the diagnosis. Patient’s dramatic response to steroids confirmed our diagnosis. IgG4 related disorder should be considered in the differential of neck swelling in a patient who did not respond to antibiotics. Our Patient’s repeat neck ultrasound one month later showed significant improvement in the size of the mass.

Disclosures – There are no disclosures
Can’t walk, Can’t breathe – A Case report on Anti MDA 5 antibody Positive Dermatomyositis

Anusha Vuppala, MD, Swathi Chalasani, MD, Sarwat Umer, MD

Introduction

Weakness and rash in a pediatric patient can be the presenting sign of a rheumatological disorder. The symptoms can be non specific which leads to delay in seeking care. We present a unique case of atypical dermatomyositis with rapidly progressive Interstitial Lung disease in a 8 yr old patient. It is a very uncommon presentation in Pediatrics with complicated course. Management posed a challenge with delicate balance of antibiotics and intensive immunosuppressive therapy and subsequent good outcome for our patient.

Case Presentation

Patient is a 8 yr old African American Male presented with symptoms of weakness for about 5 months which progressed to a point where patient couldn’t walk, persistent fever upto 103 F almost every other day for about 4 months prior to presentation. It gradually progressed to joint pain, mainly involving knees, elbow with erythema and swelling. Mother noted rash around eyes and on hands about a month prior to presentation. Patient was also noted to have heliotrope rash and gottron papules, significant weakness of bilateral upper and lower extremities ( 3/5 ). MRI of lower extremity was done which showed extensive inflammation. So, treatment was initiated without muscle biopsy due to extent of muscle inflammation. Patient was started on pulse steroids and methotrexate subsequently with initial improvement of weakness. However, weakness and rash worsened. Planned for repeat pulse steroids and IVIG but patient was lost to follow up. About 3 weeks later, patient presented with respiratory distress. CXR revealed bilateral opacities consistent with ARDS. Physical examination notable for swollen right wrist and right knee concerning for septic arthritis. He was taken to OR emergently for washout to achieve source control. Cultures grew MSSA. Due to decline in respiratory status, patient was intubated and was promptly placed on ECMO due to inability to achieve adequate saturation on highest positive pressure. Decision was made to initiate on plasmapheresis as well with concern for rapidly progressive ILD. Anti MDA 5 antibody was tested at the same time to evaluate for Rapidly Progressive Interstitial Lung Disease. It was noted to be positive. He was initiated on rituximab soon after results of antibody testing. Patient’s respiratory status significantly improved on ECMO following plasmapheresis. He was able to be extubated and respiratory status remained stable on minimal supplemental oxygen. Repeat MDA 5 antibody titres declined. HRCT done 1 month later showed thickening of interlobular septa , numerous cystic appearing lesions, interstitial thickening opacities which are consistent with Interstitial Lung disease

Discussion

Juvenile dermatomyositis is a rare autoimmune vasculopathy characterized by proximal muscle weakness and pathognomic rash. Anti MDA 5 antibody is associated amyopathic or hypomyopathic dermatomyositis, ischemic cutaneous ulcerations, rapidly progressive ILD and poor prognosis. We describe a very sick, complicated pediatric patient presenting with Rapidly Progressive ILD and noted clinical improvement with timely starting of ECMO, plasmapheresis before rituximab. The presentation
is well described in adult population, whereas it is very rare in pediatric population. Rheumatologist needs to be aware of the anti MDA 5 antibody associated ILD in patients with dermatomyositis and the aggressive nature of disease course and intense immunosuppression needed for favourable prognosis. Similar cases were described in literature which had poor outcomes. Timely recognition and management made a significant impact on outcome for our patient.

Disclosures – There are no disclosures.
Statin-Induced Autoimmune Myopathy: A Clinical Case Report
Shahrzad Zonoozi, Michelle Rumbaugh, Tanuja Yalamarti, David Chetrit, David Chen

1. University of Pennsylvania Health System, Pennsylvania Hospital, Philadelphia, Pennsylvania
2. Division of Rheumatology, University of Pennsylvania

1. Introduction
Since their introduction in 1986, statins have become widely prescribed in the management of cardiovascular disease. Though generally safe and well-tolerated, a spectrum of statin-induced myopathies are becoming well-recognized. We report the case of a patient who presented with progressive muscle weakness while on atorvastatin.

2. Case Report
A 71 year old Chinese male with a history of stroke, diabetes mellitus, hypertension and hyperlipidemia who was in his usual state of health until returning from a trip to South East China. Upon his return to the US, he noted new fatigue, lower extremity weakness and shortness of breath. During initial hospitalization, he was noted to have an elevated creatine kinase (CK) and transaminitis. Muscle biopsy showed scattered myonecrosis with mild inflammation and nuclear bags. Given concern for statin-induced myopathy, atorvastatin was discontinued. He subsequently re-presented to hospital with worsening shortness of breath.

Physical exam revealed symmetrical proximal weakness of upper and lower extremities with 2/5 strength in deltoids and hip flexors. Repeat labs showed a persistently elevated CK to 2766 and ongoing transaminitis with aspartate aminotransferase (AST) 227 U/L, alanine aminotransferase (ALT) 227 U/L, normal alkaline phosphatase and bilirubin. Myositis work-up revealed a rheumatoid factor of 13, ANA and ANCA negative, dsDNA antibody < 10, aldolase 24-42 during hospitalization prior to treatment. Anti-HMGCR antibody was strongly positive and increased from 54 to 126 units on consecutive testing. Extended myositis panel was negative.

3. Discussion
Based on the above findings, the patient was diagnosed with autoimmune necrotizing myositis secondary to atorvastatin use. He was started on intravenous methylprednisolone 1g for 3 days and subsequently continued on prednisone 60mg daily. He received a 5 day course of intravenous immunoglobulin G (IVIg), and was started on methotrexate as a steroid sparing agent. Upon re-evaluation, laboratory tests normalized and the patient made clinical improvement.

4. Conclusion
The pathogenesis of statin-induced autoimmune necrotizing myositis remains unclear and there appears to be an association with human leukocyte antigen (HLA) DRB1*11:01. Statin exposure upregulates HMGCR and in genetically susceptible individuals, may result in autoimmunity against HMGCR. Once an autoimmune response is activated, high HMGCR levels in regenerating muscle cells may continue to drive autoimmunity even after statin is discontinued. Whilst no clinical trials have been conducted to establish effective treatments for statin-induced autoimmune myopathy, the mainstay of treatment is discontinuation of the medication and immunosuppression with high dose glucocorticoids as well as maintenance therapy including other agents such as IVIg, methotrexate, azathioprine, mycophenolate mofetil or rituximab.
5. Clinical Implications
Given the increasing and widespread use of statins, it is important to be aware of the less common side effects of these medications, their diagnosis and available treatments. This is especially important as discontinuation of statins is not necessarily sufficient to stop the disease process.

Disclosures – None

References