Introduction

- Lemierre syndrome, or jugular vein suppurative thrombophlebitis, was common before the discovery of antibiotics. (1)
- The incidence of Lemierre syndrome decreased after use of penicillin and was nearly forgotten for 30 years, hence, also referred to as the forgotten disease. (1)
- Although still rare overall, the incidence is now starting to rise. (2)

Case Description

- A healthy 36-year-old woman is evaluated at urgent care for acute pharyngitis and dyspnea then sent home with supportive treatment.
- Symptoms progress over a week to include right neck pain, fever and chills.
- Presented to ED with respiratory distress, found to be febrile, hypoxic with leukocytosis and thrombocytopenia.
- CXR notable for RLL airspace disease, CT neck w/ non-occlusive in R internal jugular vein (Figure A).
- Admitted to MICU for hypoxic respiratory failure requiring intubation, broad spectrum antibiotics and anticoagulation started.
- Blood cultures grew Fusobacterium necrophorum.

- Developed pleural effusions requiring bilateral chest tubes (Figure C).
- Fevers persisted despite clinical improvement, CT C/A/P negative for occult abscess.
- Discharged to rehab after antibiotics course. Represented 3 weeks later with severe lower back pain found to have L4-S1 osteomyelitis w/ epidural phlegmon on MRI (Figure D).
- After extended antibiotic course she recovers and returns to work.

Imaging

Figure A: CT Neck showing non occlusive Right IJ Thrombus. Figure B: Graphic demonstrating pathogenesis of bacteria induced thrombus formation. (4)

Lemierre Syndrome Illness Script

Presentation
- Young healthy adolescents
- High fever, Chills, Sore throat, neck pain
- Respiratory distress

Diagnostics
- Internal jugular Vein thrombophlebitis on CT Neck w/ Contrast
- Anaerobic bacteremia (e.g. Fusobacterium spp)
- Septic emboli

Management
- Empiric Antibiotics
- Source control
- Anticoagulation (Debatable)

Discussion

- Lemierre Syndrome (also termed The Forgotten Disease), is defined as post-pharyngitic suppurative jugular vein thrombophlebitis, with metastatic spread of disease (bone, lung, brain, joints)(Figure B).
- Up to 80% of cases are caused by the gram negative anaerobic bacteria Fusobacterium necrophorum, with the remainder caused by a variety of organisms. (5)
- Fusobacterium spp. are common microbiologic isolates in non-streptococcal pharyngitis and peritonsillar abscesses, accounting for about 20% of community onset pharyngitis, (Table 2), and result in more morbidity & mortality (up to 5%) than streptococcal pharyngitis(5).

<table>
<thead>
<tr>
<th>Species</th>
<th>Patients (n = 312)</th>
<th>Asymptomatic Students (n = 180)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fusobacterium necrophorum</td>
<td>64 (20.5)</td>
<td>17 (9.4)</td>
</tr>
<tr>
<td>F. necrophorum and GAS</td>
<td>47 (15.1)</td>
<td>16 (9.9)</td>
</tr>
<tr>
<td>F. necrophorum and GCS/GGS</td>
<td>9 (2.9)</td>
<td>6 (3.3)</td>
</tr>
<tr>
<td>F. necrophorum, GAS, and Mycoplasma pneumoniae</td>
<td>7 (2.2)</td>
<td>1 (0.6)</td>
</tr>
<tr>
<td>Fusobacterium polymorphus</td>
<td>1 (0.3)</td>
<td>-</td>
</tr>
</tbody>
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- Lemierre syndrome should be suspected in young healthy adults with preceding pharyngitis that rapidly progresses to fever, rigors, chills, neck pain and respiratory distress.
- Additionally counseling in ‘return precautions’ should focus on these symptoms.
- The incidence of this “forgotten disease” may continue to rise in the era of increasing antimicrobial resistance and stewardship. (1)(2)

Teaching Points

- In the era of increasing antibiotic stewardship clinicians should be vigilant about the potential of developing Lemierre syndrome.
- Remember The Forgotten Disease, a morbid & mortal complication of pharyngitis in young healthy individuals with rapidly progressive systemic syndromes preceded by upper respiratory symptoms.

References