Clinical Characteristics and Outcomes of Patients with Otosyphilis and HIV Infection

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Background

Cochleovestibular dysfunction secondary to infection with Treponema pallidum (otosyphilis) is a reversible cause of sudden hearing loss, found more commonly in HIV infected patients, however, there is not an established criteria for diagnosis. It is hypothesized that infection of treponemal spirochetes into the central nervous system occurs early in the course of disease, perhaps occurring in as high as 100% of infected patients. The clearance of the organism from the CSF is multifactorial, including factors such as host immune status, presentation to health care provider, and duration of infection. Otosyphilis is thought to potentially occur at any stage of syphilis, however may proceed through two separate processes/mechanisms: (1) through 8th cranial nerve involvement in syphilitic meningitis or (2) through direct invasion of the spirochetes into perilymph of the inner ear. The mechanism of disease may influence the positivity of CSF studies, confounding proposed methods of diagnosis. Intuitively, inflammation of the cranial nerves in syphilitic meningitis will produce less isolated symptoms than localized invasion of auditory structures. In this case series of six HIV positive patients with otosyphilis, the differences in presentation, treatment, and course of disease will be discussed in an attempt to illuminate the variability found in the otosyphilis syndrome.

Methods

We performed a retrospective chart review from 2007 through 2016 from a tertiary academic center in Washington, DC and identified six patients who met the following inclusion criteria: HIV infection, cochleovestibular complaints, positive RPRs drawn within 6 months of presentation to the Otolaryngologist, performance of an LP and treatment for presumed otosyphilis. We excluded patients with a history of noise exposure, recurrent otologic infections, prior exposure to ototoxic medication, known CNS lesions prior to onset of symptoms, poor documentation of prior work-up. All patients were followed by both Infectious Disease physicians and Otolaryngologist. Data on medical history, clinical presentation, laboratory/imaging findings, audiograms, treatment courses, and outcomes were collected.

Results

Table 1: Selected clinical characteristics of the 6 patients studied and their subjective outcomes after treatment

<table>
<thead>
<tr>
<th>Patient</th>
<th>Serum RPRs within 6 months of diagnosis (pre-treatment)</th>
<th>CSF WBC count</th>
<th>CSF Protein</th>
<th>CSF VDRL</th>
<th>Brain MRI/CT Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1:128</td>
<td>25</td>
<td>40</td>
<td>Negative</td>
<td>7th and 8th CN enhancement and otolesly bilaterally</td>
</tr>
<tr>
<td>2</td>
<td>1:128</td>
<td>30</td>
<td>40</td>
<td>Negative</td>
<td>No significant findings</td>
</tr>
<tr>
<td>3</td>
<td>1:16</td>
<td>28</td>
<td>12</td>
<td>Negative</td>
<td>Increased FLAIR MR signal in 6 Visible</td>
</tr>
<tr>
<td>4</td>
<td>1:16</td>
<td>28</td>
<td>12</td>
<td>Negative</td>
<td>Increased FLAIR MR signal in 6 Visible</td>
</tr>
<tr>
<td>5</td>
<td>1:128</td>
<td>25</td>
<td>15</td>
<td>Negative</td>
<td>No significant findings</td>
</tr>
</tbody>
</table>

Figure 2: A graph of RPR titers responses to treatment for each patient studied, labeled with each graph is the treatment each patient received. The background of the graph corresponds with response to treatment: green being resolution of symptoms, gray being intermittent response, and red being no improvement/progression of symptoms.

• 5/6 patients presented with subjective hearing loss, 1 patient presented with ataxia from vestibular dysfunction.
• 4/6 patients had audiograms performed.
• Audiograms revealed low through high frequency hearing loss of the affected side in 2 patients and high frequency hearing loss in 1 patient (>1.5 to 2 kHz). One patient had no objective hearing loss on audiogram to match his symptoms.
• 3/4 had bilateral high frequency hearing loss, even though their symptoms were localized to only one ear.
• 4/4 patients had audiograms that correlated with their symptoms.
• 5/6 patients initiated treatment only after 4 weeks of symptoms
• Treatment consisted of 14 days of Penicillin G IV (1 patient received ceftriaxone)
• 3/6 patients had symptom improvement/resolution in 6 months
• 2/6 had no improvement/worsening of symptoms
• 1/6 had temporary improvement of symptoms
• 5/6 patients received steroids along with IV penicillin
• 2/6 had no improvement/progression of symptoms
• 1/6 had temporary improvement of symptoms
• 2/6 had symptom improvement/resolution in 6 months
• 2/6 patients did not have improvement
• 1 patient was later diagnosed with CNS lymphoma
• 1 patient had no objective findings on audiogram (patient 2)
• Response to treatment did not seem to correlate with HIV viral load or CD4 count

Conclusions

• The presentation of otosyphilis is varied and requires a high index of suspicion, especially in the presence of HIV infection.
• Either high or low frequency hearing loss can occur and often occur simultaneously
• Treatment with IV penicillin and steroids can improve symptoms
• Need further studies to determine whether prompt diagnosis and prolonged therapy would improve outcome in those without symptom improvement.

References


Clinical characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Gender</th>
<th>CD4 Count</th>
<th>HIV Viral Load</th>
<th>Initial or Primary Symptoms</th>
<th>Lateral Subjected</th>
<th>Duration</th>
<th>Associated Symptoms</th>
<th>Audiology</th>
<th>Clinical Outcomes of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>44</td>
<td>M</td>
<td>466</td>
<td>20</td>
<td>Hearing loss</td>
<td>Unilateral</td>
<td>4 Months</td>
<td>Ear pain, tinnitus</td>
<td>Available</td>
<td>Temporary improvement of symptoms (resolved after 2 weeks)</td>
</tr>
<tr>
<td>2</td>
<td>51</td>
<td>M</td>
<td>482</td>
<td>20</td>
<td>Hearing loss</td>
<td>Bilateral</td>
<td>1 Month</td>
<td>Tinnitus</td>
<td>No</td>
<td>No improvement of symptoms</td>
</tr>
<tr>
<td>3</td>
<td>24</td>
<td>M</td>
<td>547</td>
<td>20</td>
<td>Hearing loss</td>
<td>Bilateral</td>
<td>1 Month</td>
<td>Tinnitus</td>
<td>Yes</td>
<td>Improvement/resolution of symptoms</td>
</tr>
<tr>
<td>4</td>
<td>48</td>
<td>M</td>
<td>17</td>
<td>28230</td>
<td>Hearing loss</td>
<td>Unilateral (Right ear)</td>
<td>1 Week</td>
<td>N/A</td>
<td>Yes</td>
<td>Improvement/resolution of symptoms</td>
</tr>
<tr>
<td>5</td>
<td>28</td>
<td>M</td>
<td>148</td>
<td>30</td>
<td>Hearing loss</td>
<td>Unilateral (Right ear)</td>
<td>1 Month</td>
<td>Ear pain, tinnitus, and headache</td>
<td>Yes</td>
<td>Progression of symptoms</td>
</tr>
<tr>
<td>6</td>
<td>23</td>
<td>M</td>
<td>62090</td>
<td>N/A</td>
<td>Ataxia</td>
<td>Not applicable</td>
<td>3 Months</td>
<td>N/A</td>
<td>No</td>
<td>Improvement/resolution of symptoms</td>
</tr>
</tbody>
</table>

Figure 1a-1d: Audiogram results for those patients who presented for testing depicted as hearing level in decibels versus frequency in hertz. The right ear is depicted in blue with the left ear depicted in red. The side of symptomology is labeled along with the patient number in the bottom right corner.