INTRODUCTION

Otosyphilis is a rare complication of Syphilis caused by the spirochete Treponema pallidum. It is transmitted through sexual contact or vertically from mother to fetus, and spread through cerebrospinal fluid (CSF) and the endolymphatic system of the inner ear.

Secondary, tertiary, and congenital Syphilis can manifest with the audiovestibular symptoms of Otosyphilis, such as sudden unilateral or bilateral sensorineural hearing loss (SNHL), tinnitus, and vertigo.

High risk populations include men who have sex with men (MSM) and those with HIV infection.

Rates of Syphilis in the U.S. have risen annually since 2006, with a rate increase of 178.6% among women during 2015-2019, suggesting a rapid increase in heterosexual Syphilis.

An increasing incidence over the past decade may indicate an underdiagnosis in the ear nose throat (ENT) community.

Diagnostic aids include audiograms, confirmatory serological testing of rapid plasma reagin (RPR) or Treponemal enzyme immunoassays, and CSF analysis.

CASE DESCRIPTION

48-year-old Hispanic male with a past medical history of HIV infection presented to the Emergency Department (ED) 3 months after sudden sensorineural hearing loss in the left ear with tinnitus, and vertigo.

He failed to show audiologic improvements after a high dose oral prednisone taper and two intratympanic dexamethasone injections with an ENT physician.

After a recent positive RPR titer, the patient was advised by his primary care physician in coordination with infectious disease specialists to come to the ED for workup of a suspected diagnosis of Otosyphilis.

HISTORY

Past Medical History:
- HIV positive with recent CD4 count within normal limits per primary care physician
- Recent Syphilis diagnosis
- Mandibular osteomyelitis
- Malignancy
- Tinea versicolor

Medications:
- Etravirine/tenofovir alafenamide-ribovirine 200/25-35 mg per oral daily
- Multivitamin per oral daily

Allergies:
- No drug allergies

Surgical History:
- Unremarkable

Family History:
- Unremarkable

Social History:
- History of unprotected sex with multiple men
- 10 pack-year history of smoking tobacco
- Review of Systems:
  - Tinnitus in the left ear
  - Intermittent flushing lights in his bilateral temporal visual field for 2 weeks
  - Remainder of the review of systems was unremarkable

PHYSICAL EXAM

Vitals:
- Blood Pressure: 116/73 mmHg
- Pulse: 70 beats per minute
- Temperature: 98.6 degrees Fahrenheit (oral)
- Respirations: 18 breaths per minute
- Oxygen Saturation: 100% on room air

Skin: no lesions or rashes noted

HEENT: Weber test lateralized to right ear, Rinne test showed air > bone conduction. Tympanic membranes showed no effusions, orrrhea, or rupture. Pupils were equal round and reactive to light and accommodation

Neck: Full range of motion of the neck woth no evidence of stiffness

Cardiovascular: regular rate and rhythm, no murmurs

Pulmonary: lungs sounds clear to auscultation in all lung fields bilaterally

Musculoskeletal: 5.5 strength in upper and lower extremities bilaterally

Neuro: CN II-VII intact. Alert and oriented to person, place, and time. Sensory and motor intact. Reflexes 2+ throughout. No limb or truncal ataxia noted

DIAGNOSTICS

Routine CBC and chemistries were within normal range
- ESR: 25 mg per oral daily
- Lumbar puncture was performed, and CSF analysis showed a mildly elevated white cell count (33% monocytes, 62% lymphocytes, 4% neutrophils) with 6 nucleated cells/μl. 92 red blood cells/μl. Protein 37.5, glucose 60
- CSF culture showed no organism growth, a nonreactive VDRL, and negative Rapid Strep Simplex Virus polymerase chain reaction
- Brain MRI with and without contrast was unremarkable and showed no acute intracranial abnormalities with unremarkable internal auditory canals (Figure 3)
- Brain MRA was unremarkable (Figure 3)

Recent audiogram showed profound SNHL from 250-8000 Hz in the left ear and normal hearing sensitivity in the right ear (Figure 1)

Tympanometry was unremarkable and suggested a normal middle ear function in both ears

Figure 1: Audiogram pre-antibiotic treatment

Figure 2: Audiogram post-antibiotic treatment

Figure 3: Normal brain MRI/MRA ruled out ototoxicity, meningitis, and vertebrobasilar cerebrovascular accident (CV A)

RECOMMENDED TREATMENT GUIDELINES FOROTOSYPHILIS

1. First line treatment includes prompt hospital admission for intramuscular penicillin G sodium 10 to 14 days.
2. Treatment delay can result in permanent hearing loss.
3. Adjunct corticosteroid therapy may be beneficial.
4. Close individualized follow-up with serial audiograms is recommended to evaluate response to treatment.

CONCLUSION

Early diagnosis and treatment is critical in potentially reversing SNHL due to Otosyphilis.

Given the rise of Syphilis in recent years, clinicians should consider Otosyphilis in any sexually active patient presenting with sudden SNHL, particularly MSM and those with HIV.

If suspected, early consult with ENT and infectious disease specialists should be implemented to prevent permanent hearing loss and further neurologic sequelae.

REFERENCES