



Otosyphilis: A Timely Diagnosis

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INTRODUCTION

- Otosyphilis is a rare complication of Syphilis caused by the spirochete *Treponema pallidum*¹
- 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 ear¹
- 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 vertigo¹⁻⁴
- High risk populations include men who have sex with men (MSM) and those with HIV infection^{1,2}
- Otosyphilis has an estimated prevalence of 653 per 100,000 among patients with otologic presentations⁴
- Rates of Syphilis in the U.S. have risen annually since 2000, with a rate increase of 178.6% among women during 2015-2019, suggesting a rapid increase in heterosexual Syphilis⁵
- An increasing incidence over the past decade may indicate an underdiagnosis in the ear nose and throat (ENT) community^{1,2}
- Diagnostic aids include audiograms, confirmatory serologic testing of rapid plasma reagin (RPR) or Treponemal enzyme immunoassays, and CSF analysis^{1,4}
- CSF analysis may show pleocytosis, elevated protein, and normal glucose with or without a positive venereal disease research laboratory test (VDRL)^{4,6}
- Audiometry curves often show low frequency involvement of the affected ear, suggestive of endolymphatic hydrops⁷
- Otosyphilis is one of the few forms of SNHL that are potentially reversible with early recognition and treatment^{4,8}
- Table 1 includes the differential diagnosis for Otosyphilis¹⁻⁶

Table 1: Differential diagnosis

- Acoustic neuroma
- Meniere's disease
- Otosyphilis
- Vertebrobasilar cerebrovascular accident (CVA) or transient ischemic attack (TIA)
- Autoimmune inner ear disease
- Meningitis
- Vestibular neuritis

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
- He had 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 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
- Medications:
 - Emtricitabine/tenofovir alafenamide-rilpivirine 200/25-25 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 flashing 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, erythema, or rupture. Pupils were equal round and reactive to light and accommodation
- Neck: Full range of motion of the neck with no evidence of stiffness
- Cardiovascular: regular rate and rhythm, no murmurs noted
- Pulmonary: lung sounds clear to auscultation in all lung fields bilaterally
- Musculoskeletal: 5/5 strength in upper and lower extremities bilaterally
- Neuro: CN II–XII 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
- Serologic RPR titer was reactive at 1:64 with a reactive *Treponema pallidum* IgG antibody index of 52, and a reactive total *Treponema pallidum* antibody (IgG and IgM)
- Lumbar puncture was performed, and CSF analysis showed a mildly elevated white cell count (33% monocytes, 62% lymphocytes, 4% neutrophils) with 6 nucleated cells/uL and 92 red blood cells/uL, protein 37.5, glucose 60
- CSF culture showed no organism growth, a nonreactive VDRL, and negative Herpes 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

PATIENT MANAGEMENT

Recommended Treatment Guidelines for Otosyphilis

- First line treatment includes prompt hospital admission for intravenous penicillin G sodium for 10 to 14 days⁸
- Treatment delay can result in permanent hearing loss^{3,8}
- Adjuvant corticosteroid therapy may be beneficial^{3,8}
- Close individualized follow-up with serial audiograms is recommended to evaluate response to treatment⁴

Case Outcome

- Patient was diagnosed with Otosyphilis and admitted for intravenous penicillin G sodium 3 million units every 4 hours for 10 days with neurology and infectious disease consultation
- He was continued on his antiretroviral therapy and given deep vein thrombosis prophylaxis during hospitalization with enoxaparin 40 mg subcutaneous injection daily
- With daily neurologic checks, patient's ocular symptoms had improved. No subjective improvement in hearing was noted
- He was discharged home with instructions to follow up with outpatient ENT, neurology, and ophthalmology specialists
- Patient's follow up audiogram with ENT one month later showed no improvements in hearing in the left ear (Figure 2)
- There is now discussion of invasive surgical procedures such as a bone-anchored hearing aid or cochlear implant

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

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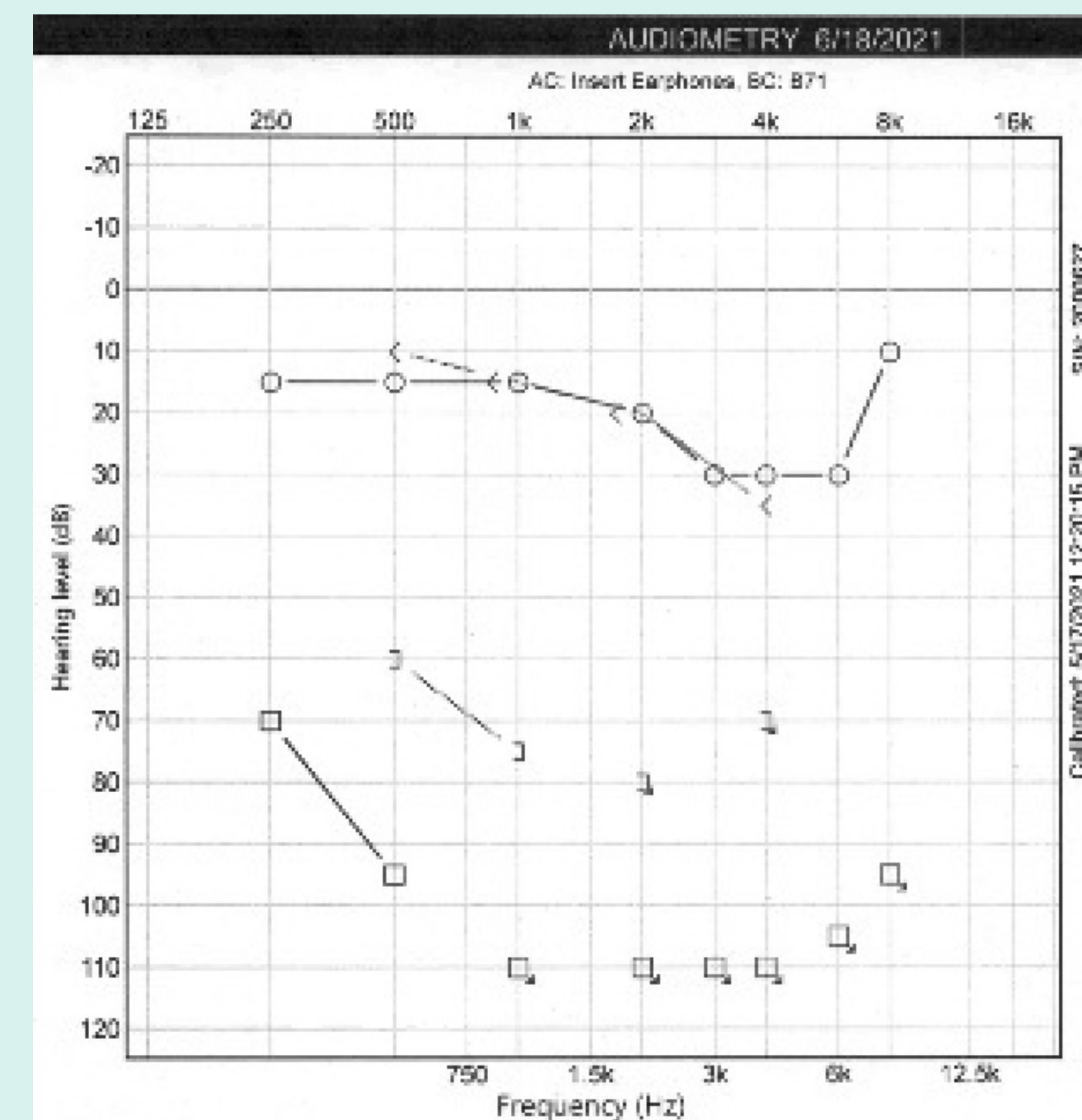


Figure 1: Audiogram pre-antibiotic treatment

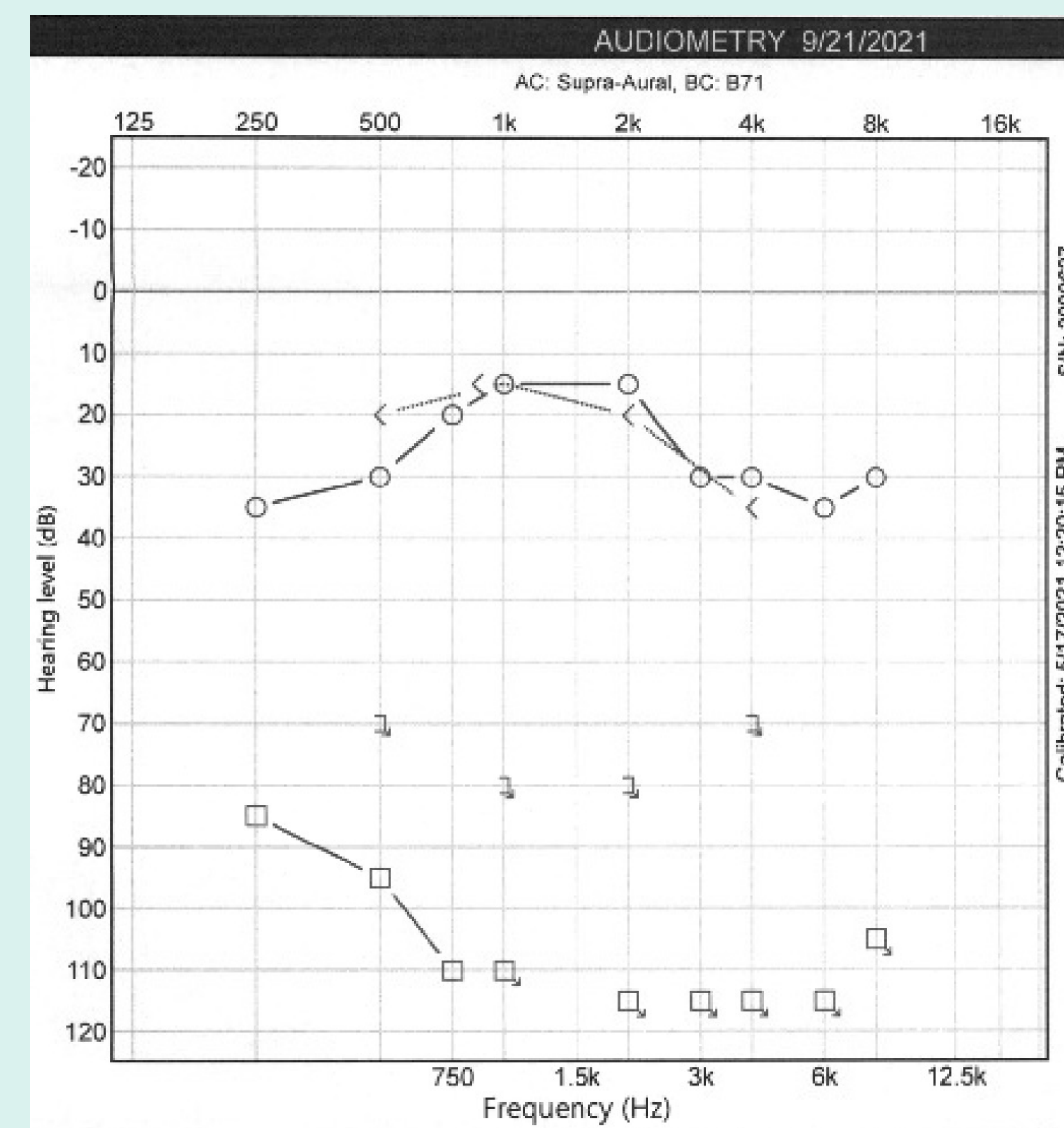


Figure 2: Audiogram post-antibiotic treatment

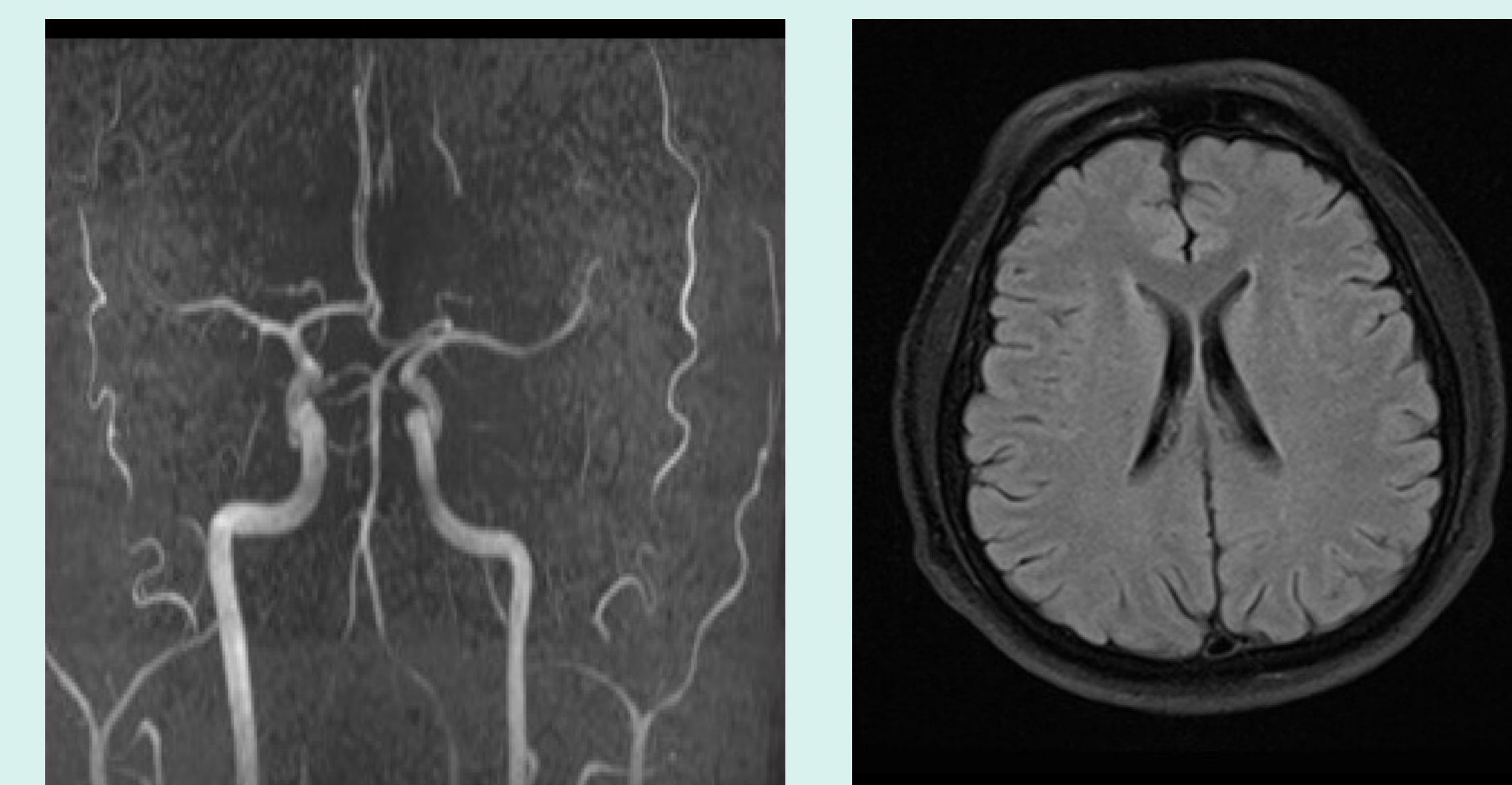


Figure 3: Normal brain MRI/MRA ruled out acoustic neuroma, neurosyphilis, or CVA