

## Case Report

## Otosyphilis a Case Report

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## Introduction

Syphilis is a Sexually Transmitted Infection (STI) or congenitally acquired disease caused by the *Treponema pallidum* subspecies *pallidum*. The “great imitator”, syphilis can have varied clinical presentations including genital ulceration (painful or painless), rash, neurologic dysfunction (cerebral vascular accident, meningitis), and stillbirth [1]. Some of the most debilitating consequences of infection include neurological manifestations that can occur at any time, even years to decades after the initial infection. Prior studies report an increased risk of neurosyphilis in persons with HIV (PWH), especially in those with low CD4 counts and not on Antiretroviral Therapy (ART) [2,3].

Otosyphilis is a less common form of neurosyphilis whereby inflammation of the vestibulocochlear nerve, cochleovestibular apparatus, and/or temporal bone may cause Sensorineural Hearing Loss (SNHL) and/or vertigo [1]. Otosyphilis is often challenging to diagnose, because it can present without other symptoms classically associated with syphilis. The recommended treatment regimen for neurosyphilis in the United States is Intravenous (IV) penicillin G. In this study, we describe a case of a delayed diagnosis of otosyphilis with a concurrent new diagnosis of HIV that illustrates this important and often misdiagnosed disease.

## Case

A 30-year-old married woman and mother of three children, a homemaker, has been undergoing treatment for latent syphilis for the past 10 years and has been placed on intravenous Penicillin G. She came for a consultation due to bilateral progressive hearing loss over the last 10 years, accompanied by non-pulsatile bilateral tinnitus. There is no history of dizziness induced by changes in pressure or exposure to high-intensity sounds.

On otoscopy, the eardrums were intact and normal.

The vestibular examination was normal. During the neurological examination, no deficits were noted in the examination of the cranial nerves and muscles. The individual exhibited a normal gait, coordination, and balance.

A pure-tone audiometry was performed, revealing severe bilateral sensorineural hearing loss on the right and profound loss on the left (Figure 1).

Impedance audiometry showed a central tympanogram with a present stapedia reflex on both the right and left sides.

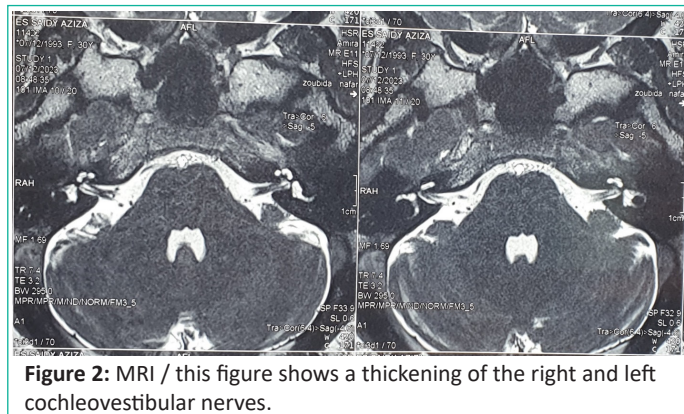
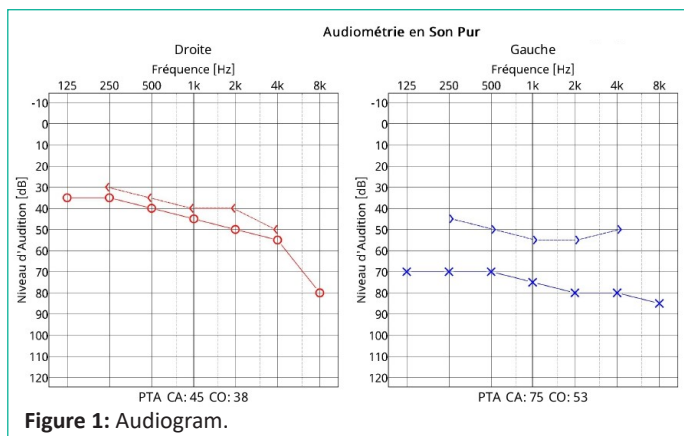
The blood syphilitic serology indicated a true positive result: TPHA (*Treponema Pallidum* Hemagglutination Assay) = positive at 1/2560; VDRL (*Venereal Disease Research Laboratory*) = positive at 1/2.

The lumbar puncture showed normal results, with syphilitic serology being negative in the cerebrospinal fluid (CSF). Ophthalmological evaluation revealed no ocular abnormalities.

The assessment did not reveal any co-infections, particularly no HIV infection.

The patient underwent a temporal bone CT scan, revealing bilateral mastoid opacification consistent with bilateral mastoiditis.

The Magnetic Resonance Imaging (MRI) showed thickening of the cochleo-vestibular nerve at the cisternal segment and its division branches at the internal auditory meatus, more pronounced on the right, without contrast enhancement after contrast agent injection. Given the context, this suggests non-active otosyphilis (Figure 2).



**Table 1: Multiple cranial involvements with MRI abnormalities (cranial nerves thickening and gadolinium enhancement).**

<b>Tumoral causes:</b>
- Carcinomatous meningitis
- Meningeal involvement in lymphomas
- Neurofibromatosis type 2
<b>Autoimmune causes:</b>
- Sarcoidosis
- Wegener's disease and other granulomatoses
- Connective tissue diseases
<b>Infectious causes:</b>
- Viral
- Tuberculosis
- Syphilis
- Lyme disease
<b>Traumatic causes</b>

**Discussion**

Otosyphilis is a rare manifestation of systemic *Treponema pallidum* bacterial infection, with the most common clinical symptoms including sensorineural hearing loss, tinnitus, and vertigo [4]. Bilateral hearing loss typically occurs acutely with rapid progression, accompanied by vestibulocochlear symptoms [5]. The presentation can also vary between individuals, spanning unilateral and bilateral involvement, asymmetric hearing loss, rapidly progressive to fluctuating hearing loss, spontaneous or provoked disequilibrium, and generalized unsteadiness [6,7]. Audiometric studies often involve the low- and high-frequency ranges [5,8] while sparing middle frequencies [9]. Otosyphilis and neurosyphilis can be diagnosed in all phases of syphilitic infections [4,10]. *Treponema pallidum* spirochetes can infect the central nervous system, causing hearing loss by syphilitic inflammation of the perilymphatic space, degeneration of the inner ear structures and temporal bone, and impairment of the eighth cranial nerve [4,9,11-14]. Thus, all patients with a confirmed diagnosis of disseminated syphilis should undergo a thorough ocular, otologic, and neurologic examination [13].

The diagnosis of otosyphilis is inherently challenging as it requires a high index of clinical suspicion even in the context of confirmatory serology. There does not exist a practical way to perform a treponemal test on the perilymphatic fluid nor a histologic examination of the temporal bone [4,12,16]. Diagnosis is multistep, typically requiring a combination of compatible neurologic, cochlear, and vestibular symptoms, a positive serologic test, CSF abnormalities, and/or audiologic studies [4,6,13]. Otosyphilis is also a diagnosis of exclusion, requiring that hearing loss is not caused by another discernable organic inner ear pathology [4,6,11,15].

Cette patiente a présenté une atteinte subaiguë des nerfs cochléo-vestibulaire avec à IRM un épaississement et une prise de contraste pathologique de celui-ci correspondant probablement à une infiltration nerveuse inflammatoire lymphocytaire péri-vasculaire. De telles anomalies IRM sont aspécifiques et peuvent se rencontrer dans des pathologies très diverses (Table 1) comme une méningite carcinomateuse, une localisation méningée de lymphome, une neuro-sarcoïdose, une granulomatose de Wegener, une tuberculose neuromeningé, une neurosyphilis, une neuro- borreliose ou dans certaines infections virales (Smith et Anderson, 2000 ; Corr, Bhigjee & Lockhat, 2004).

Otosyphilis is typically treated as symptomatic neurosyphilis with intravenous penicillin regardless of CSF findings [4,6,9]. Intramuscular penicillin typically fails to obtain appropriate treponemicidal levels in the cerebrospinal and perilymphatic space [8,9,12,16]. In addition, corticosteroids are sometimes administered to improve the chances of remission of hearing loss by reducing inner ear inflammation [6,9]; however, corticosteroids are typically avoided in HIV-infected individuals to avoid further suppression of cell-mediated immunity [8]. Unfortunately, definitive conclusions on treatment outcomes are difficult given the low incidence of this rare disease [7].

The prompt diagnosis of otosyphilis is critical as early treatment can arrest the progression of audiologic disease [12,16]. Treatment may lead to improvement of hearing function and even complete restoration of hearing abilities [7,10,11] given that syphilis is one of the rare reversible causes of sensorineural hearing loss [12]. Meaningful recovery is more likely to happen with a shorter duration of symptoms [11]. Older patients with chronic, persistent vestibulocochlear and audiologic disturbances are less likely to recover [2,15]. Hearing loss is less likely to improve compared to vestibular symptoms [15], with the literature observing an 80% improvement in tinnitus and vertigo compared to 20%-25% of patients with hearing improvement after appropriate antibiotic therapy [8,12].

**Conclusion**

It is important for health care professionals realize that syphilis is still with us and the incidence of otosyphilis in adults may increase in coming years as we see syphilis rates continue to climb upwards. Otosyphilis is one of the few forms of SNHL that can improve with antibiotic treatment, and it is potentially curable. Therefore, routine screening for, such as asking patients about hearing loss, tinnitus or gait imbalance, should be integrated into the work up for a new diagnosis of syphilis regardless of syphilis.

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