Otosyphilis: A Silent Presentation of Syphilis

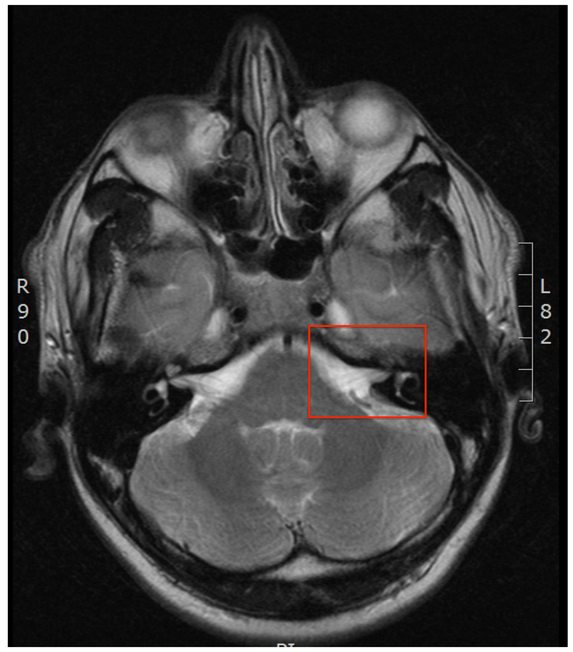
Christie Izutsu, MD, Resident – Department of Internal Medicine
Ryan Orosco, MD, Resident – Division of Otolaryngology, Head & Neck Surgery
Joseph Caperna MD, MPH; Eva Stettner, RNP

UC San Diego School of Medicine

Background

The rate of syphilis diagnoses has increased recently. Less common clinical manifestations of this disease have been seen with increasing frequency, otosyphilis being one such manifestation. Several theories have been proposed for the pathogenesis of otosyphilis, including that a hypersensitivity reaction plays a role in the hearing loss and that immunosuppressed states such as with HIV can result in reactivation of dormant treponemes secondary to T-cell involvement.[1,2] Vestibulocochlear symptoms, including sensorineural hearing loss, remain the prominent features in classic cases of otosyphilis, although symptom variability often complicates the diagnosis. It can be present in various stages of syphilis, from secondary to latent to neurosyphilis. From a cohort of 97 HIV-infected patients diagnosed with neurosyphilis[3], 4 had otosyphilis. All cases were treated with standard IV pencillin without subjective improvement. We present one severe case of otosyphilis that led to deafness. This patient received multiple rounds of treatment with penicillin and steroids without subjective improvement over a ten-month period.

Figure 1



MRI revealing "normal internal auditory canals without and with contrast. No involvement of the brainstem, cochlea, or involvement of the 7th/8th nerves bilaterally."
Note the 7th/8th cranial nerves show normal appearance on the left, similar to that on the right.

Support & Disclosures

UCSD IRB Approval #071931
Owen Clinic Master protocol: Retrospective Use of Existing Clinic Data
Supported by UCSD CFAR Core Services
Support for HNS partially supported by funds from NIH T32 training grant (DC000128)

Case Report

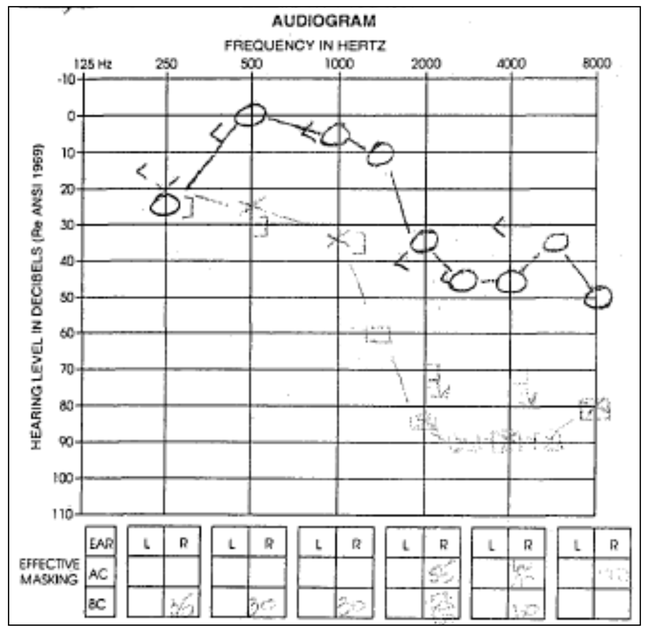
33 HIV+ M on ARVs with CD4 count 492 cells/ml & VL undetectable, history of anogenital condyloma, disseminated zoster at time of initial diagnosis, presented with three weeks of "ear infection" symptoms and associated vertigo.

RPR was positive, titer 1:32, but was nonreactive one year prior. Weber lateralized to the right, with a positive Rinne test, consistent with sensorineural hearing loss (SNHL). He was immediately started on IV PCN 2.4 million units with prednisone 60mg daily for two weeks to address neuronal inflammatory process. He had no subjective improvement following treatment, with audiogram noting normal to moderate R-SNHL and severe L-SNHL. A speech discrimination score was 56% from 36% on left (inset & figure 2).

The patient also received bicilllin injections once per week x 3 weeks as recommended by public health with resolution of vertigo; he developed right-sided tinnitus shortly thereafter, prompting additional prolonged steroid treatment as well as a second course of IV PCN.

He eventually had an MRI to rule out schwannoma (see figure 1). Following completion of a prolonged 6-week steroid course, he still had no improvement in symptoms.

Due to financial hardship, the patient was unable to follow up with head and neck surgery (HNS) and could not obtain a repeat audiogram. He was eventually lost to follow up and remained completely hearing-impaired on the right side. RPR titer 10 months after diagnosis was 1:1.



Audiogram, 2/17/11, at time of initial treatment: mild sloping to severe sensorineural hearing loss on left with poor discrimination. Right ear with mild to moderate hearing loss above 1500 Hz and 92% discrimination at 45 decibels. Also with normal tympanogram.

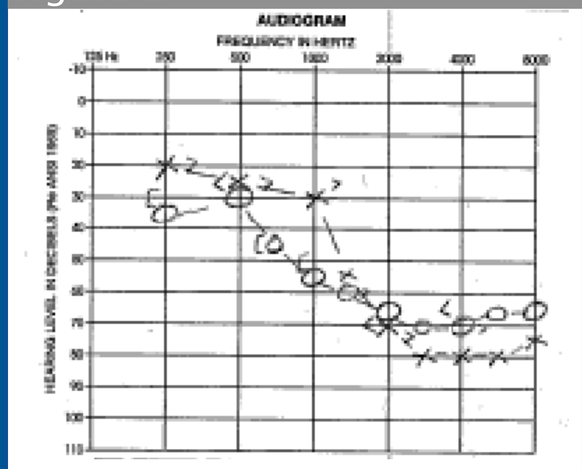
Discussion

Otosyphilis may present at various stages of syphilis and may present as an isolated syndrome distinct from neurosyphilis, although there is often secondary cranial nerve VIII involvement in as many as 57% of cases of neurosyphilis[3]; generally CSF in isolated otosyphilis is normal. It presents similarly to other inner ear disorders, making the diagnosis challenging. However, several case series suggest useful clinical criteria include the following: a) inner ear dysfunction (cochleovestibular dysfunction) confirmed by audiogram with b) confirmed positive treponemal tests.[1,2,4] The definitive diagnosis is difficult to make as it requires inner-ear fluid demonstrating a positive serology or bone histology. Audiogram may also show speech discrimination deficits. Imaging is often normal, although there have been reports of MRI revealing meningeal enhancement or infarction.

Treatment with penicillin G (either IM or IV) is often recommended based on the presumed stage of syphilis; with persistent symptoms after initial treatment, continuation at doses recommended for neurosyphilis are often instituted (aqueous crystalline penicillin G, 18-24 million units/day, administered at 3-4 million units IV every 4 hours or continuous infusion for 10-14 days)[5,6]. This raises the question of whether or not to treat as for neurosyphilis initially, regardless of CSF findings (based on high rates of co-existing neurosyphilis). It has been suggested that higher doses are required not only for CNS penetration but also perilymph penetration. It is not clear at which point adjunctive therapy with steroids should begin. However, with severe, sudden-onset hearing loss, urgent administration of corticosteroids is recommended (prednisone 40-80 mg/day for 2-4 weeks followed by taper).[2] The exact mechanism of their effect is not clear, although it is suggested that steroids may enhance passage of penicillin into inaccessible foci of organisms or suppress the immune response to organism lysis.[1] Follow up RPR titers have shown at least a four-fold decline, although this does not correlate well with symptom improvement.

Effects of otosyphilis can be reversible with early aggressive treatment. However, multiple studies have failed to show consistent improvement in auditory symptoms even with what is thought to be adequate treatment. Many groups have subsequently attempted to characterize those patients who are most likely to benefit from interventions. In one study of 48 patients, those least likely to regain hearing were those with severe symptoms at onset of treatment, although these findings, too, have been inconsistent in other studies.[4] Unfortunately, this patient did not regain his hearing in despite several courses of penicillin and adjunctive steroids administered within a timely manner.

Figure 2



Repeat audiogram, 3/24/11, with minimal left-sided improvement (56% discrimination) and significant worsening of right side, now similar to the left.

Conclusions

This case exemplifies both the diagnostic and therapeutic dilemmas that delay the appropriate diagnosis and treatment of otosyphilis. Without a suspicion for syphilis, patients may go a mean of 10 months without a diagnosis, increasing the risk of permanent auditory dysfunction. Equally concerning is that despite treatment, a substantial proportion of patients will not have any improvement. While efforts might be helpful to characterize those patients who are less likely to respond to therapy are helpful, they do not provide a solution to this ongoing problem.

Treatment to date has been based on protocols for particular stages of syphilis (i.e. late latent vs neurosyphilis), but there have been no studies comparing outcomes among different regimens. Also, there is no data to guide clinicians about use of adjunctive steroids and alternative treatment for refractory cases. Additional studies are needed to help standardize treatment for this disease process.

References

[1] "Otosyphilis mimics immune disorders of the inner ear." *Acta Oto-Laryngologica*, 2006;126:679-684.
[2] "Otosyphilis in HIV-Coinfected Individuals: A Case Series from Toronto, Canada." *AIDS Patient Care & STDs*. Vol 22, No 3, 2008.
[3] Dan, J et al. ACT HIV 2013 abstract.
[4] "Otosyphilis: A review of 85 cases." *Otolaryngology-Head and Neck Surgery* (2007)136,67-71.
[5] CDC Sexually Transmitted Diseases Treatment Guidelines, 2010. "Diseases Characterized by Genital, Anal or Perianal Ulcers."
[6] "Neurosyphilis: A Historical Perspective and Review." *CNS Neuroscience & Therapeutics*. 16(2010) e157-e168.
[7] Pasricha JM et al. "Otosyphilis: a cause of hearing loss in adults with HIV." *MJA* (2010)193:7;421-422.
[8] Little JP, et al. "Otosyphilis in a patient with human immunodeficiency virus: Internal auditory canal gumma." *Otolaryngol Head Neck Surg* (1995) 112(3):5.
[9] Mckenzie et al. "Otosyphilis and HIV: therapeutic dilemma and outcome of treatment." *BMJ Case Rep*. 2009;bcr01. 1464.