Auditory Disorder in Central Nervous System Miliary Tuberculosis: Case Report

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Abstract
We evaluated a 28-year-old female with a unilateral hearing loss of unusual pathogenesis, that of central nervous system miliary tuberculosis. Audiologic and otologic findings were consistent with left retrocochlear disorder, characterized by a profound hearing sensitivity loss, absent acoustic reflexes, normal otoacoustic emissions, and the presence of only wave I of the auditory brainstem response. Imaging studies revealed the presence of multiple punctate lesions, one of which was extra-axial and located at the left cerebellopontine angle. The pattern of audiometric test results, particularly the combination of normal otoacoustic emissions and profound hearing sensitivity loss, contributed importantly to the investigative sequence leading to the final diagnosis.

Key Words: Hearing loss, sensorineural, tuberculosis

Abbreviations: ABR = auditory brainstem response, CNS = central nervous system, DPOAE = distortion-product otoacoustic emission, F = frequency, LLR = late latency response, MLR = middle latency response

A uditory disorder associated with tuberculosis is rare. Conductive hearing loss has been reported as a result of Mycobacterium tuberculosis of the temporal bone (Birrell, 1973; Skolnik et al, 1986; Yaniv et al, 1986). The loss is usually associated with chronic otitis media with purulent effusion, thickened tympanic membrane and middle ear mucosa, ossicular destruction (Yaniv et al, 1986), and/or tympanic membrane perforation (Skolnik et al, 1986). Sensorineural hearing loss associated with tuberculosis has been reported due to labyrinthitis secondary to meningitis (Vernon, 1967) or osteomyelitis of the temporal bone (Kearns et al, 1985). It can also occur as a result of ototoxic medications used to treat a primary infection (Vernon, 1967).

Tuberculosis is an infectious disease caused by Mycobacterium tuberculosis. It is characterized by the formation of tubercles, or small, rounded nodules, in tissue. Although the most common site of tuberculosis infection is the lungs, the infection can be carried through the blood stream to other organs from the primary infection site. One form of tuberculosis results in the formation of numerous small tubercles distributed throughout the central nervous system (CNS). These tubercles are referred to as miliary lesions because of their resemblance to millet seeds. CNS miliary tuberculosis occurs as a result of the dissemination of the tuberculous bacilli through the brain by the blood stream.

Although miliary tuberculosis has the potential to affect any portion of the CNS, we are unaware of any reports of auditory disorder secondary to the disease. This article summarizes clinical data from the audiologic, otologic, and radiologic evaluations of a patient with a unilateral hearing disorder secondary to CNS miliary tuberculosis. The auditory disorder was characterized by a profound hearing sensitivity loss of retrocochlear origin.
CASE REPORT

Subject

Patient HC is a 28-year-old female of Taiwanese descent. She was referred to the California Ear Institute at Stanford for audiologic and otologic consultations as a result of a sudden hearing loss in her left ear. At the time, she was being treated by the Infectious Diseases Department, Stanford University Medical Center, for long-standing tuberculosis.

Four weeks prior to the referral, the patient reported noticing that she could not hear out of her left ear on the telephone. She also reported feeling a “heaviness” on the left side of her head. Her hearing history was otherwise unremarkable. There was no family history of hearing loss or history of other risk factors for hearing loss.

The neurotologic evaluation, including otoscopic examination, was normal. She had no significant otologic history and no reported dizziness or tinnitus. Significantly, the patient had a long history of tuberculosis and had recently begun medical therapy for miliary tuberculosis involving her nervous system. The medical therapy consisted of a drug regimen that included isoniazid, rifampin, streptomycin, and ethambutol.

Clinical Results

Immittance audiometry indicated normal middle ear function bilaterally. Crossed and uncrossed acoustic reflexes were absent when the eliciting signal was presented to the left ear. Results are shown in Figure 1. Tympanometry was carried out with conventional clinical instrumentation (Grason-Stadler GSI-33) using a 220-Hz probe tone. Tympanograms for both the right and left ears were normal Type A, and static immittance was within normal limits bilaterally. Right crossed and right uncrossed reflexes were present at normal levels. Left crossed and left uncrossed reflexes could not be measured at or below 110 dB HL. This reflex pattern is consistent with an afferent abnormality on the left, indicating either a significant cochlear or retrocochlear disorder on that side.

Pure-tone and speech audiometric results are shown in Figure 2. Left ear results revealed a profound hearing loss. Hearing sensitivity could only be measured at 250 and 500 Hz at 105 and 110 dB HL, respectively. No responses were obtained at any other frequencies, and no responses were obtained to bone-conducted signals at equipment limits. The speech awareness threshold was 105 dB HL. Word recognition ability could not be measured. Right ear results
showed normal hearing sensitivity from 250 to 4000 Hz and a minimal sensitivity loss at 6000 and 8000 Hz. The word recognition score was 100 percent at 80 dB HL. Pure-tone and speech audiometric measures were carried out with conventional clinical instrumentation (Grason-Stadler GSI-16), calibrated to American National Standards Institute's specification for audiometers (ANSI, 1996). Insert earphones (Etymotic ER-3A) were used. NU-6 word lists were used to determine word recognition scores.

As a matter of routine clinical procedure in the case of unilateral hearing loss, a Stenger test was carried out to assess the organicity of the hearing loss. The result of a speech Stenger test was negative for functional hearing loss. As a further indication of the organic nature of the loss, a shadow curve was noted on the left audiogram at expected levels for insert earphones when the right ear was not masked.

At this point in the evaluation, the results indicated the presence of a sudden, organic, profound, sensorineural hearing loss on the left ear. Absent reflexes, a shadow curve, and a negative Stenger verified the organicity of the response. The only concern about these results arose from the seeming incongruity between the extreme degree of loss and the passive nature of its discovery by the patient. A hearing loss of this magnitude, if it did occur suddenly, might be expected to be noticed more generally than during telephone use. As a result, additional testing was carried out to verify the nature of the disorder.

Distortion-product otoacoustic emissions (DPOAEs) were measured to assess cochlear function. DPOAE amplitudes as a function of $F_2$ frequency are plotted in Figure 3. Results showed substantive emissions across the frequency range.
Auditory evoked potential measurements were carried out in an effort to clarify the disparity between results of otoacoustic emissions testing and those obtained with behavioral and immittance audiometry. Auditory evoked potentials, including the auditory brainstem response (ABR), middle latency response (MLR), and late latency response (LLR), are shown in Figure 5. Right ear responses were well formed, with component peaks at normal absolute and interpeak latencies. Left ear results were abnormal. No MLR or LLR responses were recorded. For the ABR, only component wave I was observable. The absolute latency of wave I was 1.5 msec in both ears. Wave I was reliably recorded down to 70 dB nHL. The presence of Wave I on the left is consistent with the otoacoustic emissions results indicating near-normal cochlear function. The absence of later waves suggests a site of lesion at the proximal end of the VIIIth nerve or low auditory brain stem. Auditory evoked potentials were recorded with conventional instrumentation (Otodynamics ILO-92) for both the right and left ears. These results are consistent with normal cochlear outer hair cell function in both ears. To further assess the left ear emissions, input-output functions were determined at 2000 and 5000 Hz. Results are shown in Figure 4 and confirmed that, despite the presence of a profound hearing sensitivity loss on the left, cochlear function, or at least outer hair cell function, was normal. DPOAEs were recorded with conventional clinical instrumentation (Otodynamics ILO-92). The frequency ratio of F2 to F1 was 1.2. F1 amplitude was 65 dB SPL; F2 amplitude was 55 dB SPL.

Figure 4 Distortion-product otoacoustic emission input-output functions at an F2 frequency of 2000 Hz (A) and 5000 Hz (B) from the left ear of patient HC, a 28-year-old female with CNS miliary tuberculosis.

Figure 5 Auditory evoked potentials in patient HC, a 28-year-old female with CNS miliary tuberculosis, including the auditory brainstem response (ABR), middle latency response (MLR), and late latency response (LLR).
Clinical instrumentation (Nicolet Spirit). ABR recordings were made in response to alternating polarity clicks presented at 90 dB nHL at a rate of 23.7/sec; MLR recordings were made in response to tone bursts presented at 60 dB nHL at a rate of 0.5/sec.

Magnetic resonance imaging studies revealed the presence of multiple bilateral supratentorial punctate lesions, confirming a diagnosis of CNS miliary tuberculosis. Results are shown in Figure 6A. One of the lesions, and the likely cause of the auditory disorder, was extra-axial and located at the left cerebellopontine angle. Results are shown in Figure 6B.

**COMMENTS**

HC is a 28-year-old female with a unilateral hearing loss of unusual pathogenesis, that of CNS miliary tuberculosis. Audiologic and otoacoustic emissions results were consistent with left retrocochlear disorder, characterized by profound sensorineural hearing loss, absent acoustic reflexes, normal DPOAEs, and the presence of only ABR wave I. Imaging studies revealed the presence of multiple punctate lesions, one of which was extra-axial and located at the left cerebellopontine angle.

HC's hearing sensitivity loss appeared to be due to retrocochlear rather than cochlear disorder. Audiometric and immittance measures were equivocal in that the profound hearing sensitivity loss and absent acoustic reflexes could be attributable to either cochlear or retrocochlear disorder. However, auditory evoked potential results were in agreement with otoacoustic emissions in implicating the VIIIth nerve or auditory brain stem. DPOAEs were present across the frequency range, and input/output functions suggested healthy outer hair cell function. Similarly, that the wave I was present at intensity levels as low as 70 dB in an ear with a profound hearing loss indicated that the loss was probably not cochlear in nature. Of course, the fact that a patient has CNS miliary tuberculosis does not preclude that person from having a sudden idiopathic hearing loss of cochlear origin. However, if the loss in this case were of cochlear origin, both the DPOAEs and the ABR would be expected to be absent.

Tuberculosis as a cause of sensorineural hearing loss has been reported infrequently in the literature. Although sensorineural hearing loss has been reported previously in association with meningitis secondary to tuberculosis, it is difficult to separate the effects of the disease from the effects of the treatment, which often consists of ototoxic antibiotics. Regardless, in the case of HC, there was no indication of meningitis, and
the unilateral nature of her disorder was inconsistent with any effects of ototoxic medications.

HC is a case of sudden sensorineural hearing loss secondary to tuberculoma involving the root-entry zone of the VIIIth cranial nerve. To our knowledge, a similar case has not been previously reported. Although intracranial tuberculoma has been previously reported (Skolnik et al., 1986), sensorineural hearing loss as a result has not. Magnetic resonance imaging allowed us to localize the lesion to the VIIIth cranial nerve at the cerebellopontine angle. Previous cases of tuberculosis with sensorineural hearing loss may have had similar lesions that were not localizable prior to the advent of such imaging.

Although sudden sensorineural hearing loss is seldom reported in patients with tuberculosis, this case demonstrates that the disease, as a result of the formation of a tubercle at some precise point in the auditory nervous system, can cause a hearing sensitivity loss of retrocochlear origin.

The use of otoacoustic emissions was helpful in the diagnosis of the disorder. Upon initial assessment, the sudden sensorineural hearing loss appeared unrelated to the known history of tuberculosis. Without the DPOAE results, the hearing loss might have been viewed as an idiopathic sudden hearing loss, with immediate medical intervention aimed at the possible underlying causes. With the DPOAE results, the loss was immediately viewed as retrocochlear in nature, and clarification was sought with imaging studies.

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REFERENCES


