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From silence to sound – a case of autoimmune ear disease

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ABSTRACT

Autoimmune inner ear disease (AIED) is a rare rheumatological cause of sensorineural hearing loss, accounting for less than 1% of worldwide hearing loss. It presents with rapidly progressive, unilateral or bilateral SNHL, often fluctuating. The progression is over weeks to months, along with vestibular symptoms. Diagnosing the condition is challenging because no adopted standardized tests are available. Hence, AIED is a challenging diagnosis based on exclusion. Clinical characteristics and how the patients respond to immunosuppressive agents are used for the diagnoses of AIED. Only a few cases have been reported in India till now. Here, we present a rare case of Autoimmune Ear Disease without any underlying systemic autoimmune disease.

Keywords: autoimmune, sensorineural hearing loss, autoimmune ear disease

INTRODUCTION

Autoimmune inner ear disease (AIED) is a rare rheumatological cause of sensorineural hearing loss, which accounts for less than 1% of hearing loss worldwide [1]. It presents with rapidly progressive, unilateral or bilateral SNHL, often fluctuating. The progression is over weeks to months, along with vestibular symptoms. The diagnosis is difficult due to the lack of standardized tests. Hence, AIED is a challenging diagnosis based on exclusion [3]. Clinical characteristics and how the patients respond to immunosuppressive agents are used for the diagnoses of AIED. Only a few cases have been reported in India till now. Here, we present a rare case of Autoimmune Ear Disease without any underlying systemic autoimmune disease

CASE HISTORY

Mr. S, a 50-year-old male with no known comorbidities, presented with complaints of unilateral hearing loss in his left ear for two weeks and a 1-year history of quiescent Chronic Suppurative Otitis Media in his right ear. Otoscopy revealed tympanic membrane perforation on the right side. MRI brain showed no abnormalities. On evaluation, the left ear tympanogram was A-type (ruling out middle ear involvement), and the right ear tympanogram was B-type (indicating middle ear pathology in the right ear).

INVESTIGATIONS

The audiometry assessment indicated sensorineural hearing loss in the left and mixed hearing loss in the right ear. C-Reactive protein was high (13 mg/l). Rheumatoid factor, Anti-Cyclic Citrullinated peptide antibody, Anti-Neutrophil Cytoplasmic Antibodies, Anti-Nuclear Antibody and VDRL were all negative. The ophthalmological exam was normal, without any signs of retinal vasculitis.

Another audiometry was done 3 weeks after his first visit, which showed improved hearing in the left ear. Speech Identification Score improved in both ears compared to the previous audiometry.

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Unmasked Air		Unmasked Bone	Unmasked Air	Masked Bone
R	○	<	△	⊏
L	X	>	□	⊐

FIGURE 1. The above figure depicts the symbols used in the following audiograms

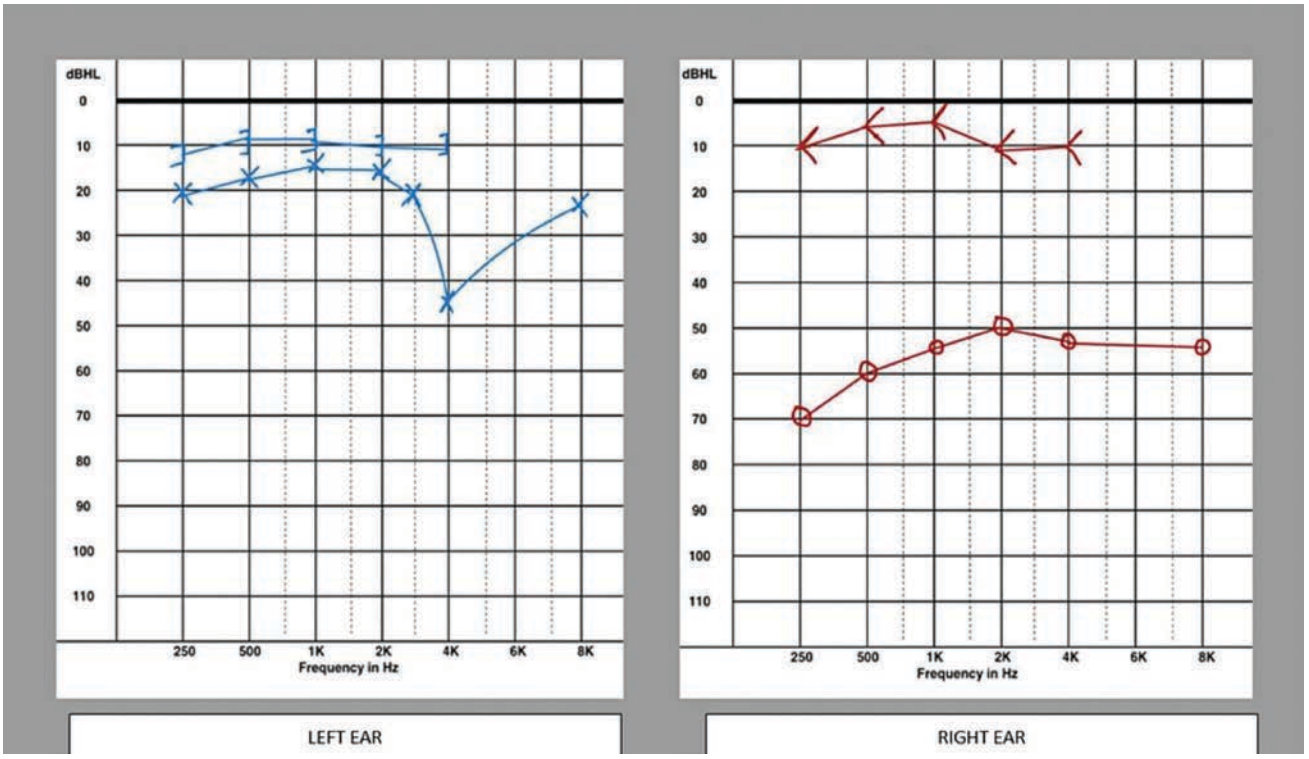

FIGURE 2. Left and right audiogram - done 1 year before diagnosis of Autoimmune Ear Disease showing conductive hearing loss

The patient reported continuous improvement in all of his symptoms.

Differential diagnosis

Autoimmune Ear Disease, Meniere's Disease, Presbycusis

Treatment

A diagnosis of right-sided Chronic Suppurative Otitis Media and bilateral sensorineural hearing loss was made. The patient was initially started on steroids at the rate of 0.5mg/kg and slowly tapered over 4 to 6 weeks. He was started on Methotrexate later.

Outcome and follow-up

The patient was called in for periodic audiograms, which revealed improved hearing in his left ear. He is stable and doing well.

DISCUSSION

AIED is often misdiagnosed as Meniere's disease. It poses a significant challenge due to the absence of standardized diagnostic criteria and reliable tests. Consequently, the diagnosis relies on a process of exclusion. The immune response can directly affect the inner ear, but it may also cause damage through the deposition of immune complexes or systemic immune-mediated diseases.

Following the activation of the immune response and the release of interleukin (IL)-1β, the autoimmune response is triggered. Activated circulating leukocytes and immunoglobulins, responding to antigenic stimuli, can chemotactically target the inner ear. The activated lymphocytes traverse the blood-labyrinthine barrier, potentially entering the cochlea through the spiral modiolar vein of the Scala tympani and ultimately reaching the endolymphatic sac [3]. The range of available treatments is restricted,

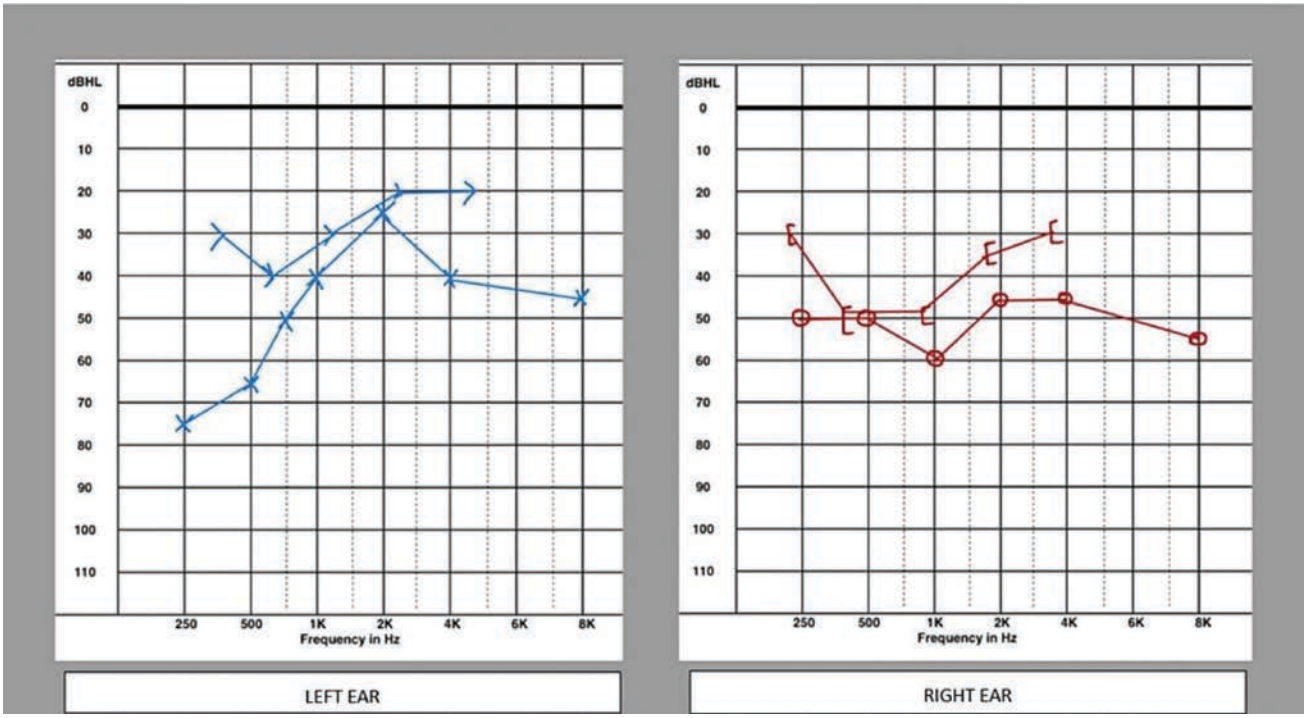

FIGURE 3. Left and right ear audiogram on presentation

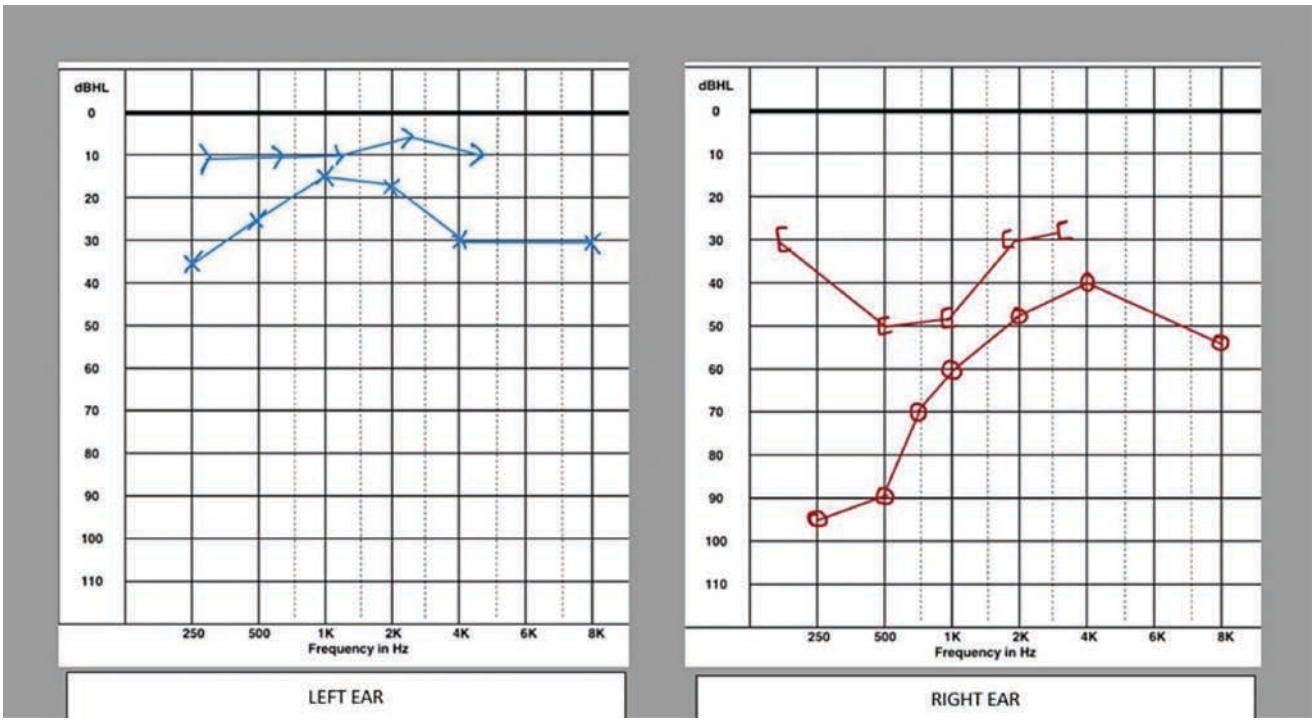

FIGURE 4. Left ear audiogram – after treatment: Improvement in hearing present compared to before treatment. Right ear audiogram – after treatment, showing mixed hearing loss without any improvement after treatment was completed

and corticosteroids are the singular validated treatment. In instances of treatment inefficacy, supplementing with other immunosuppressive agents may become essential. These alternatives encompass Methotrexate, cyclophosphamide, azathioprine, and cyclosporine A, each demonstrating superior outcomes compared to relying solely on corticosteroids [4]. The rate of hearing loss advances too swiftly to be indicative of presbycusis and unfolds too gradually to diagnose sudden sensorineural hearing loss (SNHL) definitively [5].

Given that recognizing AIED dates back to 1979, determining its incidence poses a challenge. McCabe documented a group of individuals experiencing unexplained, advancing bilateral SNHL [6]. The symptoms in AIED are bilateral hearing loss with or without giddiness, vertigo and ataxia. This patient also presented with general symptoms that point to-

wards many diagnoses. The pace of hearing loss is too rapid to align with a presbycusis diagnosis and too gradual to characterize SNHL definitively [7]. Hearing loss can result from the abnormal cochlea, auditory nerve function, or higher central auditory perception or processing aspects. Diagnosis is mainly based on clinical characteristics and response to immunosuppressive agents [8]. A possible explanation is that the right ear is affected by CSOM earlier released toxins leading to Autoimmune Ear Disease bilaterally. In the above patient, we presume that the left ear improved more than the right ear because of previous history of CSOM in the right ear, which might have delayed the recovery from Autoimmune Ear Disease.

CONCLUSION

Autoimmune inner ear disease (AIED) is a rare rheumatological sensorineural hearing loss which accounts for less than 1% of hearing loss worldwide. Sensorineural hearing loss due to immune-mediated dysfunction needs immediate treatment, which can only be started if the condition is diagnosed effectively. Only a few cases have been reported in India till now.

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