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Bilateral Vestibulopathy Due to Severe Cochlear Otosclerosis: A Well-Known Condition Without Any Favorable Solution

Case Report

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Abstract

Bilateral vestibulopathy is a rare condition. It is one of the most difficult balance problems to treat because nearly all patients experience long-term unsteadiness. In this report, a 39-year-old woman gradually developing chronic dizziness and instability due to bilateral vestibulopathy as a result of progressive severe cochlear otosclerosis was presented with review of the literature.

Keywords: Otosclerosis, hearing loss, bilateral vestibulopathy, treatment

Introduction

Inner ear involvement of otosclerosis exhibits a well-known clinical picture. Cochlear otosclerosis is either a further stage of stapedial otosclerosis with spread of oval window focus to the cochlea or an isolated presentation (1). It is also well-known that not every case proceeds to cochlear otosclerosis. It is difficult to predict the actual incidence of cochlear otosclerosis. Although it is not convenient enough, histopathological studies of the temporal bones may provide some signs.

In one of the earlier and extensive studies, Schuknecht and Barber (2) reported five cases with cochlear otosclerosis without stapedial fixation among 164 temporal bones with otosclerosis. Hinojosa and Marion (3) reported temporal bone findings in six cases with otosclerosis with sensorineural hearing loss among 125 bones. They found no stapedial fixation. Balle and Linthicum (4) reported temporal bone findings in seven cases with isolated cochlear otosclerosis. There is sufficient histopathological evidence to support the presence of otosclerotic lesions located in different regions of the otic bone without any oval window pathology.

Progressive sensorineural hearing loss and chronic unsteadiness are major clinical aspects of cochlear otosclerosis. Abnormal bone turn-over, disturbed blood circulation and venous congestion, and enzymatic activities will eventually lead to atrophy of the spiral and scarpa ganglions (5). In this report, we present a case with bilateral vestibulopathy due to severe cochlear otosclerosis. Her hearing is favorable with hearing aids. However, she is desperate due to chronic balance problem since no treatment seems to help.

Case Presentation

A 39-year-old woman had bilateral progressive hearing loss and chronic unsteadiness for five years. Hearing on the right ear worsened rapidly in the last one year. She has been followed up since the beginning of her disease. Her recent audiogram revealed bilateral profound hearing loss (Figure 1). Word discrimination score was 30% on the right ear and 42% on the left ear. She has been wearing bilateral hearing aids for five years and had good communication skills. However, she had major difficulty in daily life due to chronic unsteadiness. Her examination demonstrated difficulty in tandem gait with closed eyes. She had no spontaneous nystagmus. Romberg and Fukuda stepping tests were all abnormal. Bilateral caloric testing showed canal paresis on both sides (videonystagmographic analysis; MicroMed, Inc., Chatham, IL, USA) (Figure 2). Her temporal bone tomography revealed severe demineralization around the cochlea and the vestibule (Figure 3). She had several med-

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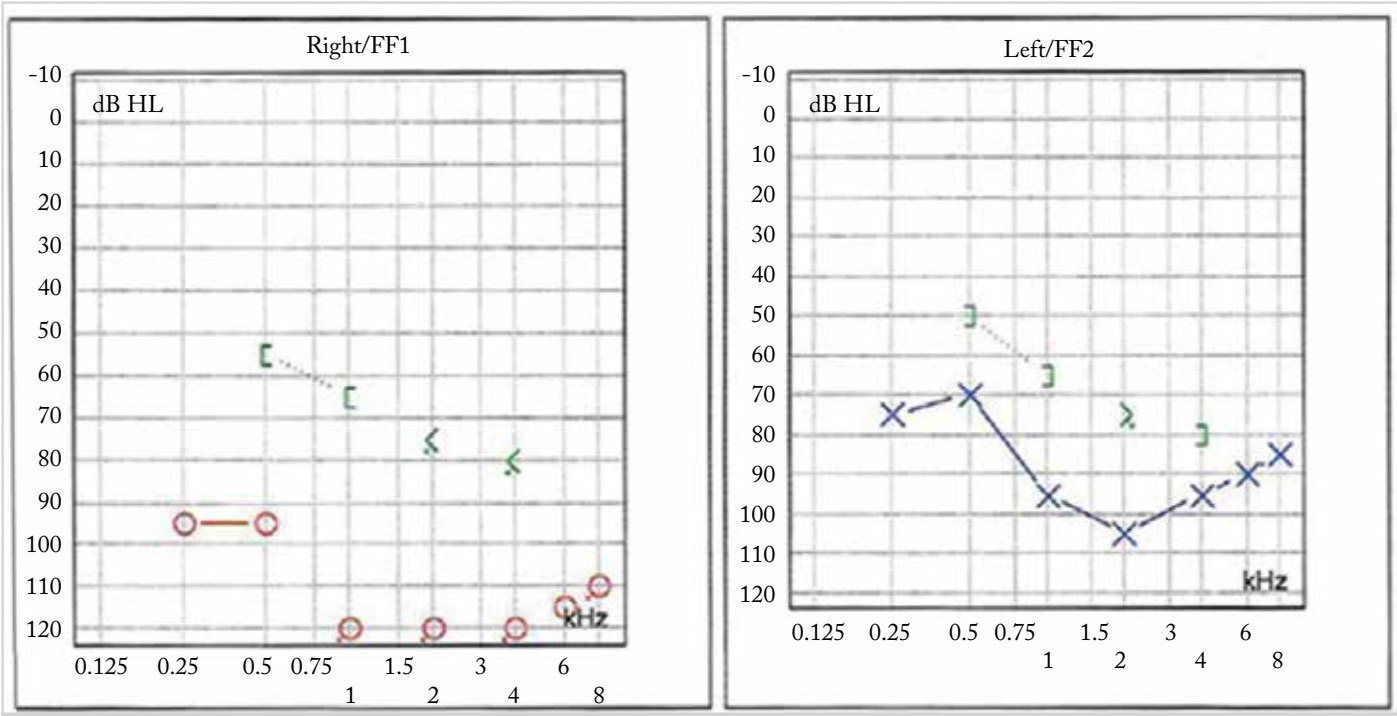

Figure 1. Audiogram of the patient demonstrating bilateral profound sensorineural hearing loss

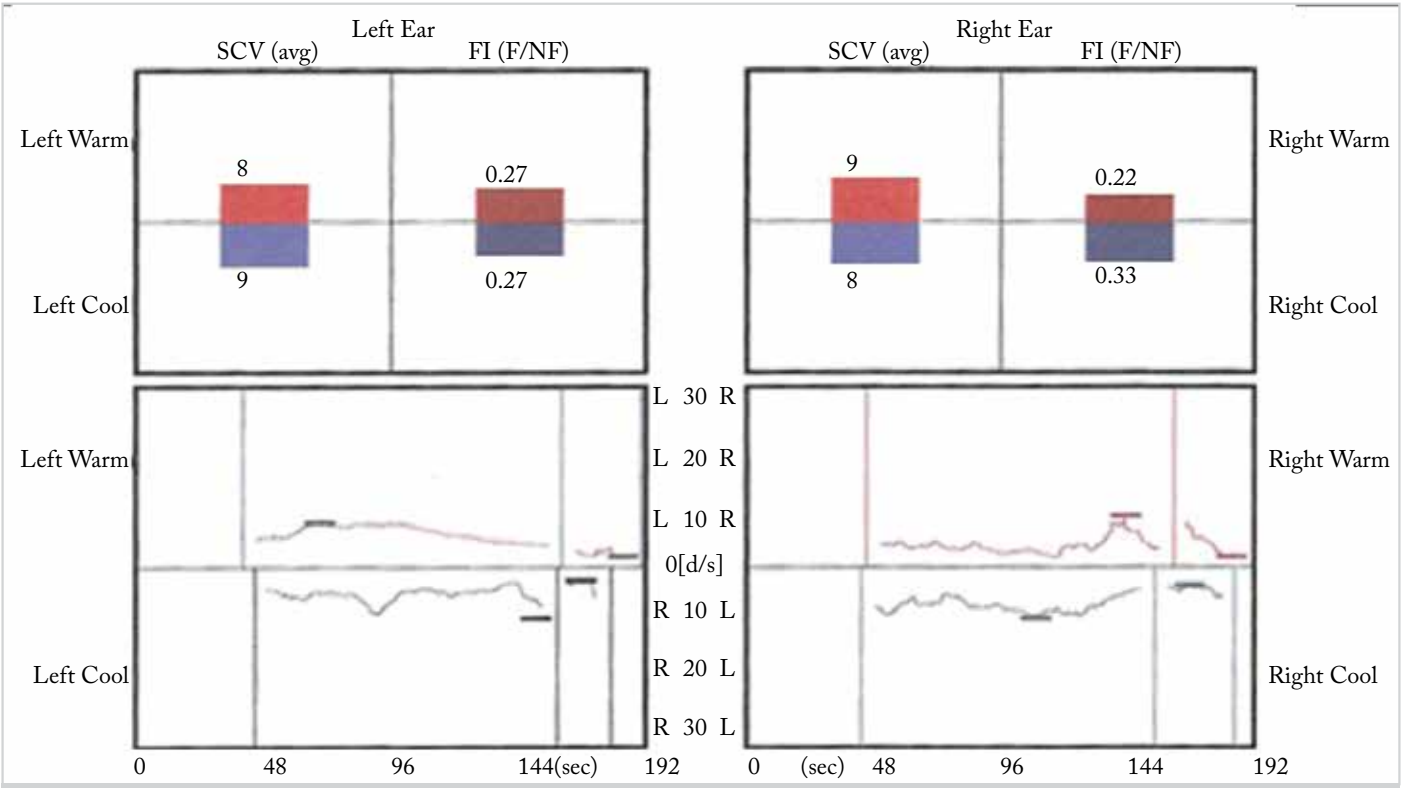

Figure 2. Bilateral caloric testing showing decreased vestibular response on both sides

ications in the past. She had sodium fluoride therapy for a total of 14 months on several intervals without any improvement. She used betahistine, corticosteroids (oral and intratympanically), piracetam, trimetazidine, ginkgo biloba, and cinnarizine. She refused to undergo any surgery for chronic vestibular problem because of the risk to interfere with her hearing. At the moment, she had vestibular exercises that appear to offer minimal relief. Informed consent was obtained from the patient.

Discussion

The precise role and mechanism whereby otosclerosis is associated with vertigo remain unclear. Vestibular complaints can be seen in 25%-45% of patients with otosclerosis (6). However, real and true vertigo is seldom. Many cases of vertigo due to cochlear otosclerosis may remain undiagnosed. Balance problems are usually associated with active stages of the disease. Postural

Figure 3. Temporal bone computed tomography revealing severe demineralization around the cochlea and the vestibule

instability is common, and problems are not related with head movements (7). Caloric analysis is usually normal. Enzymatic activities, vascular insult during active phase, toxic substances from otosclerotic focus, and biochemical change in endolymph and perilymp could be some of the reasons (8). Vestibular problems are more common in cochlear otosclerosis. It is similar to chronic unsteadiness and may last for weeks, months, and sometimes for years.

Histopathological studies of the temporal bone demonstrate several vestibular pathologies in patients with otosclerosis (9). However, it is not entirely clear how change in vestibular morphology in otoscelorosis is related with vestibular symptoms. Saim and Nadol (10) have reviewed the scarpa ganglion in 217 temporal bones with otosclerosis. They found lower cell count in patients with registered vestibular problems than those with controls and also those with otosclerosis without registered vestibular problems. There was no connection between the extension of otosclerotic focus and symptoms. It appears that vestibular lesions are more common if the otosclerotic process spreads to the inner ear (5). Loss of vestibular hair cells accounts for vestibular problems in some patients (9).

Bilateral vestibulopathy is a rare condition. It is one of the most difficult balance problems to treat because nearly all patients experience long-term unsteadiness and oscillopsia due to disturbance of the bilateral vestibulo-ocular response. Sodium fluoride has been reported to be effective in cochlear otosclerosis (6). It does not reverse an otosclerotic focus or restores bone resorption. However, it appears to be effective in preventing the development of new active areas. Our case had no benefit from any medical therapy. Surgical interventions to the labyrinth or vestibular nerve and intratympanic gentamicin application are out of option for this patient. Residual function of the vestibular system on both sides is not sufficient to promote spontaneous recovery. Vestibular implant may provide an opportunity to restore the vestibular function to be able to selectively stimulate the vestibular system in patients with disabling bilateral vestibular loss for whom there is currently no other available option. Delivering motion information to the central nervous system using electrical stimulation aids to restore gaze stabilization and postural control that will allow new clinical applications in the near future (11).

Conclusion

Treatment of bilateral loss of vestibular function is always difficult and challenging. Vestibular rehabilitation can partially restore the deficit. The reported case was desperate and had some relief after vestibular rehabilitation. However, there is still no effective and approved treatment of chronic postural instability while preserving hearing in similar patients. Vestibular implants show promising results. However, further studies are required for cochlear otosclerosis.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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Conflict of Interest: The author has no conflict of interest to declare.

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