Bilateral Cochlear Otosclerosis: Clinical and Radiological Findings

Bilateral Koklear Otoskleroz: Klinik ve Radyolojik Bulgular

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Abstract

In this report, we presented a 41-year-old female patient with unilateral hearing loss in the right ear. Previously she had a traffic accident, but had no history of ear infection. High resolution computed tomography revealed bilateral cochlear otosclerosis, with no evidence of ossicle dislocation. It was thought that cochlear otosclerosis caused mixed type hearing loss in the right ear by involving the lateral wall of the otic capsule and sensorineural hearing loss in the left ear. We think that fenestral and cochlear type otosclerosis incidentally occurred along a continuum in the right ear of the patient, and that radiological investigation is crucial in clarifying the possible concomitant pathologies in cases of hearing loss.

Keywords: hearing loss; high resolution computed tomography; otosclerosis

Öz

Bu raporda, 41 yaşında, sağ kulağında tek taraflı mikst tip işitme kaybı olan bir kadın hastayı sunduk. Hasta daha önce bir trafik kazası geçirmişti; ancak kulak enfeksiyonu öyküsü yoktu. Yüksek çözünürlüklü bilgisayarlı tomografi iki taraflı koklear otoskleroz olduğunu gösterdi, ama kemikçik dislokasyonu bulgusu saptanmadı. Koklear otosklerozun sağ kulakta otik kapsül lateral duvarını tutarak mikst tip işitme kaybına, sol kulakta ise sensörinöral işitme kaybına yol açtığı düşünülmüştür. Hastanın sağ kulağında koklear otoskleroz ile fenestral otosklerozun insidental olarak bir devamlılık arz ettiğini ve işitme kaybı vakalarında eşlik eden olası patolojilerin ortaya konması için radyolojik incelemenin önemli olduğunu düşünmekteyiz.

Anahtar Sözcükler: işitme kaybı; otoskleroz; yüksek çözünürlüklü bilgisayarlı tomografi

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INTRODUCTION

Otosclerosis is a focal osseous dyscrasia of the temporal bone that primary affects the endochondral bone of the otic capsule, cochlear capsule, or both in humans (1). Although most cases are asymptomatic, otosclerosis is one of the most common causes of conductive hearing loss. Depending on the involvement area, different symptoms might be seen, such as conductive, sensorineural, or mixed type hearing loss (2). Although the definitive diagnosis is established by histopathological investigation, high resolution computed tomography (HRCT) is the gold standard imaging modality in the diagnosis of otosclerosis (3,4).

In this report, we presented the diagnosis of bilateral cochlear otosclerosis of a patient suffering from unilateral hearing loss, and discussed the case in light of the literature.

CASE

A 41-year-old female patient presented with the complaint of unilateral hearing loss persisting for the last three years. Previously she had a traffic accident, but had no history of ear infection.

Clinical examination revealed that the tympanic membranes were intact, the Weber's sign was lateralized to the right, and the Rinne's sign was negative with the 512 Hz tuning fork. Pure tone audiogram revealed moderate-to-severe mixed type hearing loss in the right ear (Figure 1) and mild-to-severe sensorineural hearing loss in the left ear (Figure 2). Acoustic impedance revealed a type A graphic.

HRCT imaging was performed to elucidate the etiology. Low-density areas like a double-ring sign were observed in both pericochlear areas, which is diagnostic for bilateral cochlear otosclerosis (Figure 3). There was no evidence of ossicle dislocation, and acoustic tympanogram did not exhibit a type Ad graphic. Accordingly, it was thought that cochlear otosclerosis caused mixed type hearing loss in the right ear by involving the lateral wall of the otic capsule and sensorineural hearing loss in the left ear, and that fenestral and cochlear type otosclerosis occurred along a continuum in the right ear. The patient refused the exploratory surgical options offered to her, i.e., tympanotomy/stapedotomy. She is still in our follow-up program.

DISCUSSION AND CONCLUSION

Otosclerosis is the focal osseous dyscrasia specific to the endochondral bone of the otic capsule that consists of circumscribed foci of new, softer and more vascular bone, instead of the avascular bone of the adults (1,2). It is believed that the cartilaginous remnants trapped within the endochondral bone of the otic capsule are remodeled and replaced by highly vascular, immature bone. The resorption of the perivascular bone in the spongiotic phase and the deposition of immature, collagen-deficient and dense bone in the sclerotic phase are simultaneous and continuous within a focus of the disease (2,5). It is revealed that genetic and environmental factors trigger the bone remodeling. The role of the measles virus has been investigated (6). Intense hormonal activity in situations like pregnancy has been reported to exacerbate otosclerosis (6).

Otosclerosis was described about two centuries ago; Valsalva first described stapes fixation in 1735. Politzer discovered the true pathology in the labyrinthine capsule in 1893, but the term otosclerosis remained in use (2). It is an autosomal-dominant hereditary disease with variable penetrance. It is about two times more frequent in females (19%) than in males (7%). The symptoms develop in the third and fourth decades of life (7). It accounts for 5–9% of all hearing losses and 18–22% of conductive hearing losses, and in 80–90% of all cases otosclerosis occurs bilaterally (8).

Depending on the topography of the lesions, otosclerosis can be categorized into two types: fenestral and retrofenestral/cochlear. Fenestral lesions are in the lateral wall of the otic capsule; the round window, promontory, and tympanic segment of the fallopian canal may be affected. The most common location is the anterior part of the oval window. The cochlear type affects the labyrinthine capsule. These lesions may be found in the pericochlea, auditory canal, and vestibular aqueducts (9,10). The most common location of osteosclerotic lesions is the fissula ante fenestram, the anterior part of the oval window (7). Although most cases are asymptomatic, otosclerosis is one of the most common causes of conductive hearing loss. It can spread across the stapedial annular ligament and fix the stapes, causing conductive hearing loss. It can also result in sensorineural hearing loss due to an oto-

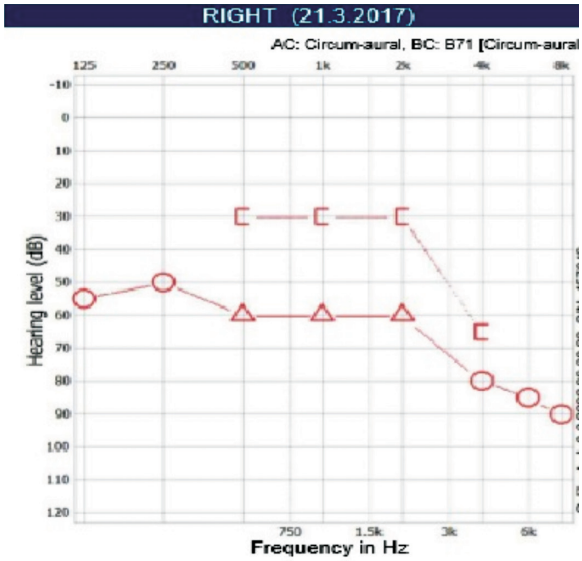

Figure1. Pure tone audiogram: moderate-to-severe mixed type hearing loss in the right ear.

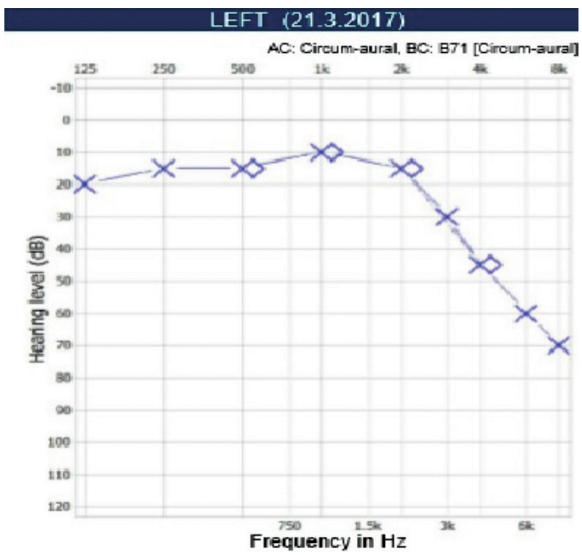

Figure 2. Pure tone audiogram: mild-to-severe sensorineural hearing loss in the left ear.

sclerotic focus in the cochlear capsule (2). Proteolytic enzymes in the cochlea may be responsible for injury of the cochlear capsule and cause hyalinization of the spiral ligament (11). A rare variant of otosclerosis is the malignant otosclerosis that affects not only the lateral wall of the otic capsule, but also the labyrinthine capsule, causing progressive mixed hearing loss (12). This variant shows that the two types of otosclerosis might not be two separate entities, but a continuum.

The medical history, physical examination, audiological testing and imaging play a significant role in the diagnosis of otosclerosis. The clinical findings include conductive, mixed or rarely sensorineural hearing loss, tinnitus and vertiginous symptoms with no sign of inflammation in the middle ear (11,13). The conductive hearing loss and tinnitus are the main symptoms (14). Our patient presented to our clinic with the complaint of unilateral hearing loss in the right ear, which was revealed by pure audiometry to be mixed type hearing loss (Figure 1). She also had sensorineural hearing loss in the left ear (Figure 2). There was no tinnitus. Although the past traffic accident might have been resulted in ossicle dislocation, this was not supported by the tympanometric evaluation. During further investigation the cochlear otosclerosis was diagnosed by the characteristic double-ring sign and there was no evidence of ossicle dislocation, ossicle fixation or cholesteatoma in temporal bone HRCT.

HRCT is the gold standard imaging modality in the diagnosis of otosclerosis, with a high specificity and positive predictive value (1,3,4). Recent studies report that HRCT has >90% sensitivity in otosclerosis diagnosis. It shows the demineralized lesions in the otic capsule (stapes footplate, cochlea, and labyrinth) and the lesions that appears as a lucent or hypodense focus (1,3,4,11,15). A thickened footplate, narrowed oval window or round window niche are the HRCT findings in the fenestral type of otosclerosis (15). The double-ring sign is the classical imaging appearance of the cochlear type of otosclerosis in HRCT. It reveals the hypointense demineralized lesions surrounding the cochlea (3,11,15). Osteogenesis imperfecta, Paget's disease, ankylosing spondylitis, rheumatoid arthritis, and syphilis are the other rare diseases that demineralize the cochlear capsule, and they can mimic the appearance of cochlear otosclerosis in HRCT (3,11,15). HRCT may also play a significant role in distinguishing otosclerosis from other pathological conditions like tympanosclerosis, cholesteatoma, ossicular fixation, and congenital malformations that cause conductive hearing loss (1,3,4). In our case, low-density areas that caused the double-ring sign were observed in both pericochlear areas (Figure 3A–B).

Bilateral cochlear otosclerosis is usually asymptomatic. It can be diagnosed incidentally by HRCT

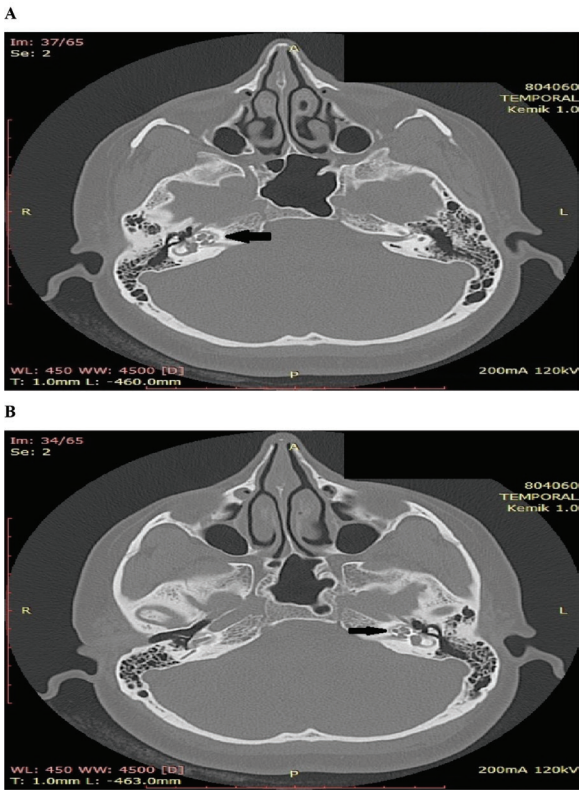

Figure 3. HRCT highlights the differences in the density of the capsule's outline, showing the "double-ring sign," a low-density demineralized endochondral defect outlining the cochlea (axial plane HRCT imaging of the temporal bone) **A.** The double-ring sign in the right ear (black arrow) **B.** The double-ring sign in the left ear (black arrow).

imaging. Accordingly, meticulous investigation should be performed in patients with hearing loss, together with radiological investigation to clarify the possible concomitant pathologies.

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