Case 1945

Fenestral and cochlear otosclerosis
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Section: Head & neck imaging
Imaging Technique: CT
Case Type: Clinical Cases
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Patient: 72 years, male

Clinical History:

The patient presented with a progressive bilateral hearing loss.

Imaging Findings:

The patient presented with complaints of a bilateral hearing loss (on the left side more than on the right side), progressive over the last 40 years. There were no complaints of vertigo or tinnitus. An otoscopic examination showed normal findings. Audiometry revealed mixed conductive and sensorineural hearing loss, more on the left side than on the right side. The patient's daughter was known to have otosclerosis. The patient was referred for a CT study of the temporal bones. The CT study showed a hypodense region in the labyrinthine capsule on both sides, just anterior to the oval window, abutting the vestibule. These lucent areas extended bilaterally, reaching the three turns of the cochlea. No other inner-ear abnormalities were seen. The middle-ear cavities, including the ossicles, appeared normal on both sides. The patient was diagnosed to have fenestral and cochlear otosclerosis. Surgery was performed on the left ear, and fixation of the stapedial footplate was noted. The treatment involved stapedectomy. A stapedial prosthesis (Schuknecht prosthesis) was inserted. Post-operative audiometry showed improved conductive hearing. If the left side responds to the treatment and shows signs of improvement, then stapedectomy of the right side will be performed later.
Discussion:

Fenestral otosclerosis usually occurs in young adults and is mostly a bilateral disease (in 85% of cases). Patients present with progressive conductive hearing loss and exhibit normal findings on otoscopic examination. Two-thirds of patients complain of tinnitus early in the course of the disease. Histologically, the enchondral bone is replaced by foci of a spongy new bone, later calcifying into a dense, ossific plaque. Fenestral otosclerosis typically starts in the labyrinthine capsule, just anterior to the oval window. The conductive hearing loss is caused by fixation of the stapedial footplate. Fenestral otosclerosis is treated with stapedectomy and insertion of a stapes prosthesis. The most common finding on CT in the early stage of fenestral otosclerosis is a lucent area just anterior to the stapedial footplate. Sometimes footplate thickening can be seen. Later, the oval window may be narrowed or even obliterated by calcified foci. Although the opposite ear may be asymptomatic, CT findings are mostly symmetrical. Cochlear (or retrofenestral) otosclerosis is much less common, but is always associated with fenestral otosclerosis. Patients present with mixed conductive and sensorineural hearing loss. The foci of the spongy new bone extend around the cochlea and sometimes even around the vestibule, the semicircular canals and the internal auditory canal. The sensorineural hearing loss is thought to be caused by cytotoxic enzyme diffusion into the fluid of the membranous labyrinth. Cochlear otosclerosis is treated with sodium fluoride or a cochlear implant. In cochlear otosclerosis, CT shows focal lucencies in the otic capsule, possibly extending in a ring-like manner around the cochlea. Later, in the sclerotic phase, these foci may undergo remineralisation and become indistinguishable from the normal otic capsule.

Differential Diagnosis List: Fenestral and cochlear otosclerosis.

Final Diagnosis: Fenestral and cochlear otosclerosis.

References:

Description: Axial CT image through the right temporal bone. A hypodense area is seen in the labyrinthine capsule (arrows), abutting the cochlear turns and vestibule, as well as the stapedial footplate (arrowhead). Origin:
Figure 2

*Description:* Axial CT image through the right temporal bone, a few millimetres caudal to that in Fig. 1. The hypodense area (arrow) is seen to extend caudally, between the basal and middle cochlear turns.

*Origin:*
Description: Axial CT image through the left temporal bone. Similar to the right side, a hypodense area is seen in the labyrinthine capsule (arrows). The stapedial footplate appears slightly thickened and hypodense (arrowhead). Origin: