Fenestral and cochlear/retrofenestral otosclerosis
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Section: Neuroradiology
Imaging Technique: CT
Case Type: Clinical Cases
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Patient: 60 years, male

Clinical History:

A 60-year-old male presented with a bilateral mixed hearing loss that was both conductive and sensorineural.

Imaging Findings:

A 60-year-old male presented to our ENT Department with a profound bilateral hearing loss, which was slowly progressive over several years. Audiometric tests that were done revealed a severe mixed (conductive and sensorineural) hearing loss. A high-resolution Computed Tomography (CT) of the temporal bone was performed, which showed diffuse demineralization of the otic capsule around the cochlea. A second osteolytic focus appeared anterior to the oval window. The anular ligament/stapes footplate was found to be thickened (see Fig. 1). The niche of the round window was also obliterated by an otosclerotic plaque (see Fig. 2) that was present. Both ears were involved. Based upon the clinical data and the CT findings, the diagnosis of diffuse fenestral and cochlear otosclerosis was established.

Discussion:

Otosclerosis is known to be an osseous dysplasia of the otic capsule. In Caucasians, it is found in 7% of male and 10% of female temporal bones. In only 12% of these cases, do the stapes get fixed accounting for clinical symptoms of conductive hearing loss. This represents circa 1% of the population. Other races have a lower incidence of otosclerosis. In the first phase of otosclerosis or otospongiosis, the dense, ivory-like osseous labyrinth is replaced with immature spongy, vascular foci of the haversian bone. In the subsequent stage of sclerosis, new ossification follows the demineralization. When the lesion is immature, it is easily detected as a hypoattenuating focus. Mature lesions are, quite difficult to recognize on CT because their density is too similar to that of the normal surrounding bone. Fenestral otosclerosis is the most common form of otosclerosis. The first site usually involved is the region of the fissula ante fenestram, an area located just anterior to the oval window. It is an embryonic rest of cartilage and connective tissue connecting the vestibular endosteal and tympanic periosteal layer. Involvement of the anular ligament/stapes footplate causes a conductive hearing loss by fixation of the stapedio-vestibular joint. Another region of interest in fenestral otosclerosis is the round window niche. Obliterative round window ossification is known to be a rare manifestation of advanced fenestral otosclerosis. In cochlear (retro-fenestral) otosclerosis, osteolysis occurs around the cochlea, appearing as a hypoattenuating halo (double-ring sign). An inflammatory reaction accompanies the demineralization. It is believed that the sensorineural hearing loss results from a diffusion of cytotoxic enzymes into the inner ear. Cochlear involvement rarely occurs without fenestral foci. In differential diagnosis, other primary bone diseases such as osteogenesis imperfecta, fibrous dysplasia and Paget's disease
Differential Diagnosis List: Diffuse fenestral and cochlear otosclerosis.

Final Diagnosis: Diffuse fenestral and cochlear otosclerosis.

References:


Description: A magnified axial CT image showing a large otospongiotic plaque in the anterior oval window region or fissula ante fenestram (indicated by a blue arrow) with obliteration of the oval window niche. The anterior crus of the stapes is also seen to be thickened. Note the diffuse demineralization of the otic capsule around the cochlea (indicated by a green arrow). Also note the presence of some debris/granulation tissue around the malleus and the incus. Origin:
Description: A magnified coronal CT image showing an otosclerotic plaque occluding the round window niche (indicated by a green arrow). Origin:
**Description:** A magnified axial CT image showing an otosclerotic plaque occluding the round window niche (indicated by green arrow). **Origin:**