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CAVITATING OTOSCLEROSIS

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Otosclerosis consists of one or several circumscribed foci of new, softer, and more vascular bone that replaces the remarkably avascular endochondrial bone of the adult labyrinthine capsule.¹ There is simultaneous and continuous resorption of perivascular bone (otospongiosis) and deposition of immature and collagen deficient bone (otosclerosis) within a focus.² It is an active remodeling process of bone that occurs within the endochondrial layer of the temporal bone, which under normal circumstances undergoes virtually no remodeling following development.³ Otosclerosis can occur independently at the stapes footplate, cochlear capsule, or both, which result in different clinical presentations and histopathological changes.

Otosclerosis is sometimes seen in other genetic bone disorders such as Paget's disease and osteogenesis imperfecta, which leaves many unanswered questions regarding the disease's genetic linkage and etiology. There is strong evidence that this is an autosomal dominant hereditary disorder with incomplete penetrance (roughly 40%), but this has been difficult to determine since approximately 90% of people with otosclerosis do not have clinical manifestations, and the percentage with isolated cochlear otosclerosis (CO) is considerably smaller.

CO is defined as a sensorineural hearing loss in the presence of, and presumably due to, an otosclerotic focus in the cochlear capsule, or an enzymatic impairment of the membranous structures of the cochlea.¹ CO is well known and has been described many times in the literature. However, we have found no mention of cavity formation in CO foci other than a single image recorded in Schuknecht's *Pathology of the Ear*.² Here we describe a case with a large cavity in a CO focus.

CASE REPORT

A 60-year-old patient who presented in 1967 with an 80dB hearing loss was diagnosed with otosclerosis of the stapedial footplate of the right ear and successfully treated with surgery. Her hearing acuity declined further, and 3 years later she was diagnosed with cochlear otosclerosis based on a polytome X-ray.

HISTOPATHOLOGY

The right temporal bone shows a large cavity (C) surrounding the middle and apical turns of the cochlea (Fig. 1). It is filled with a very fine fibrillary precipitate. The promontory is represented by a very thin layer of otosclerotic bone. The wall of the cochlea is extremely thin, in some places being merely a layer or two of cells. Adjacent to this area the spiral ligament is hyalinized. The rest of the lesion is a mixture of otosclerosis and spongiosis (Figs. 1 and 2, A and B, respectively). The spongiosis is quite active with high cellularity and multinucleated cells (arrow, Fig. 2).

The cavity extends superiorly to the level of the tensor tympani tendon while the otosclerosis extends superiorly thinning the tegmen. Below the level of the oval window, the cavity has eroded the bone of the promontory leaving just a thickened submucosa covering it.

References

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Fig. 1.
H&E, 10x. (A) otosclerosis. (B) otospongiosis. (C) cavity. (CA) internal carotid artery. (IAC) internal auditory canal. (S) stapes footplate.

Fig. 2.
H&E, 100x. (A) otosclerosis. (B) otospongiosis. (C) cavity. (arrow) multinucleated osteoclast.