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COCHLEAR OTOSCLEROSIS AND ENDOLYMPHATIC HYDROPS

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Otosclerosis is a progressive and dysplastic metabolic bone disease known to affect only human beings and can affect both the conductive and sensorineural pathways through changes in the middle and inner ear. It is a common cause of acquired hearing loss, characterized by resorption of perivascular bone that results in enlargement of the perivascular space, followed by deposition of immature and collagen deficient bone. These occur simultaneously and continuously within a focus.¹ Under light microscopy, the active stage where bone structure is being replaced by new bone can be distinguished from the inactive stage in which the bone becomes highly mineralized and thickened.

Endolymphatic hydrops, a histopathological finding related but not exclusive to Ménière's disease, refers to an increased volume of endolymph within the cochlear duct. It can be caused by trauma, infection, obstruction, or have an unknown cause. It can acutely alter hearing and balance through ruptures of Reissner's membrane and changes in endolymphatic electrolyte concentration, producing the triad of complications (fluctuating hearing loss, vertigo and tinnitus) of Ménière's disease.¹

There have been many studies assessing the relationship of these two diseases since 1955, when Cawthorne described the recurrence of vertiginous attacks in 24% of patients with otosclerosis, and it is now known that severe otosclerotic lesions involving the vestibular duct obstruct endolymphatic absorption, resulting in hydrops.^{2,3}

CASE REPORT

A 53-year-old patient with a history of bilateral conductive hearing loss and a successful fenestration operation on his left ear underwent a stapedectomy of his right ear. This was followed one year postoperatively by a sudden drop in hearing acuity in the right ear as well as imbalance.

HISTOPATHOLOGY

Histology of the right inner ear (Fig.1) shows a cochlea surrounded by active otospongiosis (AO) within an inactive otosclerotic focus (IO). The spiral ligament shows little to no hyalinization throughout the cochlea, and the stria vascularis (S) shows only mild to moderate atrophy in the apical segment and posterior turn of the middle segment.

Reissner's membrane is displaced towards the scala vestibuli, a sign of endolymphatic hydrops (H). The posterior basal and apical turns of the cochlea show evidence of rupture and secondary healing of Reissner's membrane, creating outpouches (arrow). The organ of Corti appears healthy, except in the apical turn where there is marked atrophy (arrow, Fig.2). The sensory epithelium of the utricle and saccule appear normal, even though the saccular membrane is dilated.

The endolymphatic duct and sac are patent with no surrounding otosclerosis, which in this particular case eliminates the possibility of an obstructive process.

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Fig. 1.
Active otospongiosis (AO), inactive otosclerosis (IO), endolymphatic hydrops (H), outpouches of Reissner's membrane (arrow), normal stria vascularis (S)

Fig. 2.
Active otospongiosis (AO), atrophic organ of Corti (arrow)