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Endolymphatic Hydrops and Blockage of the Endolymphatic Duct

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Endolymphatic hydrops, or dilation of the endolymphatic space, was first described independently in 1938 by Yamakawa (1) and Hallpike and Cairns (2) as the foremost pathologic correlate to Meniere's disease. Guild (3) had described the movement of endolymph toward the endolymphatic sac, noting that endolymph is formed by the stria vascularis, flows into the sacculus and through to the endolymphatic duct and sac, and exits the labyrinth by passing into vessels in the wall of the pars intermedia. Kimura and Schuknecht (4) later demonstrated that destruction of the endolymphatic sac in the guinea pig produced hydrops in both the cochlear and vestibular systems but not Meniere's disease. Here, we examine a case of endolymphatic hydrops without Meniere's disease.

CASE REPORT

The patient experienced hearing loss after an ear infection as a child. When first tested in the office, at the age of 67 years, there was a nonfluctuating sensorineural hearing loss of the right ear averaging 57 dB, with an ascending configuration.

Electronystagmography revealed a 67% reduced vestibular response on the right side. There was no spontaneous nystagmus. The patient experienced some disequilibrium later in life presumably due to the diminished function of the right labyrinth. She died at 96 years of age, with only a slight progression of her hearing loss bilaterally that is consistent with presbycusis.

Histopathology

In the right temporal bone, there is extensive hydrops that has prolapsed through the helicotrema as far into the cochlea as the middle turn of the scala tympani (Fig 1). In the basal segments of the cochlea, Reissner membrane is distended to the cochlear walls and obliterates the scala vestibuli perilymphatic space. The apical basilar membrane is depressed onto the interscalar septum. The saccule dilated and pressed against the footplate. There is hydrops of the utricle with some prolapse into the posterior limb of the horizontal canal. The endolymphatic duct is obstructed by new bone (Fig 2). Findings in the left ear were essentially normal for the patient's age.

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Temporal Bone Histopathology Case of the Month

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Fig.. 1.
Distended Reissner membrane (small arrows). Dilated saccular membrane (large arrows;
hematoxylin and eosin, $\times 20$).

Fig. 2.
Obliterated endolymphatic duct (hematoxylin and eosin, $\times 40$).