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The Endolymphatic Sac in the Mondini Disorder*

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Summary. The endolymphatic sacs are described in temporal bone specimens from a 31-year-old man with bilateral Mondini disorder. The ducts and sacs are thin-walled, cyst-like structures with complete absence of loose vascular perisac tissue, and are directly apposed to the bone of the vestibular aqueduct. Histological evidence of severe bone erosion is present in these specimens and is most marked in the intermediate and distal portions of the vestibular aqueduct. It is also present in the foveal region of the posterior temporal bone surface underlying the sac. Erosion of the bony wall of the paravestibular canaliculus (PVC) is demonstrable, with incorporation of the vein of the PVC inside the margin of the widened vestibular aqueduct. These findings suggest a causal relationship between pressure within the endolymphatic duct and sac and erosion of the surrounding bone. The absence of endolymphatic hydrops of the cochlea and vestibular organs in the Mondini disorder constrast significantly with the endolymphatic hydrops seen in Meniere's disease.

Key words: Mondini – Vestibular aqueduct – Endolymphatic sac – Endolymphatic hydrops

Introduction

In temporal bones with endolymphatic hydrops, the endolymphatic duct and sac are not distended. The only condition demonstrating enlargement of these structures is the Mondini disorder. It was therefore considered of interest to examine the endolymphatic duct and sac more closely in this latter disorder as a contrast to its appearance in Meniere's disease.

Case Report

The patient was a 31-year-old white man who was known to have the Mondini disorder bilaterally. Clinical and temporal bone findings have been published previously [3, 4] and only the endolymph system will be described in this report.

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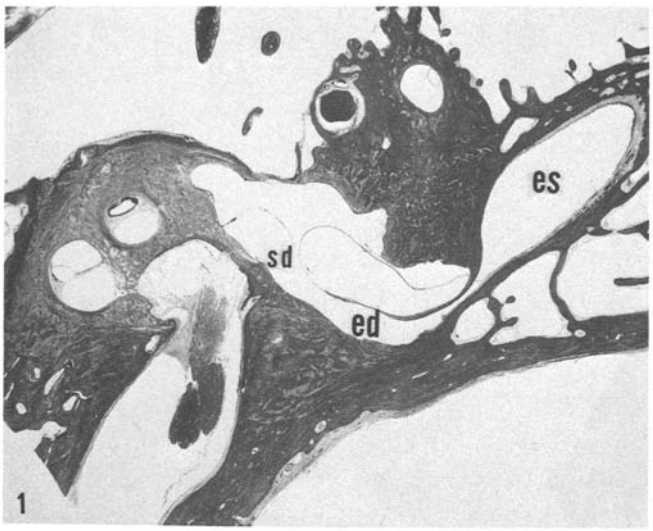

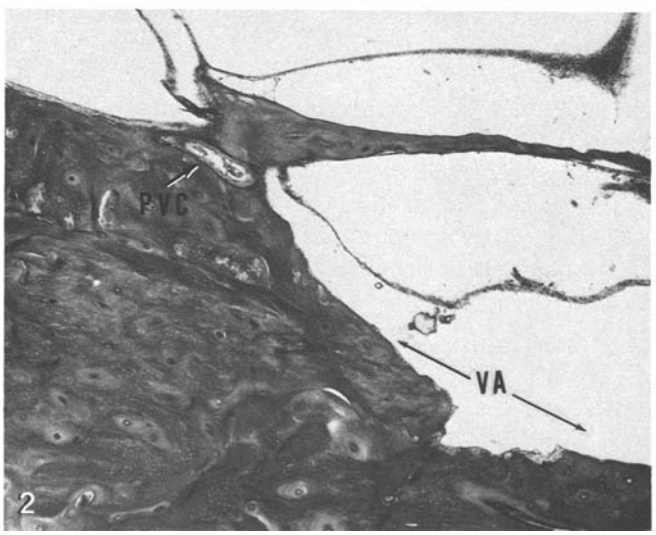

Fig. 1. Right side. Marked distension of endolymphatic duct and sac *(ed, es)* can be noted with loss of perisac tissue and erosion of vestibular aqueduct. The saccule duct *(sd)* is wide, but the saccule is normal. The cochlear duct is normal. H&E, × 4

Fig. 2. Right side. Intact para-vestibular canaliculus (PVC) with vein near vestibule. Periductal tissue is atrophic and there is focal erosion of the vestibular aqueduct *(VA)*. H&E, × 28.8

Fig. 3A–C. A Left side. Tangentially vertical section showing markedly enlarged endolymphatic duct and sac *(ed, es)*. There is deep erosion of the foveal region of the posterior surface and thinning and erosion of bone of the posterior semicircular canal. *Arrow* denotes erosion of PVC wall and vein of PVC incorporated into widened vestibular aqueduct. H&E, × 5.6. **B** Left side, high power. PVC is present with vein. Congested vein *(V)* is incorporated into the markedly eroded vestibular aqueduct. H&E, × 27.7. **C** Left side. Intact vein *(V)* of PVC is present in eroded wall of vestibular aqueduct distal to that seen **B**. H&E, × 64

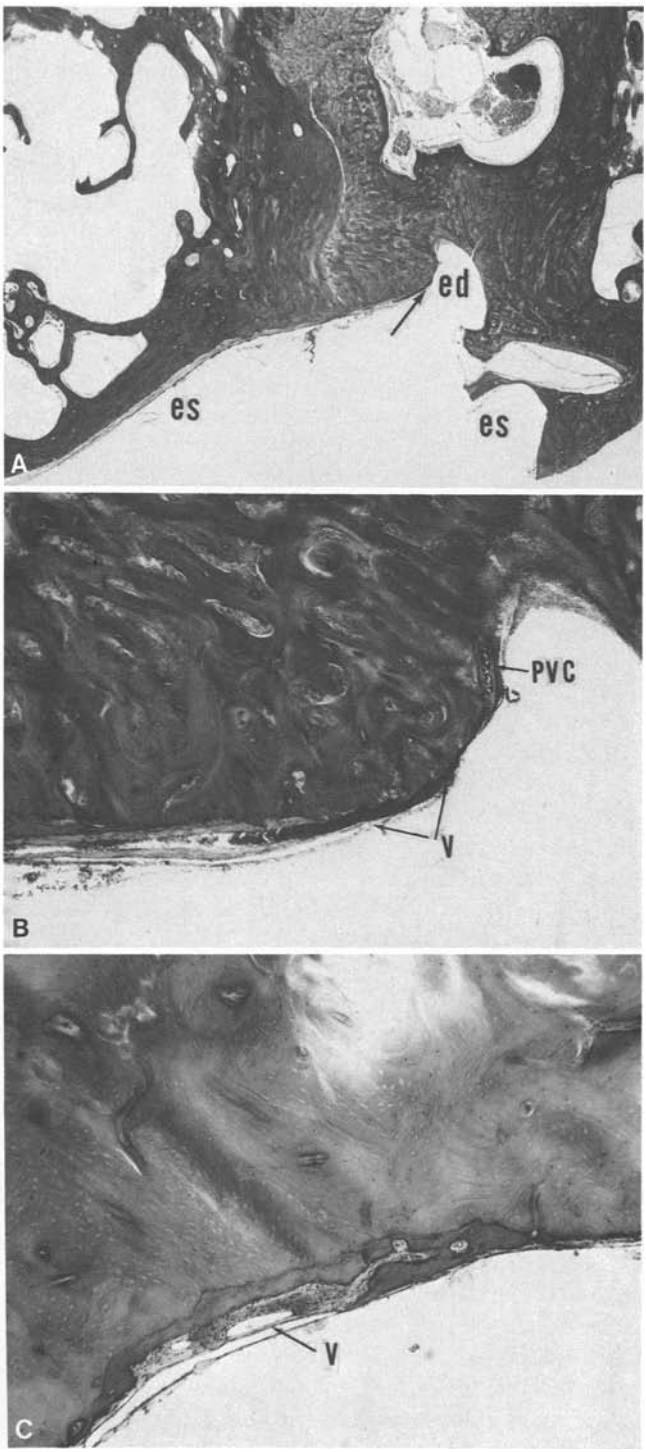

Fig. 3A–C.

Each endolymphatic duct and sac was very markedly enlarged, with complete loss of the loose, vascular periductal and perisac soft tissues throughout its length. The thin epithelial wall of the duct and sac was apposed to the bony margin of the vestibular aqueduct. The duct and sac had the appearance of a large thin-walled cystic structure with flattened epithelium and no rugal or papillary foldings (Figs. 1–3).

The vestibular aqueduct was markedly widened throughout its length. In some areas, especially in its intermediate and distal portions, this structure demonstrated uneven, serrated and deeply scalloped surfaces (Fig. 2). The bone of the adjacent paravestibular canaliculus (PVC) was completely eroded. As such, the vein of the PVC could be followed in its entirety, and was found to be incorporated just inside the widened vestibular aqueduct wall. Only at its origin at the vestibule was short, intact bony PVC present (Figs. 2 and 3).

The posterior foveal surface of the temporal bone underlying the distal enlarged endolymphatic sac was very deeply eroded, and this erosion also involved the bony wall of the posterior semicircular canal (Fig. 3A).

The cochlear duct was of normal caliber, and did not show evidence for endolymphatic hydrops. The saccule duct was wide, as a direct continuation from the distended endolymphatic duct, and the junction of the saccule duct with the saccule was also widened (Fig. 1). Proximal to this area, however, the saccule was of normal caliber, with no evidence of endolymphatic hydrops. The utricle and semicircular canals were normal.

Discussion

Although enlargement of the vestibular aqueduct has been noted from the earliest descriptions of the Mondini disorder, it was not until 1944 that Secrétan [9] described in some detail the histological appearance of the endolymphatic sac in this condition. He found the entire endolymphatic duct and sac to be dilated, with the enlargement most marked in the intradural region. In the intraosseous portion, the epithelial-lined wall of the sac was described as being against the bony margin of the vestibular aqueduct, with perisac tissue no longer present. From these studies, Secrétan believed that there was a defect in the endolymphatic duct and sac, whereby fluid resorption through the wall did not occur, with resultant dilatation of the duct and sac.

In the present case, the duct and sac are thin-walled. There is no loose vascular perisac tissue remaining, nor any identifiable histological pattern other than flat epithelium on a thin basement membrane type of wall. The structure most resembles a large cyst.

In the specimen, the large cyst-like structure of the endolymphatic duct and sac is apposed to a bony margin, which shows considerable histological evidence of erosion. This particularly involves the adjacent wall of the PVC, the intermediate and distal portions of the vestibular aqueduct, the opercular lip of the vestibular aqueduct posterior fossa opening, and the foveal region of the posterior surface of the temporal bone. The latter erodes deeply to involve the bony wall of the posterior semicircular canal.

Erosion of the bone about the endolymphatic duct and sac in the case described suggests a causal relationship of increased pressure within the duct and sac. The loss of loose vascular periductal and perisac tissue can be explained on the basis of pressure atrophy. The enlargement of the sac may be more pronounced into the dura since distension can occur more readily into an area of lesser resistance, such as the dura, where there is no bone. The incorporation

of the PVC vein into the widened vestibular aqueduct is further evidence of an eroding process.

It is very possible that an abnormality of the endolymphatic duct and sac can be present from earliest development, and that their enlargement begins before cartilage and bone are formed. However, evidence has been presented that the process continues beyond that stage with continuing enlargement and erosion of the surrounding vestibular aqueduct and posterior surface of the temporal bone.

The endolymph channels of the cochle, saccule, utricle, and semicircular canal ampullae were of normal caliber in the case described. The saccule duct was wide as a direct continuation from the distended endolymphatic duct. The entrance of the duct into the saccule at that point was widened, but the saccule was otherwise normal.

In other reported cases of the Mondini disorder, most have been without hydrops [6–9]. Certain of these cases have contained wide structures which are more in keeping with malformation than with hydrops [2]. In the few cases describing hydrops [1, 10], there was absence of membraneous portions of the cochlear wall or similar malformation of the saccule and utricle. In these specimens, it is possible that regions of endolymph resorption were compromised. Despite descriptions of hydrops, actual photographs from some of these cases have suggested retrograde widening. Such was seen in the present case.

It is a paradox that there is no endolymphatic sac distention in Meniere's disease, where the endolymphatic sac is generally thought to be abnormal, but there is hydrops of the saccule and cochlear duct. In contrast, endolymphatic sac distension in the Mondini disorder exerts pressure great enough to erode the bony vestibular aqueduct and enlarge the distal endolymhatic sac balloon-like into the posterior fossa. However, there is minimal or no retrograde pressure effect and the vestibular organs and cochlear duct are not hydropic.

In considering vascular mechanisms in Meniere's disease, the author has recently suggested [5] that insufficiency of the venous drainage of the vestibular organs may occur via the vein of the PVC. This can result in insufficient resorption of endolymph, with resultant endolymphatic hydrops if collateral venous drainage does not develop. In the present patient with the Mondini disorder, the PVC vein was preserved bilaterally, and there was no endolymphatic hydrops. One must consider the possibility that hydrops does not occur in this disorder because venous drainage of the vestibular organs is normal.

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