

Ménière's disease. Histopathological changes: A post mortem study on temporal bones

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Abstract. – The histopathological changes in the temporal bones of two deceased donors individuals with concomitant Ménière's disease have been studied. In one temporal bone we have found a blockage of the endolymphatic duct by abnormal bone. The histopathological modifications and the different therapeutic options are discussed. Clinical guidelines are proposed.

Key Words:

Ménière's disease, Hydrops, Endolymphatic duct.

Introduction

The pathology of the vestibular system in man has been even less adequately investigated than the auditory one. Table I lists conditions the pathological bases of which have been established by direct observation at autopsy or surgery; affections such as "vestibular neuronitis", in which the possible structural changes have been only guessed at from the clinical symptoms, are not considered.

Materials and Methods

We have studied the histopathological changes in the temporal bones of two deceased individuals with Ménière's disease.

These patients were donors and agreed during their life to donate post mortem their temporal bones to the House Ear Institute as a contribution to a better knowledge of temporal bone diseases.

We have removed the temporal bones in our usual way¹⁻³.

Results and Discussion

Hydrops and its causes

Ménière's disease is an affection of both the hearing and balance organs of the inner ear, characterized by episodes of vertigo, hearing loss and tinnitus. Its pathological basis is now firmly established as "hydrops" i.e. distention of the endolymphatic spaces of the labyrinth by fluid. The cause of the hydrops in Ménière's disease is unknown. There are, however, other diseases of known pathogenesis in which hydrops may be present as a complication. The common feature of these conditions is the presence of inflammatory or neoplastic involvement of the perilymphatic spaces. Thus otitis media complicated by perilymphatic labyrinthitis, syphilitic involvement of the labyrinth, or leukaemic deposits in the perilymph spaces, may be associated with hydrops⁴.

Pathological appearances of hydrops

The hydrops of Ménière's disease may affect one or both inner ears. In most cases the cochlear duct and saccule are involved, but the utricular and semicircular ducts are usually not. In some cases the cochlear duct alone is hydropic. A rare and debatable form of Ménière's is thought to affect the vestibule, but not the cochlea. Symptoms are those of vertigo, but not hearing loss.

In the hydropic cochlear duct Reissner's membrane, which is elastic, shows a variable degree of bulging. In the most severe cases, the membrane reaches the top of the scala vestibuli and may be in contact with a wide area of cochlear wall. In the apical region it may bulge to such an extent that it fills the helicotrema. In this way the distended scala media may even enter the scala tympani. The

Table I. Pathology of the vestibular system.

Malformations
Aging changes
Trauma
Ototoxicity
Infections: viral, bacterial, syphilis
Osseous lesions: Paget's disease, otosclerosis
Hydrops: Ménière's disease, syphilis, bacterial infection
Positional vertigo: atrophy of superior division of vestibular nerve; atrophy of utricle; "cupolithiasis"
Neoplasms: acoustic neuroma, metastatic carcinoma

sacculle swells up from its position on the medial wall of the vestibule and frequently touches the vestibular surface of the footplate of the stapes.

The utricle may be compressed in the process. In some cases the swollen sacculle may herniate from the vestibule into the semicircular canals. Less frequently the utricle may be distended, sometimes with small infoldings producing a scalloped appearance⁵.

Changes in walls of membranous labyrinth

Changes may be seen in the thin distended membranes of the hydropic endolymphatic spaces. Ruptures may be present particularly in Reissner's membrane, and the terminal end of the ruptured membrane may be curled up. Such ruptures have been incriminated as possible pathological basis of the fluctuations in pure tone thresholds which patients with Ménière's disease may suffer. It has been suggested that the flooding of the perilymph with endolymph with its high potassium level may inhibit the bioelectric activity of the cochlea⁶. It is likely, however, that most of these ruptures are artefactual. They may be found in non-hydropic labyrinths. They are often multiple in the same membranous labyrinth. Outpouchings are often seen in which dilatation of part of the wall of the membranous labyrinth takes place and a lining is present here that is thinner than elsewhere. These outpouchings have been explained as healed ruptures, but, because of their regular features, it is more likely that they are simply areas of increased distension of parts of the labyrinthine wall, which are normally thinner (Figure 1). The presence of

fibrous tissue in cases of Ménière's hydrops external to the endolymphatic space has been described in the scala vestibuli by Hallpike and Cairns⁷ and in the vestibule deep to the footplate of the stapes by Schuknecht⁶. It is possible that the foci of connective tissue in these two situations are reactions to the irritation produced by repeated distension and subsidence of the adjacent cochlear duct and saccule respectively.

Changes in vestibular aqueduct and endolymphatic duct

While hydrops involving the scala media and saccule is accepted by all as a basic feature of the pathology of Ménière's disease, there is no such unanimity with regard to the alterations in the endolymphatic duct and its surrounding vestibular aqueduct. There have been many descriptions of obstructive or potentially obstructive lesions of these structures associated with Ménières disease whereby restriction of the flow of endolymph may have caused the hydrops. The following list gives a brief indication of the lesions discussed at length in the literature:

- a) Fibrosis^{8,9}.
- b) Metastatic breast carcinoma¹⁰.
- c) Decreased vascularity¹¹.
- d) Partial atresia of the intermediate portion of the vestibular aqueduct with decreased amounts of endolymphatic duct tissue¹².
- e) Irregularity of the osseous wall of the vestibular aqueduct, sometimes with blockage of the orifice of the vestibule¹³.

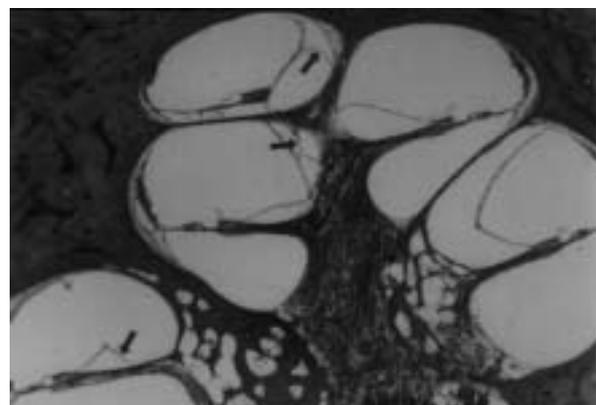


Figure 1. Meniere's disease. Ruptures of the hydropic membrane are represented by redundancies in the membrane (arrows). ×35

f) Blockage of the vestibular aqueduct by syphilitic microgummata – “perivascular round cell infiltrations”¹⁴.

g) Tumour-like papillary lesion resembling choroid plexus, which was removed surgically from the endolymphatic sac; no Ménière's symptoms were present in this case¹⁵.

Lesion similar to that mentioned under (g) were described in cases of Ménière's disease by Schuknecht within the ductus reuniens region of the hydropic cochlear duct⁶, by Gussen⁹ in the hydropic crus commune and by Hassard et al.¹⁶ at operation for possible insertion of an endolymphatic shunt. It is possible that such structures may secrete an excess of endolymph and so produce lesions and symptoms of hydrops. In contrast to these observations it must be pointed out that in many careful studies of some Ménière's hydropic temporal bones no changes whatever have been noted in the endolymphatic duct or vestibular aqueduct¹⁷.

Ultrastructural alterations of a degenerative nature have been observed in the epithelial cells of the endolymphatic sac in cases of Ménière's disease¹⁸, but similar changes were also seen in cases of acoustic neuroma, suggesting that they are the result of the disease process and not its cause.

We have found in a temporal bone endolymphatic hydrops due to a blockage of the endolymphatic duct by abnormal bone (Figure 2).

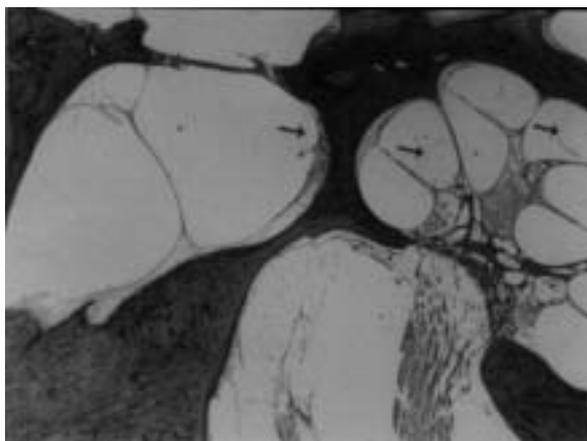


Figure 2. Endolymphatic hydrops due to a blockage of the endolymphatic by abnormal bone (*white arrow*). The saccular and Reissner's membranes are distended (*black arrows*). $\times 15$

Changes in the sensory epithelia of the labyrinth

Alterations of the sensory cells of the organ of Corti are mentioned by Hallpike and Cairns⁷, but they warn that such changes may be the result of post-mortem autolysis. The effect of acid used in decalcification of the temporal bone is another possible source of damage to these cells after death. Such possibilities of artefacts have been ignored in some reports. Changes, particularly in the apical region, have been described and associated with low frequency hearing loss⁶. In a recent study of 23 temporal bones from 17 patients with Ménière's disease, however, no direct correlation was found between endolymphatic hydrops and hair cell loss¹⁴. Atrophy of the macula of the saccule may also be found, which does not appear to be artefactual.

Relationship of symptoms to pathological changes

Image analysis of the areas in histological section of the cochlear duct (corresponding to volume in the whole structure) has been carried out in two studies and related to the hearing loss. In the study of Antoli-Candela¹⁹ the area of the cochlear duct was significantly increased in relation to the degree of hearing loss. Losses of over 70 dB showed a particularly high degree of hydropic expansion. In the study of Fraysse et al.¹⁴ a similar relationship was found between cochlear duct size and the total average hearing loss. There was also a correlation of those dimensions with the duration of the disease: the longer the history of symptoms the more pronounced the cochlear duct dilatation. A relationship also seemed to be present between (a) the amount of dilatation of vestibular structures and (b) the response to caloric tests and the presence of positional nystagmus, but this was less definite than the cochlear duct/hearing loss association.

Pathogenesis

Ablation of the endolymphatic sac in guinea-pigs results in endolymphatic hydrops within 3 months. In cats the same result can only be attained with survival times of 6 months to 3 years²⁰. In humans, operations for drainage of endolymph into the subarachnoid space are sometimes of value in the treatment of Ménière's disease. These facts suggest that obstruction of the endolymphatic

duct may play a part in the pathogenesis of Ménière's disease. Endolymphatic duct obstruction is sometimes, but not consistently, present on histopathological examination of serial sections of temporal bones in cases of hydrops. It is possible that studies of further cases at autopsy using modern techniques such as X-ray of microsliced bones, histochemistry and electron microscopy may yet establish a definite lesion of the endolymphatic duct as the cause of the hydrops.

Conclusions

There is histopathological evidence that the saccule in Ménière's disease may swell up from its position on the medial wall of the vestibule and touch the vestibular surface of the footplate of the stapes, causing a transmissive hearing loss, thus mimicking an otosclerosis. We suggest that in patients suspected of otosclerosis, a history of vertigo in recent months be considered as a contraindication to stapes surgery because it can indicate the probability of endolymphatic hydrops, with the risk of further cochlear loss should the labyrinth be opened.

Brackmann states: "Endolymphatic sac (ES) mastoid shunt is my procedure of choice in early Ménière's disease"²¹. There is considerable controversy about the physiological basis for endolymphatic sac surgery. There is evidence that the ES absorbs endolymph. The rationale for an endolymphatic shunt is to drain excess endolymph into the mastoid or spinal fluid. It has been proposed that an endolymphatic mastoid shunt with silastic sheeting works by enlargement of the lumen of the endolymphatic sac, removal of bony pressure, improvement of blood supply and enlargement of the absorptive area of sac.

In our opinion the finding in our temporal bones of an endolymphatic hydrops due to a blockage of the endolymphatic duct by abnormal bone (Figure 2) indicates that no benefit can result shunting the excess of endolymph through the endolymphatic sac. In fact in these cases the endolymphatic hydrops forms in the membranes of the vestibular system and do not pass through the endolymphatic duct, which is obstructed, and do not reach the ES.

Contrast enhanced Magnetic Resonance is required in case of Ménière disease to assess the degree of involvement of the membranous system. In case of reduction of the endolymphatic duct, no shunting procedure should be attempted. In these cases, the gold standard for the control of vertigo remains in our opinion the vestibular nerve section, which provide relief of vertigo in more than 90% of these patients.

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