Obstructing lesions of the endolymphatic sac and duct mimicking Ménière’s disease

Ryan C. Cmejrek, MD; Cliff A. Megerian, MD

Abstract
In this retrospective case series and literature review, we demonstrate that temporal bone lesions that obstruct the endolymphatic sac or duct can cause symptoms of Ménière’s disease. This finding is likely attributable to endolymphatic hydrops; initially, such cases typically masquerade as Ménière’s disease. Between July 1995 and April 2002, a total of 379 patients were treated for an initial diagnosis of Ménière’s disease at our institution. Among this group, 3 patients were found to have an obstructing lesion of the endolymphatic sac or duct that we felt was causally related to their Ménière’s-like symptomatology. We reviewed these cases and noted the similarities in each patient’s presentation, including a common pathophysiology. On imaging studies, each patient had a different pathologic lesion that involved the endolymphatic sac or duct: patient 1 had a jugular megabulb, and she was ultimately treated with vestibular nerve section; patient 2 had a cholesterol granuloma, which was treated with surgical excision; patient 3 had an endolymphatic sac tumor that was treated with surgical excision. As has been suggested in previous reports, not all cases of Ménière’s disease are idiopathic. We conclude that obstruction of the endolymphatic sac or vestibular aqueduct by a mass lesion or vascular anomaly can lead to vestibulocochlear pathology that mimics Ménière’s disease.

Introduction
Ménière’s disease is defined as the idiopathic syndrome of endolymphatic hydrops that results in hearing loss, tinnitus, and episodic vertigo, and aural fullness. However, there is continuing controversy in the literature regarding the underlying pathophysiology of Ménière’s disease, and most authors agree that not all cases are idiopathic. Recent reports have suggested many possible etiologies, including autoimmune disease, viral infection, allergy, and otosclerosis. In any event, impaired absorption or overproduction of endolymph is presumed to cause endolymphatic hydrops and associated Ménière’s symptomatology. Similarly, vascular compression of the endolymphatic sac and duct has been shown to be associated with Ménière’s symptomatology in the presence of anatomic variants such as jugular megabulb and an anomalous vein of the vestibular aqueduct.

Between July 1995 and April 2002, a total of 379 patients had been seen in the otology–neurotology clinic with a diagnosis of Ménière’s disease. In addition to the typical medical and audiologic workup that was administered to all patients, those with unilateral hearing loss underwent imaging studies in order to rule out retrocochlear disease.

In this article, we describe the cases of 3 patients who initially presented with fluctuating sensorineural hearing loss, tinnitus, episodic vertigo, and aural fullness and who were initially diagnosed with Ménière’s disease. Further evaluation and diagnostic imaging revealed that each had a mass lesion of the temporal bone that had obstructed the endolymphatic system. Patients 1 and 2 exhibited compression and erosion of the endolymphatic sac and duct by a jugular megabulb and a cholesterol granuloma, respectively. Patient 3 had two endolymphatic sac tumors that led to an internal obstruction of the sac bilaterally.

We propose that any abnormal space-occupying lesion of the temporal bone has the potential to cause endolymphatic hydrops by obstructing the endolymphatic duct or sac.

Case reports
Patient 1. A 35-year-old woman presented with a 9-year history of Ménière’s disease; she had a right-sided sensorineural hearing loss that had been sudden in onset and slowly progressive over the years. The hearing loss was accompanied by fluctuating nonpulsatile tinnitus, episodes of vertigo that initially occurred a few times monthly, and...
Aural fullness. Her symptoms had begun during a pregnancy, and they were presumed to be of viral etiology. She had sought intervention when her episodes of vertigo began to occur almost daily. An audiogram showed essentially normal hearing in the left ear and a sloping mild-to-profound sensorineural hearing loss in the right ear (figure 1, A). Her speech discrimination score on the right was 0%. Magnetic resonance imaging (MRI) was negative for retrocochlear pathology.

After 3 years of conservative medical therapy (a low-salt diet and a diuretic), the patient returned with worsening vertigo and drop attacks. Findings on electronystagmography were normal. However, computed tomography (CT) of the temporal bones detected an enlargement of the right jugular bulb with encroachment on the endolymphatic sac and duct (figure 1, B). The patient underwent an endolymphatic shunt procedure, but her vertigo actually worsened and was unresponsive to gentamicin therapy delivered to the middle ear. Labyrinthectomy was recommended, but the patient was hesitant to sacrifice the slight degree of auditory perception that remained in her right ear. Instead, she underwent a middle fossa vestibular nerve section. At the 4-year follow-up, she remained free of disabling vertigo.

**Patient 2.** A 45-year-old woman presented with a diagnosis of Ménière’s disease and a long history of fluctuating hearing loss and tinnitus on the left. She reported a 7-year history of episodic vertigo that had worsened over the preceding 3 years; she also complained that sound was distorted. She sought an otologic evaluation when she began to experience an increasing feeling of pressure and pain deep in her left ear.

An audiogram revealed a mild high-frequency sensorineural hearing loss on the right and a mild-to-moderate low-frequency sensorineural hearing loss on the left (figure 2, A). MRI detected a complex mass on the left petrous bone that extended to the posterior fossa and obstructed the endolymphatic sac (figure 2, B). A slight dural erosion was noted, but the internal auditory canal and cochlea were not involved. Electrocochleography revealed a summating-potential to action-potential (SP/AP) ratio of 0.56 on the left and 0.15 on the right, findings that are consistent with endolymphatic hydrops of the left ear.

The patient underwent a transtemporal resection of the lesion and dural reconstruction. The mass was not only locally invasive, it had invaded the endolymphatic sac and duct, as well. Final pathology was consistent with a cholesterol granuloma. Postoperatively, the patient’s hearing declined slightly, but her vertigo had completely resolved.

**Patient 3.** A 37-year-old woman with von Hippel-Lindau disease presented with a diagnosis of Ménière’s disease that manifested as hearing loss and tinnitus on the left over several years and a recent increase in the frequency of vertigo attacks to one a day. These symptoms had been unresponsive to diet, diuretics, and other medications. The patient had no other medical problems related to her von Hippel-Lindau disease, and she was taking no medications. Findings on physical examination were significant only for a Weber test that lateralized to the right ear at 512 Hz. An audiogram showed an upsloping sensorineural hearing loss in the left ear (figure 3, A). Her speech discrimination score on the left was 72%. MRI detected a left temporal bone lesion posterior to the posterior semicircular canal and a similar, smaller lesion on the right (figure 3, B).

The patient underwent a left mastoidectomy and exploration of the left endolymphatic sac with tumor resection and...
preservation of the labyrinth and dura. Final pathology was consistent with an endolymphatic sac tumor (previously known as an aggressive papillary tumor). Postoperatively, her vertigo resolved and her hearing improved. Although electronystagmography 6 months postoperatively showed a residual peripheral deficit on the left, the vertigo remained absent.

Over the course of 3 years, the patient’s speech discrimination score fell to 0% in the left ear despite the absence of tumor regrowth on imaging studies. Electrocochleography showed an increase in the SP/AP ratio in both ears (right: 0.48; left: 0.53), and she began noticing a fluctuation in the hearing in her right ear and a return of the vertigo. There was also a subtle increase in the size of her rightsided endolymphatic sac tumor. She elected to continue medical management.

A noteworthy aspect of this case is that it was included in an earlier series describing hearing preservation surgery in von Hippel-Lindau disease–related endolymphatic sac tumors. Since that report was written, new electrocochleographic data presented herein further support the hypothesis that the affected ears are indeed hydropic.

Figure 2. Patient 2. A: Preoperative audiogram reflects the mild high-frequency sensorineural hearing loss on the right and the mild-to-moderate low-frequency sensorineural hearing loss on the left. Hearing levels for bone conduction approximated those of air conduction (±10 dB). B: T2-weighted axial MRI shows the cholesterol granuloma on the left petrous apex (arrowhead).

Figure 3. Patient 3. A: Preoperative audiogram demonstrates normal hearing on the right and an upsloping mild-to-moderate sensorineural hearing loss on the left. Hearing levels for bone conduction approximated those of air conduction (±10 dB). B: T1-weighted contrast-enhanced axial MRI shows the enhancing lesion of the endolymphatic sac and duct on the left and a smaller, similar lesion on the right.
Discussion

The role of endolymphatic hydrops in Ménière’s disease has been a topic of much discussion in the otorhinolaryngology literature since it was first described by Hallpike and Cairns in 1938. To date, the most reliable animal model of hydrops has been the guinea pig, the model initially described by Kimura and Schuknecht. However, studies of endolymphatic duct obstruction in primates have not strongly supported its role as a principal inciting pathologic entity in Ménière’s disease in humans. The limitations of detecting hydrops in living patients has made it difficult to definitively establish a cause-and-effect relationship between endolymphatic obstruction, Ménière’s disease, and its proposed histopathologic correlate, endolymphatic hydrops.

Our findings, combined with those of Jahrsdoerfer et al and Hosseinzadeh et al, support the concept that obstruction of the endolymphatic sac and duct by a number of causes (e.g., a tumor, vascular anomaly, or internal sac disease) can produce symptomatic Ménière’s-like disease, likely via the production of endolymphatic hydrops, as has been seen in the guinea pig. This concept is supported by the presence of the electrocochleographic abnormalities in 2 of our patients and the relief of their vertigo symptoms by surgical decompression or VIIIth nerve section.

Our findings and those of temporal bone histologic studies support the concept that even microscopic violation of the endolymphatic duct can potentially incite hydrops. Yoon et al were able to show that endolymphatic hydrops and otosclerotic obstruction of the vestibular aqueduct can coexist. Rizvi and Gibbin demonstrated that vestibular aqueduct obstruction secondary to fracture-induced osseousification also has the potential to produce hydrops. These findings support the concept that any disturbance of longitudinal outflow of endolymph can lead to the development of endolymphatic hydrops and Ménière’s symptomatology.

The cause of idiopathic Ménière’s disease in the absence of obvious endolymphatic sac or duct obstruction remains unclear. Data on the underabsorption or overproduction of endolymphatic fluid are still being accumulated. Clinical data presented in our report and others indicate that sac and duct malfunction by reason of gross obstruction appears to be an important contributing cause of hydrops production in some cases. Hence, the evidence indicates that a variety of lesions that cause obstruction of the endolymphatic sac by external or internal luminal obstruction can lead to clinical Ménière’s disease symptomatology.

Our 3 case reports provide further evidence to support the idea that endolymphatic hydrops represents the final common pathway in the production of Ménière’s symptomatology. However, they also underscore the fact that Ménière’s disease is a diagnosis of exclusion. Careful physical and radiologic assessment of each patient is warranted even when classic Ménière’s disease seems to be the most likely diagnosis. Further research into the underlying mechanisms of endolymphatic hydrops will likely lead to refinement of our current understanding of the link between hydrops and Ménière’s disease in the coming years.

References