MENIERE'S DISEASE

Robert W. Baloh, MD
University of California at Los Angeles

Case illustration. A 43 years old woman presented with recurrent episodes of vertigo lasting 3 to 4 hours. These episodes were not positional but continue no matter what she did. The vertigo attacks were accompanied by nausea and vomiting and by decreased hearing, fullness and tinnitus (a roaring sound) in her left ear ear. Over time the hearing in that ear progressively got worse.

Historical perspective. Prosper Meniere was a French physician who while working at a deaf mute Institute in Paris was struck by the observation that vertigo and hearing loss often occurred together. In his initial report before the Imperial Academy of Medicine in 1861 he described a young man who had violent attacks of vertigo, nausea and vomiting without any apparent cause. Later the young man noticed loud noises in the ears along with decreased hearing.

Did this young man described by Meniere actually have what has become known as Meniere's disease. The modern diagnostic criteria for Meniere's disease include attacks of vertigo with one sided ear fullness, roaring tinnitus and hearing loss typically lasting hours. The key to the diagnosis is to document a decrease in hearing in the involved ear during an attack and a gradual progressive hearing loss between attacks. The young man described by Meniere had vertigo attacks followed by tinnitus and fluctuating hearing loss but apparently the hearing loss involved both ears. The hearing loss with Meniere's disease typically begins in one ear and remains so for many years and only a small percentage of patients go on to develop involvement of both ears after many years. It is possible that the young man had some other inner ear disorder such as autoimmune inner ear disease but Meniere provided too few details to be sure.

One problem that plagued Meniere and the other physicians who wanted to study the inner ear at autopsy in the mid 19th century was the denseness of the bone that surrounds the inner ear. They could not make thin cuts through the tissue of the inner ear as was typically done to study other organs of the body. A German Otolaryngologist Karl Wittmaack solved this problem and revolutionized the study of the inner ear diseases in the early 1920's. At autopsy he removed a cylinder of bone containing the inner ear and slowly decalcified the bone with 1% nitric acid over several months. Once x-ray analysis showed the bone was completely decalcified he imbedded the tissue in cellulose and then made very thin serial cuts. Physicians from all around the world visited his laboratory to learn the technique including a young Otolaryngologist from England, Charles Hallpike.

In December of 1934, a 63 year old man was referred to the department of neurosurgery at the London Hospital to have his vestibular nerve cut in an attempt to stop his debilitating attacks of vertigo caused by Meniere's disease. Along with the vertigo he had tinnitus and progressive hearing loss in the left ear. At that time it was common to group all inner ear causes of vertigo under the label of Meniere's disease. Whether there was a Meniere's disease with specific pathology or whether there were many diseases that caused the same combination of symptoms was unknown. Regardless, in this case it was reasonably clear that the symptoms were originating from the left side and prior cases had been successfully cured of vertigo by cutting the vestibular nerve.

The operation was performed by Dr. Hugh Cairns, one of the few neurosurgeons in the world able to perform such a delicate procedure. The surgery on the man was performed on December 18, 1934. Cairns noted that the opening was smaller than usual because of the thickness and hardness of the bone but he was able to cut the left vestibular nerve. The patient had unusually high blood pressure during the operation and that evening he became restless and then unresponsive. Despite heroic efforts to relieve pressure in the brain with a second operation the patient died 3 days later. An autopsy performed 6 hours after death showed massive bleeding into the brain. Cairns cut out the inner ears in their bony capsule with a saw and placed them in storage in formaldehyde solution.

The bone pieces containing the inner ears of the man sat in formaldehyde at the London Hospital for more than a year before Cairns became aware of Hallpike's interest in inner ear pathology. The decalcification process took about 6 months and finally Hallpike was able to cut the specimens and examine the tissue under a microscope. He observed a prominent distention of the entire inner ear sac – hydrops. Hallpike and Cairns published what they felt was the first description of the pathology in Meniere's disease in October of 1939. Unknown to them, a Japanese otolaryngologist Kyoshiro Yamakawa reported similar findings of hydrops in a

patient with Meniere's disease who died of pneumonia. Yamakawa, who also studied with Wittmaack in Germany, initially presented the findings at a medical congress in Kyoto, Japan in April of 1938 and then in a short article in German later in the year.

So by the 1940's there was general consensus on the clinical features and the pathology of Meniere's disease but little was known about the mechanism for producing hydrops and how hydrops causes the typical attacks. Subsequent autopsy studies in patients with Meniere's disease have reliably found hydrops but occasionally hydrops has been found in patients without typical symptoms of Meniere's disease. Assuming increased fluid pressure in the inner ear is the cause of Meniere's disease clinicians have tried many ways to lower the inner ear pressure. A possible relationship between migraine and Meniere's disease was first suggested by Meniere when he noted that many of his patients with vertigo and hearing loss also had migraine. Subsequently there have been multiple reports indicating that migraine is more common in patients with Meniere's disease than in the general population. The nature of this relationship is poorly understood, however. Does migraine lead to hydrops or does it simply mimic Meniere's symptoms? Likely migraine is just one of several risk factors for developing Meniere's disease.

Symptoms of Meniere's disease. Meniere's disease is a syndrome, defined by a combination of symptoms and findings. Attacks typically begin with a sensation of fullness and pressure along with decreased hearing and roaring tinnitus in one ear. Vertigo rapidly follows reaching maximum intensity within minutes and then gradually resolving over hours. In the early stages the one sided hearing loss is reversible between attacks but over time the hearing loss continues between attacks. The tinnitus that is often compared to the sound of the ocean or a seashell next to the ear can be constant but usually becomes louder with an attack. Nausea and vomiting typically accompany the vertigo and balance is impaired. As noted above, variations in this typical pattern of symptoms is not uncommon. Isolated attacks of vertigo or hearing loss can precede the characteristic combination of symptoms for months and rarely years. With one variant called delayed Meniere's disease episodes of vertigo develop many years after the patient has experienced a sudden one sided deafness.

Diagnosis of Meniere's disease. By far, the most important test is an audiogram. The diagnosis of Meniere's disease rests on finding a characteristic hearing loss in the low frequencies. Other audiometric tests such as electrocochleography (ECog) and otoacoustic emissions may be abnormal but there are too many false positives and false negatives to be reliable. Vestibular tests such as VNG and ENG can identify damage to the vestibular part of the inner ear and are most helpful when considering surgical options.

What is the role of MRI in diagnosing Meniere's disease? At present its main role is to rule out other causes of a one sided hearing loss such as a benign tumor of the vestibular nerve called an acoustic neuroma or vestibular schwannoma. These benign tumors rarely cause vertigo but typically present with a gradually progressive one side hearing loss. If they grow inward they might damage the inner ear and mimic Meniere's disease. When an MRI is done to rule out an acoustic neuroma it must be done with contrast (a dye injection) since these small tumors can be missed without contrast enhancement. Current MRIs do not visualize the tiny inner ear well enough to see hydrops but new techniques with a special coil near the ear are being developed to improve resolution of the images. Preliminary studies at UCLA have shown that resolution of hydrops on MRI correlates with improvement in clinical symptoms. MRI could revolutionize both the diagnosis and treatment of Meniere's disease. It would provide a "gold standard" to evaluate and compare different treatments.

Treatment for Meniere's disease? The first line of treatment for Meniere's disease is a medication to suppress symptoms at the time of an attack. Symptomatic treatments such as meclizine and diazapam do not prevent attacks but rather "take the edge off" of attacks. These medications should be taken at the earliest sign of onset before severe nausea and vomiting occur. Once vomiting occurs medications can be given via suppositories or injection since oral medications will not be absorbed. Salt restriction diets and diuretics were the earliest treatments and continue to be the most prescribed treatments for preventing Meniere's attacks. Clearly, some patients with Meniere's disease are salt sensitive but this is a subset so that severe prolonged salt restriction in all patients is unwarranted. However, a trial of salt restriction should be the first line of treatment for Meniere's disease. The effectiveness of diuretics has never been proven in adequately controlled treatment trials but most clinicians feel these medications are effective at least in some patients. Hydrochlorthiazide or acetazolamide are recommend for a 3 to 6 months course before deciding on efficacy. Since migraine is common in patient's with Meniere's disease acetazolamide has the added benefit of treating both migraine and Meniere's disease.

Based on the finding of hydrops in patients with Meniere's disease, surgeons have tried a wide range of procedures to drain fluid from the inner ear (endolyphatic shunt surgery) but these procedures have largely been abandoned because of the technical difficulty of maintaining a functioning drainage tube in such a small fluid

compartment (about 1 cc of fluid). Assuming there is an inflammatory component to Meniere's disease physicians have injected steroids into the middle ear through the ear drum where the medication can make its way into the inner ear. There is currently a controlled treatment trial underway to see if this treatment works but it will be awhile before we know the results. Cutting the vestibular nerve is a reliable way to stop the vertigo attacks with Meniere's disease since the abnormal signals set off by the hydrops can not be passed on to the brain. Surgical techniques have improved since the time of Cairns so major complications are rare. Of course this surgery does not treat hydrops so the hearing loss and ear noises continue to progress even though vertigo stops. Most patients will compensate for the loss of vestibular function on one side but it can take months for the brain to "rewire" so that balance returns to normal. Rare patients compensate poorly after surgery and become chronically dizzy. A less invasive way to obtain the same result is to inject gentamicin into the middle ear where it slowly crosses into the inner ear. Gentamicin is selectively toxic to the vestibular part of the inner ear. This is a popular procedure in Europe where studies have found that about 50% of patients will obtain good results with a single injection. Some require multiple injections and some are not helped (presumably because the drug does not make its way into the inner ear).

Differential diagnosis of Meniere's disease. Any disorder that causes a combination of vertigo and hearing loss can mimic Meniere's disease. Autoimmune inner ear disease can start with vertigo attacks and fluctuating hearing loss but the course of symptoms is much more aggressive than with typical Meniere's disease with progressive severe hearing loss on both sides within the first few months. Typically the hearing loss will respond dramatically to steroids or other immune suppression. Autoimmune inner ear disease can be part of a systemic autoimmune disorder such as lupus or rheumatoid arthritis or it can selectively involve the inner ear. Migraine can mimic Meniere's disease and in a very small subset may even cause Meniere's disease. Chronic infections of the inner ear, including latent syphilis can also mimic Meniere's disease but as with autoimmune inner ear disease the course is much more fulminant than with typical Meniere's disease.

References

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