



Symptoms: Sound- and Pressure-Induced Vertigo

By Hamid R. Djalilian, MD

50-year-old man is seen for the complaints of hearing loss and dizziness. Loud noises make him dizzy, as does forcefully pressing his right ear with the palm of his hand, he said.

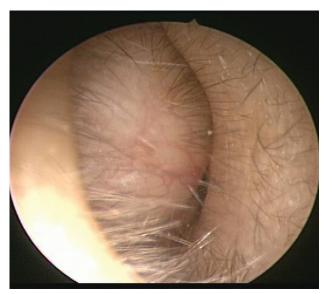
The patient notices that his hearing is a bit muffled on the right side as well. When he tries to put a cotton swab in his ear, it doesn't go in easily.

His history is significant for a tympanoplasty 15 years prior to presentation. The examination of the patient's ear is shown in the image on the right.

What is your diagnosis? See p. 16.

Read past Clinical Consultation columns in a special collection on the *HJ* website: bit.ly/HJClinicalConsultation.

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This picture shows the examination of our patient's right ear canal.

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Diagnosis: Cholesteatoma Causing Posterior Canal Dehiscence

By Hamid R. Djalilian, MD

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he complaint of sound- and pressure-induced vertigo should make the clinician think of three clinical entities: perilymphatic fistula, semicircular canal dehiscence, and a migraine-related phenomenon, which becomes the diagnosis of exclusion.

In perilymphatic fistula, intermittent leakage of perilymph occurs through the round or oval window. This condition generally occurs after direct physical trauma to the ear or head, or barotrauma.

Typically, patients with barotrauma will have symptoms after a significant pressure change on the tympanic membrane, which commonly occurs with Eustachian tube dysfunction. For example, the patient could have trouble clearing the ears on descent when scuba diving, or the patient could have an upper respiratory infection and difficulty equalizing pressure in the ears upon descent in an airplane.

Perilymphatic fistula can also occur from an increase in central venous pressure. For instance, when a patient lifts a very heavy object, holding the breath and exerting significant force, intracranial pressure can rise, leading to higher perilymphatic pressure and the occurrence of a fistula.

In semicircular canal dehiscence, the bone overlying the membranous semicircular canals erodes. This erosion can occur spontaneously, which is most commonly seen in the superior semicircular canal, followed by the posterior semicircular canal. It can also result from a cholesteatoma—most often in the horizontal semicircular canal, then in the superior semicircular canal, and, least commonly, the posterior semicircular canal.

The etiology of superior semicircular canal dehiscence has not been fully elucidated. An increase in intracranial pressure has been thought to play a role in the condition.

Posterior semicircular canal dehiscence usually happens as a result of a high jugular bulb or the exposure of the canal to the posterior fossa dura. Horizontal canal dehiscence primarily occurs as a result of an acquired cholesteatoma.

A cholesteatoma starting in the pars flaccida will gradually expand into the antrum and, from there, the horizontal semicircular canal. Damage to the horizontal semicircular canal usually results from the expansile force as well as the osteolytic molecules, which destroy bone, or the surrounding granulation tissue, which is caused by the inflammatory reaction to the cholesteatoma.

As the cholesteatoma expands in the mastoid, it can involve the superior semicircular canal, which is a few millimeters away, above the horizontal canal. A cholesteatoma that starts in the attic region rarely affects the posterior semicircular canal.

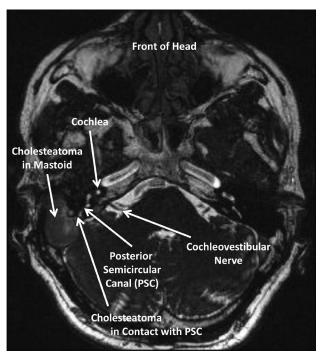


Figure 1. This axial CISS MRI of our patient shows a 3-cm mass in the mastoid that is abutting the posterior semicircular canal.

CHOLESTEATOMA CAUSED BY SURGEON

In our patient, we first obtained a CT scan to evaluate the mass that was bulging into the ear canal. The imaging showed an expansile mass that had destroyed much of the mastoid and the bone overlying the posterior cranial fossa and dura.

As the mass had continued to expand, it destroyed the bone overlying the posterior semicircular canal, leading to posterior canal dehiscence and the patient's symptoms. An MRI confirmed that the mass was a cholesteatoma and not a different tumor (see figures 1 to 3). A cholesteatoma will appear bright on diffusion-weighted imaging, indicating the presence of dead cells—in this case, dead skin cells.

This type of cholesteatoma generally results from the turning under of the skin of the lateral external auditory canal at the end of an operation. During a tympanoplasty, an incision is made in the ear canal, and the medial and lateral skin flaps are elevated.

At the end of the operation, these two skin flaps need to be straightened to make sure that the skin does not flip over. If a small piece of skin is turned over, the dead keratin will accumulate under the skin and form a cholesteatoma, termed an iatrogenic cholesteatoma (caused by the surgeon). It is critical

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FOUR BONUS VIDEOS

Read this month's Clinical Consultation case, and then watch the accompanying videos from Hamid R. Djalilian, MD, to review the patient's imaging for yourself.

In the first video, the axial (nose is at the top of the image) CISS MRI demonstrates a hyperdense (bright compared with the brain) area in the center that has the consistency of cerebrospinal fluid (CSF), indicating high water content.

The second video shows the coronal (top of the head is at the top of the image) CISS MRI of the eroding mass in contact with the posterior semicircular canal. The mass is also causing mass effect and compression of the cerebellum.

In the third video, coronal diffusion-weighted images (HASTE sequence) demonstrate a hyperintense, or very bright, area in the mastoid, indicating a high likelihood of cholesteatoma.

The T1 non-contrast axial MRI, depicted in the fourth video, demonstrates the right mastoid mass with similar density to CSF.

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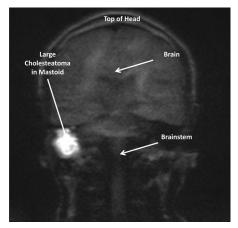


Figure 2. The coronal diffusion-weighted imaging MRI of our patient demonstrates a bright signal, indicating the presence of a cholesteatoma.

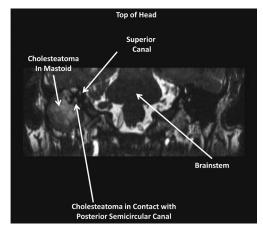


Figure 3. The coronal CISS MRI of our patient shows the cholesteatoma abutting the posterior semicircular canal, which is represented by a small white dot.

The treatment of cholesteatoma is surgical removal. In this patient, there is a very large defect. This defect can be filled using abdominal fat, bone pate or shavings, cartilage, or artificial materials. There is a risk of infection if artificial materials get exposed through the skin of the ear canal

Alternatively, a large cavity of the mastoid can be connected to the ear canal, but that would require lifelong cleanings of the ear. If obliteration of the cavity is undertaken, annual MRIs need to be obtained for at

to pay exquisite attention at the very end of the operation to ensure that the skin flaps do not flip.

least three years to evaluate for recurrence of the cholesteatoma.

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