Symptom: Gradual Hearing Loss

By Hamid R. Djalilian, MD

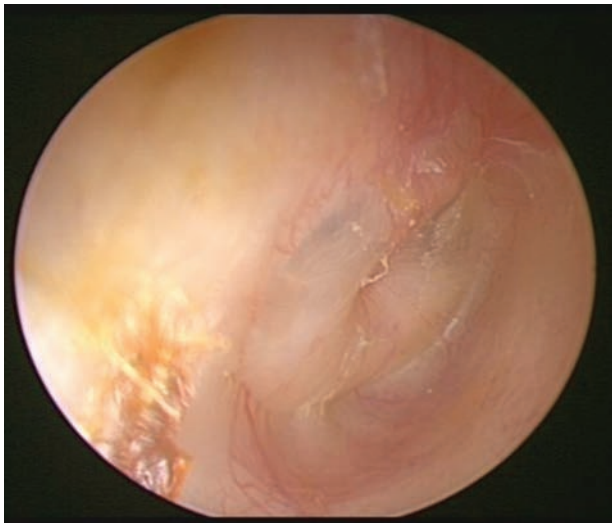
A 39-year-old female is seen in the office for hearing loss. She has noticed a gradual decline in her right ear over the past few years and has experienced difficulty hearing in noisy environments for the past 10 years, she said.

Her primary doctor treated her for an ear infection, which did not improve her hearing. This treatment course was repeated a few times without benefit.

The patient saw an otolaryngologist, who treated her with more antibiotics, but again there was no improvement. At that point, her otolaryngologist suggested a myringotomy and PE tube placement. Her otoscopic exam is shown on the right.

What is your diagnosis? See p. 10.

Dr. Djalilian is director of neurotology and skull base surgery and associate professor of otolaryngology and biomedical engineering at the University of California, Irvine.



This image shows the patient's right tympanic membrane.

Diagnosis: Neurofibromatosis Type 2

By Hamid R. Djalilian, MD

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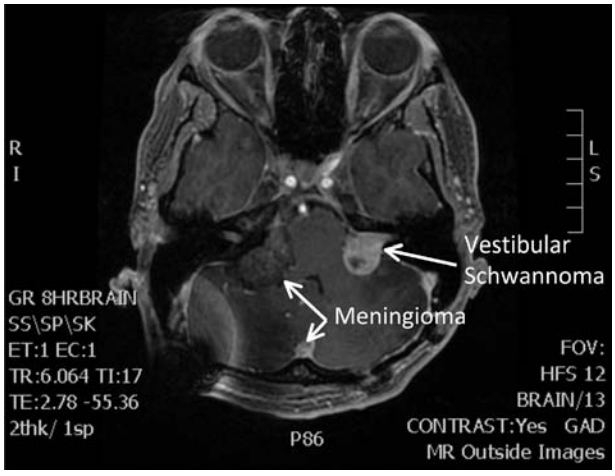
The most common etiology for the opaque appearance of the tympanic membrane is purulent or mucoid middle ear fluid. Patients with middle ear fluid usually have additional symptoms related to the underlying cause.

For example, acute otitis media generally leads to significant pain and pressure in the middle ear, and often results in systemic effects as well, such as fever and chills. In addition, an otherwise healthy adult with no previous history of Eustachian tube dysfunction should not start having acute otitis media episodes at age 39. Pure Eustachian tube dysfunction rarely causes a mucoid fluid in adults; a serous, or yellowish, middle ear fluid is most common.

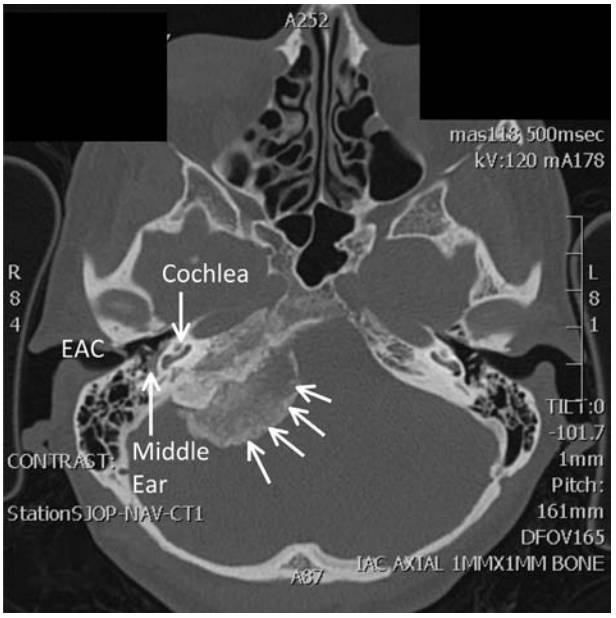
The image of the patient's right tympanic membrane on page 7 is consistent with a middle ear mass. The mass is occupying the lower three-quarters of the middle ear, with some air seen superiorly. The most common middle ear masses include cholesteatoma, an anomalous jugular vein or carotid artery, glomus tympanicum or jugulare tumors, middle ear adenoma, and facial nerve schwannoma.

When a patient presents with a middle ear mass, the first task is to define the mass with imaging. There are two primary modalities for imaging the temporal bone. The first is computed tomography (CT), which provides the best anatomical detail for the bone surrounding the ear.

In the temporal bone, CT generally cannot differentiate a mass from surrounding inflammation or fluid. Magnetic resonance imaging (MRI) provides the best differentiation between a tumor and surrounding tissue.



The patient's axial T1 post-gadolinium-enhanced MRI shows meningiomas posteriorly on the sagittal sinus and in the right (left side of the image) cerebellopontine angle (CPA). The left (right side of image) vestibular schwannoma can be seen. The right CPA meningioma does not appear bright because of its significant calcification.



This axial CT of the temporal bone demonstrates a large calcified meningioma (arrows) in the cerebellopontine angle involving the temporal bone. Tumor in the middle ear can be seen. "EAC" stands for external auditory canal.

The patient's MRI, shown on the left, revealed multiple intracranial tumors: a left vestibular schwannoma, an apparent meningioma in the right cerebellopontine angle, and a midline posterior meningioma involving the sagittal sinus area.

The CT image, which appears above, showed a large calcified meningioma involving the jugular bulb, cerebellopontine angle, and the temporal bone. The mass seen in the middle ear is the extension of the meningioma into the middle ear.

THREE DIAGNOSTIC CRITERIA

This patient has neurofibromatosis type 2 (NF2). NF2 is an autosomal-dominant condition characterized by the intracranial development of multiple neurofibromas, meningiomas, or ependymomas. It affects approximately one in 50,000 people.

Patients with NF2 most commonly have bilateral vestibular schwannomas (90%), meningiomas (50%), spinal tumors (50%), cutaneous neurofibromas, and eye lesions (90%).

The diagnosis of neurofibromatosis type 2 requires that one of the following three criteria be met: 1) bilateral vestibular schwannomas; 2) first-degree relative with NF2 and unilateral vestibular schwannoma before age 30;

or 3) first-degree relative with NF2 and the presence of two of the following: neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity/juvenile cortical cataract.

In most autosomal-dominant hereditary disorders, an inherited mutation in one of the involved genes leads to disease manifestation. In neurofibromatosis type 2, the mutated gene is only inherited half the time. In the other 50 percent of patients, it occurs spontaneously.

If the NF2 gene is suspected, prenatal testing can identify it in most cases. Screening of family members of NF2 patients is necessary to uncover silent tumors that may require treatment before they cause symptoms.

NF2 patients frequently present with hearing loss. Most often, the hearing loss is sensorineural, bilateral, progressive, and caused by the vestibular schwannomas, and these patients become postlingually deafened adults as the tumors grow or are treated surgically or with radiation. Initially, the majority of these patients will require hearing aids, but, eventually, they will lose all functional hearing.

The auditory brainstem implant (ABI) has been most commonly used for the rehabilitation of these patients. However, the development of open-set speech recognition with an ABI has been very uncommon in NF2 patients.

More recently, there has been a trend toward early intervention on the tumors and preservation of the cochlear nerve with cochlear implantation. NF2 patients with cochlear implants have had much better performance compared with those who have an ABI.

SURGICAL TREATMENT

While most of the tumors are treated surgically, recent evidence from a few small studies suggests that the anticancer drug bevacizumab may help shrink tumors in some patients with NF2. Currently, a large multi-institutional study is evaluating the drug's efficacy for this population.

Surgical therapy of the tumors depends on the presence and degree of hearing loss. A hearing preservation approach (retrosigmoid or middle cranial fossa) is performed in patients with useful hearing. A translabyrinthine approach is used in patients without functional hearing. In some occasions, the internal auditory canal is decompressed, allowing for tumor expansion without compression of the cochlear nerve to reduce the likelihood of hearing loss.

Some clinicians have used stereotactic radiation in NF2 patients. While this treatment modality is better tolerated in the short term, the patient has a higher risk of developing a malignancy in the long term. Therefore, radiation is reserved for special cases.

In cases where a cochlear implant is planned, intraoperative promontory stimulation is performed to gauge the function of the cochlear nerve. An auditory brainstem implant can be placed using a retrosigmoid or translabyrinthine approach.