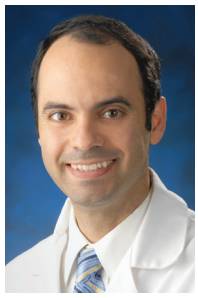


Symptom: Recurrent Otitis Media

By Hamid R. Djalilian, MD

A 25-year-old woman presents with a history of acute otitis media. During general health checks with her primary care doctor, the patient has been diagnosed with otitis media on different occasions over the past 18 months. Her medical history otherwise is not significant.

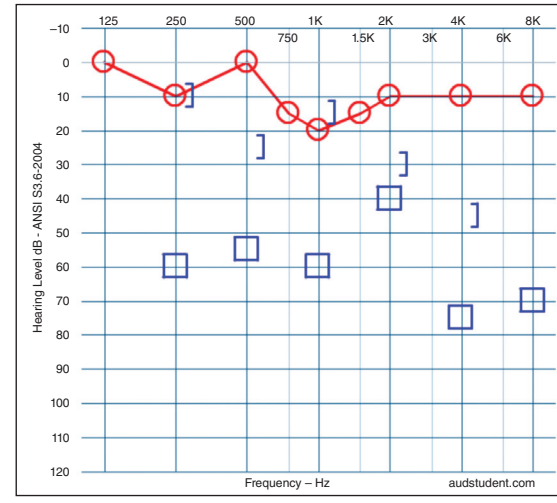
The patient's main complaint is hearing loss that she's experienced for about two years. The examination reveals a red, bulging tympanic membrane, with some overlying debris.

Palpation of the bulging portion of the tympanic membrane under microscopy demonstrates it to be hard. There is no pulsation or drainage.

The patient's audiogram is shown on the right.

What is your diagnosis? See p. 22.

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 month's patient, who has a history of acute otitis media and hearing loss, presented with the above audiogram.

Diagnosis: Middle Ear Adenoma

By Hamid R. Djalilian, MD

Continued from p. 19

While acute otitis media typically is a bacterial infection of the middle ear, it can be caused by a virus. As its nomenclature implies, the condition has an acute course, occurring over a short period of time.

In children, the diagnosis of acute otitis media requires onset within 48 hours of presentation, as well as the presence of middle ear fluid and middle ear inflammation. Signs and symptoms of middle ear fluid ideally are recognized with pneumatic otoscopy or a tympanogram, and identification of inflammation requires visualization of erythema.

In general, it is extremely uncommon for otitis media to present in somebody with no pain, fever, or acute symptom course. It is also uncommon for an adult with no history of otitis media or Eustachian tube dysfunction as a child to suddenly start developing acute otitis media.

In adults presenting with acute otitis media in the absence of previous Eustachian tube problems or chronic otitis history, the clinician should first be thinking about uncommon presentations of systemic diseases. These patients are best evaluated by an otolaryngologist.

Wegener's granulomatosis is a systemic condition that affects the lungs, kidneys, sinuses, and ears, most commonly. Patients with this condition can present with acute otitis media or serous otitis media.

Sometimes, otitis is the only presenting sign, even though there is active disease in the lungs, kidneys, and sinuses, which otherwise may have gone undetected. Patients should be questioned about recent onset of sinusitis, cough, or change in the color of their urine.

Workup generally is deferred to the primary care physician or rheumatologist. However, a quick urinalysis, erythrocyte sedimentation rate, and antineutrophil cytoplasmic autoantibodies (ANCA) test can help provide the clinician with some screening tests to speed up the diagnostic process.

It is uncommon for an adult with no history of otitis media or Eustachian tube dysfunction as a child to suddenly start developing acute otitis media.

Other systemic conditions that affect the temporal bone are tuberculosis, syphilis, histiocytosis X, and sarcoidosis, among others. Additional diseases that affect the immune system include human immunodeficiency virus and leukemia. These conditions should be included in the differential diagnosis.

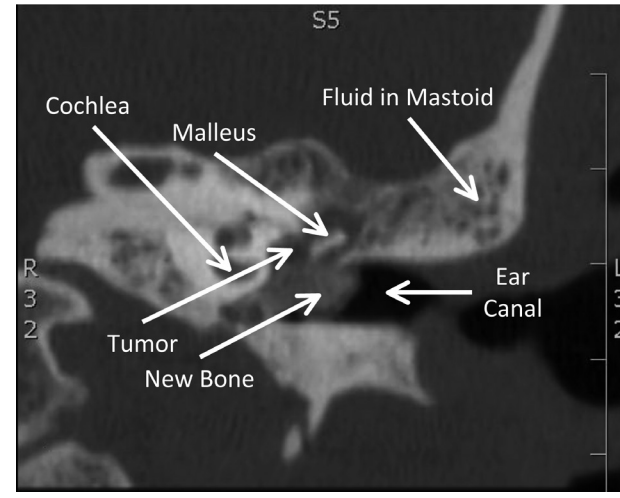


Figure 1. This axial CT of our patient's temporal bones at the level of the mid-tympanic membrane demonstrates opacification of the middle ear with surrounding bone formation bulging through the tympanic membrane.

A new onset of acute otitis media in adulthood without previous Eustachian tube dysfunction or otitis media should also prompt an evaluation of the nasopharynx, middle ear, and skull base to rule out the presence of a tumor that could either cause blockage of the Eustachian tube or fill the middle ear, creating otitis or the appearance of a red, bulging tympanic membrane.

WHAT'S IN A NAME?

Our patient was taken to the operating room, where a tumor was surgically excised. Pathology revealed the tumor to be a neuroendocrine adenoma of the middle ear (NAME), a benign neoplasm in which cells have characteristics of neuroendocrine and mucin-secreting cells. This tumor type is uncommon, encompassing two percent of temporal bone tumors.

Neuroendocrine adenomas of the middle ear have been called middle ear adenomas, carcinoid tumors, amphicrine adenomas, adenocarcinoids, and adenomatous tumors of the middle ear. The tumor can be mistakenly read on pathology results as an adenoid cystic carcinoma as well.

Patients with a NAME present most commonly with hearing loss, tinnitus, and aural pressure, and, sometimes, with discharge. While the tumor is benign, it can extend into the mastoid, Eustachian tube, and external auditory canal.

On examination, these patients usually have an intact tympanic membrane with a mass or middle ear effusion.

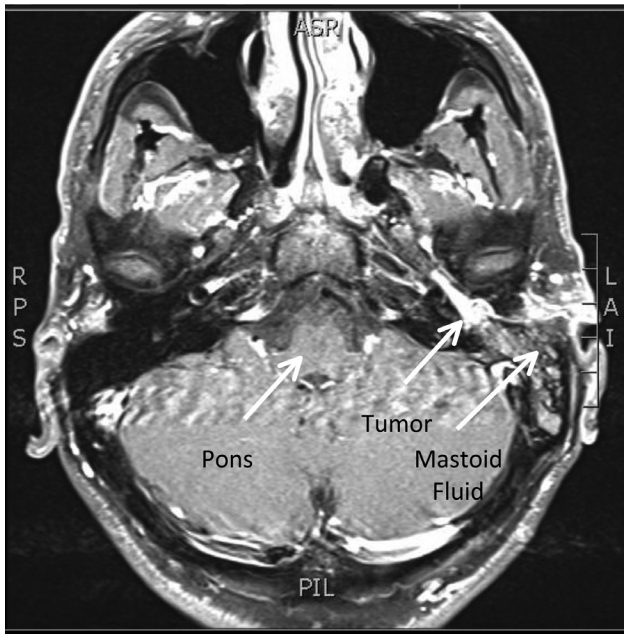


Figure 2. This axial gadolinium-enhanced T1-weighted MRI of the internal auditory canals shows enhancement of the tumor in the middle ear and the tumor's extension into the mastoid.

While this tumor is termed neuroendocrine, it does not generally secrete neurotransmitters or systemically active molecules.

Imaging with a temporal bone CT and MRI is usually necessary to appreciate fully the extent of the tumor.

On CT, the tumor will appear as an opacification of the middle ear and, possibly, the mastoid. When using CT imaging, it is difficult to distinguish the tumor from the presence of fluid in the middle ear caused by Eustachian tube blockage.

Mastoid opacification generally results from blockage of the aditus ad antrum (connection between the middle ear and mastoid). However, the tumor may extend into the mastoid through the aditus ad antrum.

Bony destruction generally is not seen in patients with a NAME, as the tumor spreads along paths of least resistance. In our patient, the tumor had led to some osteogenesis (new bone formation) in the middle ear, which was evident on the preoperative CT scan (see figure 1).

MRI can help distinguish the tumor from middle ear and mastoid fluid. Middle ear or mastoid fluid appears bright on T2-weighted images, and the tumor will appear bright on post-gadolinium T1-weighted images (see figure 2).

NAME is treated surgically, generally via a tympanomastoidectomy, but the approach depends on the extent of the tumor. If the tumor extends medially to the malleus and incus, the removal and replacement of the incus may be necessary to ensure full tumor removal.

Recurrence is most common when the disease involves the ossicles but the ossicles are left entirely intact. We have found the argon laser to be beneficial in removing microscopic disease when there is extensive infiltration of the middle ear and mastoid.