Symptom: Mixed Hearing Loss

By Hamid R. Djalilian, MD

A 37-year-old female presents with a history of hearing loss in her right ear. She states that the hearing loss has been gradual and that she is not able to hear people very well on that side.

The patient has a plugging sensation in the same ear that has been gradual in onset as well, and tinnitus and mild imbalance, which have been present for one to two years.

She is otherwise healthy and says she has never had ear surgery. She had some ear infections as a child, but never as an adult.

Visualization of the ear under microscopy shows a white colored mass behind the tympanic membrane.

What is your diagnosis? See p. 12.

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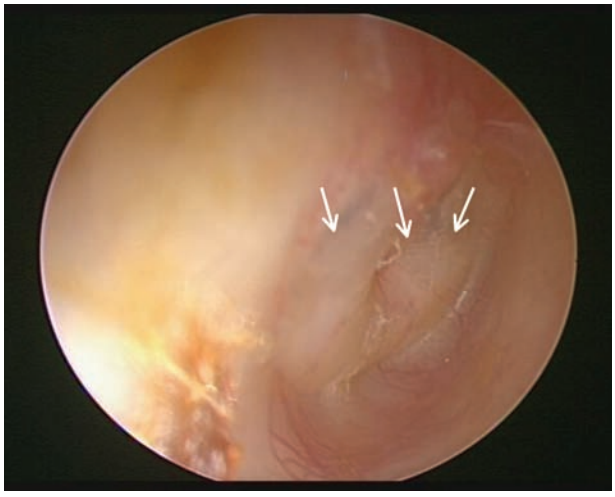

In this image of the right tympanic membrane, note the white mass, outlined at the superior edge with arrows.

Diagnosis: Meningioma in Middle Ear

By Hamid R. Djalilian, MD

Continued from p. 10

A white mass behind an intact tympanic membrane is most commonly associated with a congenital cholesteatoma. A congenital cholesteatoma is due to the persistence of epithelial rests in the middle ear during embryogenesis.

The squamous epithelium that makes up the outer surface of the tympanic membrane and the ear canal constantly generates new skin layers and sheds the dead layer of skin, called keratin.

The keratin on the outer surface of the tympanic membrane and ear canal, as well as the rest of the body, gradually is shed and washed away. The keratin produced by a congenital cholesteatoma will accumulate in the middle ear, gradually expand, and eventually destroy surrounding tissue.

A congenital cholesteatoma can lead to a mixed hearing loss, though initially it will cause a conductive hearing loss.

Another possible cause of a white mass behind the tympanic membrane is a dehiscent carotid artery. While we associate the color red with arteries, the thicker wall of the carotid artery gives it a whitish appearance in the temporal bone.

A dehiscent carotid artery is caused by an error in embryogenesis; the carotid artery develops in an aberrant fashion and is more laterally and posteriorly located. A dehiscent carotid artery would generally not cause a sensorineural hearing loss.

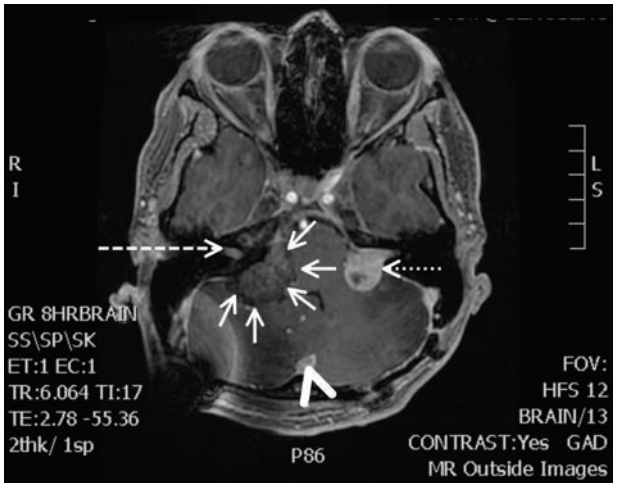

Figure 1. *This axial T1-weighted post-gadolinium MRI of the temporal bone shows the tumor (indicated by the small solid arrows), which does not take up contrast. Bilateral internal auditory canal masses are also seen, as shown by the dashed arrows, and there is a small midline posterior meningioma present as well, marked by the arrowhead.*

It is uncommon for meningiomas to invade the middle ear, but that can happen via a few routes.

A last cause of a white mass in the middle ear is involvement with a tumor or herniation of brain and meninges into the middle ear.

Brain herniation into the ear, termed encephalocele, can occur in the presence of congenital defects of the roof of the middle ear (tegmen tympani) or in obese patients, particularly middle-aged females.

Tumors that could cause this picture of a white mass behind an intact tympanic membrane include facial nerve schwannomas, meningiomas, and other primary middle ear tumors.

UNCOMMON INVASION

In this patient, an MRI showed a large, non-enhancing mass in the cerebellopontine angle on the patient's right side (left side of *figure 1*), consistent with a calcified meningioma.

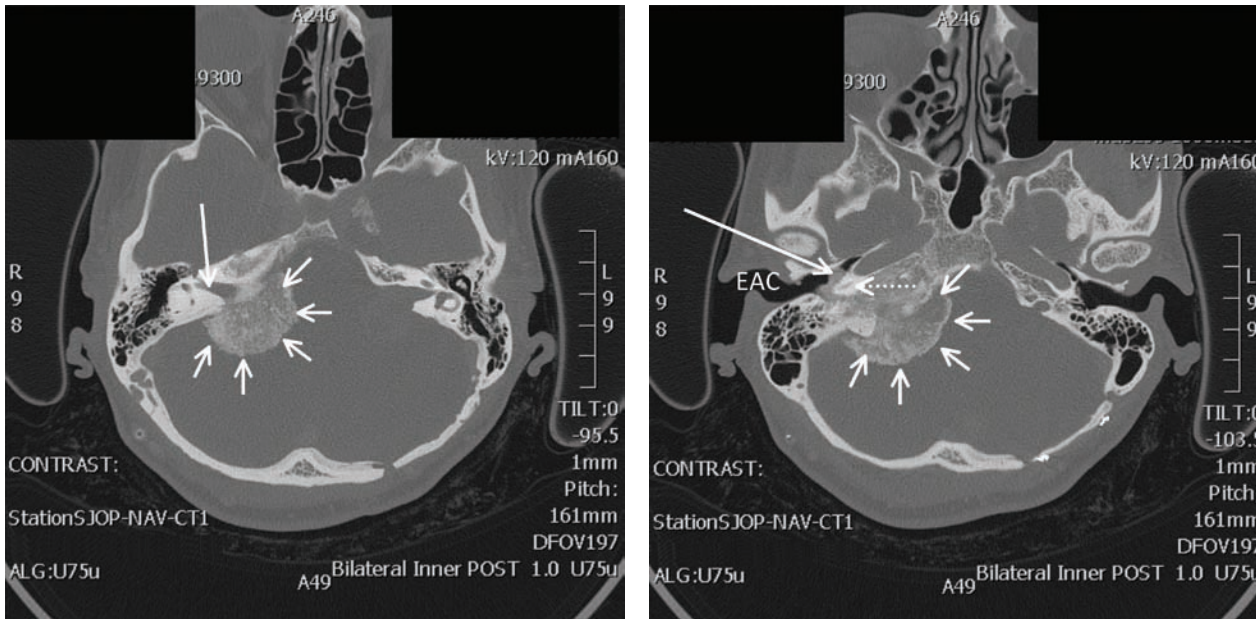

Figure 2. *Left: In this CT image of the temporal bone at the level of the internal auditory canal (long arrow), the calcified (white) mass, indicated by small arrows, is seen abutting the medial side of the internal auditory canal. Right: This lower-level CT image shows the invasion of the middle ear (long arrow) with the mass (small arrows). The basal turn of the cochlea (dashed arrows) is also shown, as is the external auditory canal (EAC), which is seen on the patient's right (left side of image).*

The patient had bilateral enhancing masses in the internal auditory canals, indicated in *figure 1* by dashed arrows, consistent with bilateral vestibular schwannomas. In addition, she had a small midline meningioma, as marked by the large arrowhead in the figure.

The long-term prognosis of neurofibromatosis type 2 generally is not good due to the mass effect of the various tumors or complications arising from their treatment.

A CT scan showed the calcified nature of the meningioma and its involvement of the middle ear (*see figure 2*).

Based on the imaging, the patient was diagnosed with neurofibromatosis type 2, a rare genetic condition that causes multiple intracranial tumors, including schwannomas, meningiomas, and ependymomas. The presence of bilateral vestibular schwannomas is diagnostic of this condition.

It is uncommon for meningiomas to invade the middle ear, but that can happen via a few routes, including through the facial nerve via the geniculate ganglion, bony openings from blood vessels, congenital defects, travel through air cells, or frank invasion of the bone.

Vestibular schwannomas generally do not invade the temporal bone or involve the middle ear. Facial schwannomas, which can occur in patients with neurofibromatosis type 2, are able to invade the temporal bone and involve the cochlea as well as the middle ear structures.

Treatment of meningiomas consists of surgery or stereotactic radiation therapy. Surgical resection is generally the treatment of choice in younger patients, though therapy is tailored to each individual patient.

Stereotactic radiosurgery using the Gamma Knife or CyberKnife device is used in cases of unresectable residual tumor or as a sole treatment modality. Sometimes, depending on the location of the tumor and the age of the patient, the tumor is observed for growth. Treatment is then instituted when growth is seen on serial imaging, which is performed every six to 12 months.

The long-term prognosis of neurofibromatosis type 2 generally is not good due to the mass effect of the various tumors or complications arising from their treatment.

Evaluation of the patient's siblings and children with an MRI of the brain and spine is necessary to identify relatives with the disease. Early treatment of these tumors can lead to a better hearing outcome.

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