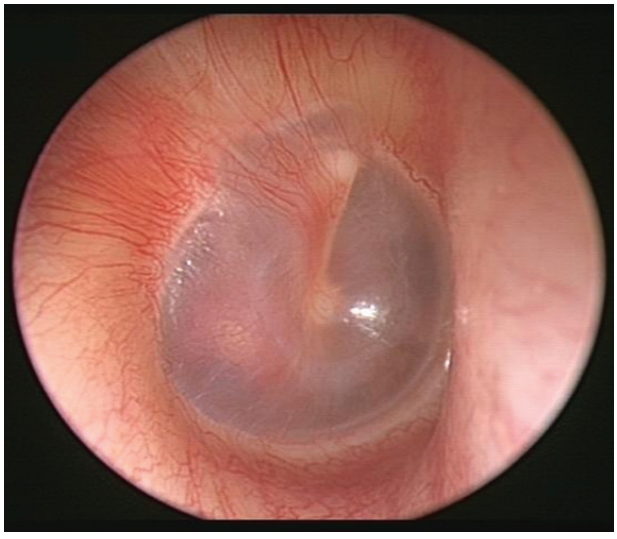
Symptom: Redness in Middle Ear

By Hamid R. Djalilian, MD



This image of our patient's tympanic membrane shows erythema of the promontory that is seen deep to the tympanic membrane.

A 45-year-old male comes into the office with a complaint of hearing loss. He had noticed this difficulty hearing over the last year, especially in noisy environments.

The patient works as a physician and sometimes has trouble understanding in a conference setting. It is also challenging for him when people mumble.

He denies having had any previous surgery on his ears or problems with ear infections as a child. His father wore hearing aids starting at age 60.

Examining the patient's ear, we find that there is a reddish hue to the middle ear structures. The Rinne exam shows bone conduction to be louder than air conduction.

What is your diagnosis? See p. 19.

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

Diagnosis: Otosclerosis

By Hamid R. Djalilian, MD

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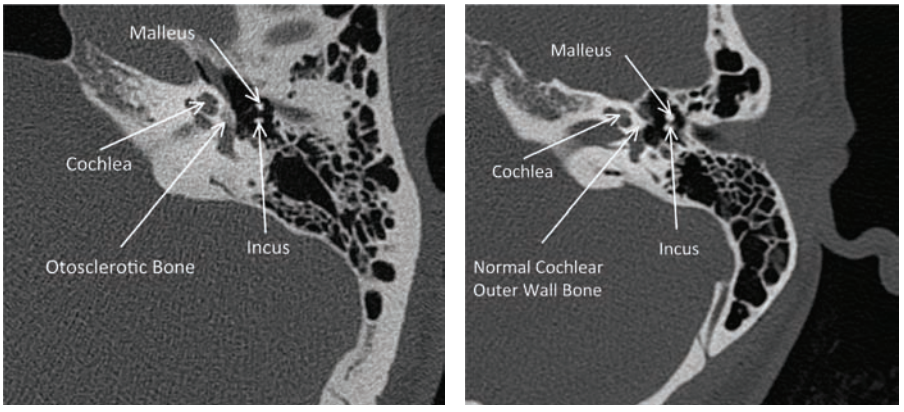
The reddish appearance on the otoscopic exam is generally associated with an infectious process. Infection causes increased vascularity of the tympanic membrane, making the membrane look red. This often occurs in conjunction with fluid in the middle ear.

Redness of the tympanic membrane in the absence of middle ear fluid can be seen in a child who is screaming during the examination. The screaming causes engorgement of the capillaries of the tympanic membrane, creating the appearance of a hyperinflated red tympanic membrane.



Left: *This axial (horizontal) CT image of the left temporal bone shows the characteristic gray bone of otosclerosis on the outer wall of the cochlea. Anterior is at the top of the image; the outer ear is on the right.* ***Right:*** *This equivalent image of a normal patient shows the normal dense white bone of the outer wall of the cochlea*

What's observed in this patient is distinct from a red tympanic membrane in that the erythema is on the promontory of the cochlea (outer wall). The tympanic membrane itself is actually normal.

This subtle exam finding is best appreciated with microscopic examination rather than the use of an otoscope. With a microscope, one is able to focus further into the middle ear and notice that the redness is from the promontory and not the tympanic membrane. This can sometimes be seen when an endoscope is used for a magnified examination of the tympanic membrane.

GENETIC ETIOLOGY

The redness of the outer wall of the cochlea in this patient is associated with a hypervascular bone of otosclerosis, termed Schwartze's sign. Otosclerosis only affects the endochondral bone of the inner ear, called the otic capsule.

The abnormal otosclerotic bone grows uncontrollably, gradually causing hearing loss in some patients. Most patients present in their 20s and 30s, and 90 percent of affected patients are under 50 at the time of diagnosis.

In studies of postmortem temporal bone histopathology, 10 percent of temporal bones from Caucasians had foci of otosclerosis. However, only 10 percent of those individuals had hearing loss.

Otosclerosis is less common in African-Americans and Asians, and more common in women. An acceleration of otosclerosis-related hearing loss is seen during pregnancy.

Multiple etiologies for otosclerosis have been theorized, but the most commonly accepted cause is genetic. Several genes associated with otosclerosis have been identified, with each on a different chromosome. An autosomal dominant pattern with variable penetrance has been seen.

iPad Video!

TREATMENT: WATCH THE SURGICAL OPTION FOR OTOSCLEROSIS

Video by Hamid R. Djalilian, MD

In the August iPad issue, watch as Hamid R. Djalilian, MD, director of neurotology and skull base surgery and an associate professor of otolaryngology and biomedical engineering at the University of California, Irvine, performs a stapedotomy, which is the surgical option for patients diagnosed with otosclerosis.

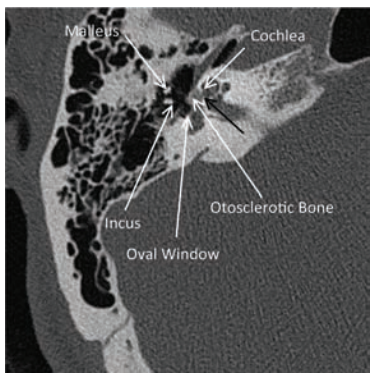
In a stapedotomy, the fixed stapes is replaced with a prosthesis. Generally, the surgery is successful at reducing the air-bone gap to less than 10 dB in about 95 percent of patients.

To see the surgery, download The Hearing Journal app, and the video, for free today at bit.ly/AppHearingJ.

Another theory is that a post-measles viral infection causes changes in the bone. Genetic material from the measles virus has been found in otosclerotic bone of some patients.

The most common area of involvement is the fissula ante fenestram, which is at the anterior edge of the oval window. Encroachment of that focus of otosclerosis onto the stapes footplate leads to the conductive hearing loss observed in patients affected by this disease.

The second most common site of involvement is the posterior aspect of the oval window, called the fossula post fenestram. Other locations include the round window and internal auditory canal. Sometimes, the otosclerotic bone will invade the cochlea and cause sensorineural hearing loss, termed cochlear otosclerosis.



This axial (horizontal) CT image of the right temporal bone demonstrates cochlear otosclerosis. Unlike the image of the left temporal bone with otosclerosis, where a narrow area of dense white bone separates the otosclerotic gray bone from the cochlear membranes, here the gray otosclerotic bone is continuous with the cochlea (black arrow).

CARHART NOTCH

Patients presenting with otosclerosis have some distinguishing characteristics on audiological testing. Most patients will have a low-frequency conductive hearing loss combined with a drop in the bone conduction threshold at 2,000 Hz, termed a Carhart notch. The cause of the drop in the 2-kHz threshold is unknown; however, a normalization of this threshold often is seen after surgical repair.

Nearly all patients with otosclerosis have an absent acoustic reflex once the conductive hearing loss has advanced beyond 30 dB in the lowest frequencies. A shallow (type As) tympanogram is seen due to the stiffness of the ossicular motion that occurs with stapes fixation.

Patients who do not have these characteristics can potentially have superior canal dehiscence and should be evaluated for that with an ultra-fine-cut CT scan of the temporal bones.

THREE OPTIONS

In the office, patients diagnosed with otosclerosis are generally presented with three options: watchful waiting, hearing aid, and surgical repair. Most commonly, older patients choose hearing aids, while younger patients opt for surgery.

The surgery, called stapedotomy, involves a transcanal approach to the middle ear. Once the stapes has been exposed, its immobility is confirmed. The stapedial tendon and posterior crus are cut by a laser.

A small (0.6-mm) opening is placed into the oval window. After measurement of the distance between the incus and the stapes footplate, a prosthesis is attached to the incus and placed into the oval window opening. Tissue or blood is used to seal around the prosthesis at the oval window.

The surgery generally is successful at reducing the air-bone gap to less than 10 dB in about 95 percent of patients.