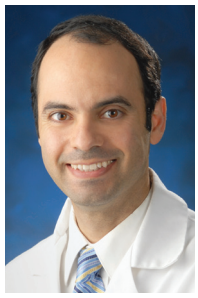


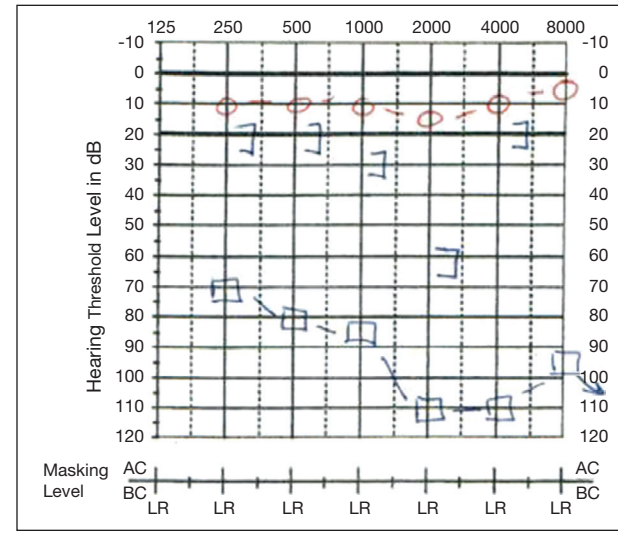
Symptom: Sensorineural Hearing Loss, Hyperacusis

By Hamid R. Djalilian, MD

A 55-year-old man presents with a history of rationally progressive sensorineural hearing loss. Hyperacusis has also been a major problem for him. He said he feels the hearing loss started approximately 15 years ago and he has been using hearing aids for the last five years. He denies any significant family history and does not have surgery. He is an English teacher and has not had substantial noise exposure, but he has been unable to work due to his hyperacusis. He denies any use of firearms or power tools. His audiogram is on the right.

What is your diagnosis? See p. 12.

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



The patient's audiogram

Diagnosis: Cochlear Otosclerosis

By Hamid R. Djalilian, MD

Continued from p. 10

This patient presented with significant progressive hearing loss and hyperacusis. We generally assume a genetic etiology in patients with progressive sensorineural hearing loss. Genetically programmed cell death is thought to be one of the causes of presbycusis. Initially, an MRI was obtained to look for a possible etiology for the patient's hyperacusis by his primary physician. The MRI images were reviewed and found to be normal. The patient, however, wanted a CT scan of the temporal bones for the superior canal dehiscence. After discussion with the patient that this was unlikely the cause of his hyperacusis, the patient still insisted on a CT scan, and it was obtained. A review of the scan showed the patient has substantial cochlear sclerosis. Further scrutiny of the CT scan showed the patient interestingly has otosclerosis involving the entire cochlea and vestibule that spares the area of the fissula ante fenestram. The fissula ante fenestram is the area anterior to the oval window. This is the most common site of otosclerosis and its involvement is the cause of the fixation of stapes in otosclerosis.

The patient's CT scan shows extensive cochlear otosclerosis, which is a process of bone remodeling that exclusively occurs in the otic capsule. Histologic otosclerosis is defined as the post-mortem finding of otosclerosis. Clinical otosclerosis occurs when the otosclerosic plaque fixes the stapes footplate and causes conductive hearing loss. The definition of cochlear otosclerosis is that of a focus of otosclerosis located in the otic capsule involving the cochlear endosteum, which causes sensorineural hearing loss (SNHL). Usually when otosclerosis is severe enough to involve the cochlear endosteum, it causes stapes fixation as well.

"Cochlear otosclerosis is clinically divided into mixed hearing loss and pure sensorineural hearing loss."

BREAKING DOWN OTOSCLEROSIS

Cochlear otosclerosis is clinically divided into mixed hearing loss and pure sensorineural hearing loss. Cochlear otosclerosis, where involvement of cochlear endosteum occurs and causes SNHL without stapes fixation, is uncommon. It is thought to be the cause of progressive sensorineural hearing loss with no conductive damage in about one percent of the patients with progressive loss.

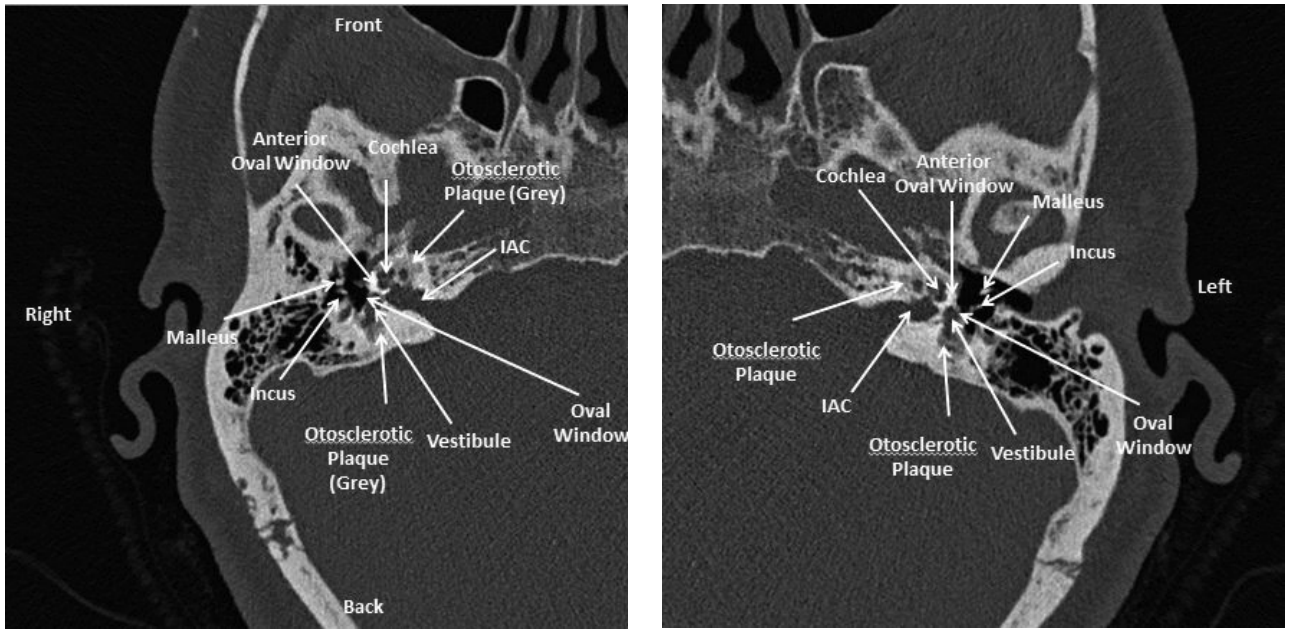


Figure 1A and Figure 1B. Axial CT image of the right (A) and left (B) temporal bones demonstrating cochlear and vestibular involvement by otosclerosis (gray blotches). The fistula ante fenestrum (the anterior part of the oval window) is not involved and appears bright white as normal cochlear bone should be.

Otosclerosis is—at its most basic level—a type of abnormal bone remodeling in the otic capsule. Generally, little bone remodeling is seen in the otic capsule under normal conditions. It is thought that remodeling of the otic capsule begins when molecular factors initiate a process that causes the problem in patients who have a genetic background for the disease and are triggered by environmental factors.

Several genes have been identified as possible loci of abnormalities in families with otosclerosis. These include *COL1A1, TGFB1, BMP2, BMP4, ACE, AGT*, and *RELN* genes. Cochlear otosclerosis, however, has not been identified to be associated with a particular gene.

It is thought that remodeling of the otic capsule begins when molecular factors initiate a process that causes the problem in patients who have a genetic background for the disease.

The measles virus has been theorized to cause otosclerosis in some patients. It has been shown to exist in the otosclerotic bone in patients with otosclerosis. Additionally, antibodies to the measles virus have been found in the perilymph in patients with otosclerosis. While progression of otosclerosis in pregnancy has been seen clinically, it has not been observed universally.

The clinical course of the disease depends on the progression of the plaques and the level of otic capsule involvement.

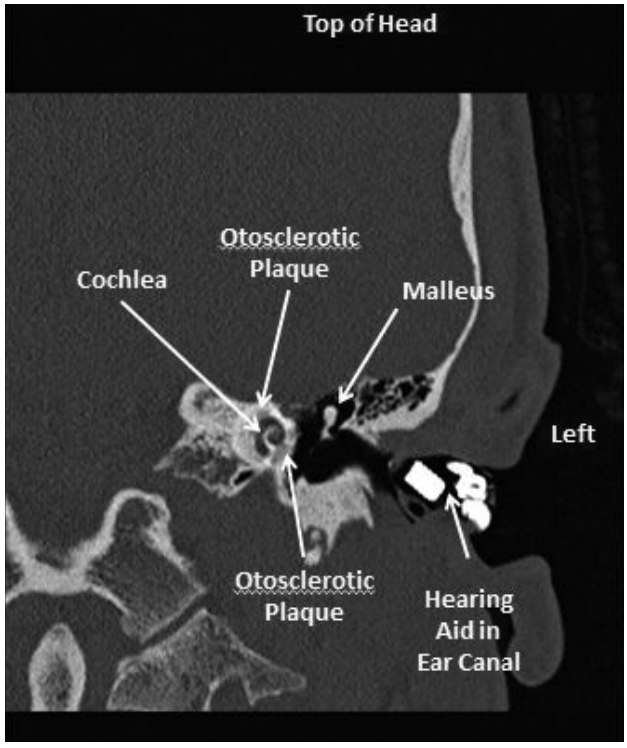


Figure 2. Coronal CT image of the temporal bone showing cochlear involvement by the otosclerotic plaque.

Some patients may develop vestibular symptoms that can be recurrent, positional, or spontaneous. These symptoms may be caused by the biochemical changes in the perilymph caused by otosclerotic involvement of the vestibular organs.

REACHING THE DIAGNOSIS, DECIDING TREATMENT

This patient had vestibular involvement but denied vestibular symptoms. Benign paroxysmal positional vertigo is seen in patients with otosclerosis and may be manifested post-stapedectomy. The presence of an active focus of otosclerosis on the promontory causes a reddish appearance on otoscopy, termed Schwartze's sign.

Computerized tomography imaging shows a hypodense (gray) double ring around the cochlea, which occurs from loss of mineralization of the bone in cochlear otosclerosis. CT imaging will only demonstrate active otosclerosic plaques because areas have a loss of mineralization. Inactive otosclerosis is not as clear on CT and may be bright white. An MRI may show a ring of intermediate signal in the areas around the cochlea or vestibular organs on T1 weighted images. Some enhancement after gadolinium administration may be seen due to the hypervascular nature of the bone. T2 weighted images may show increased signals due to the higher water content of active otosclerosis plaques compared to the dense cochlear bone, which has low water content.

Sodium fluoride has been thought to reduce the activity of the otosclerotic bone. What prompted the use of fluoride was that otosclerosis was seen more commonly in areas of northern Europe where the population primarily drank non-fluorinated water. It has been theorized that fluoride reduces the activity of the enzymes that contribute to the destruction of the native otic capsule bone and its replacement with the active vascular otosclerosis bone.

Fluoride's primary side effects include the development of joint pain, osteoporosis, and gastrointestinal issues. In our practice, the patients are screened with bone densitometry before and one year after initiation of fluoride treatment.

Bisphosphonates are a class of drugs used for the treatment of osteoporosis. They have been thought to help reduce the breakdown of bone seen in otosclerosis. The results of the studies on this treatment are mixed. A clinical trial is underway to determine its efficacy. The potential severe adverse effects of this class of drugs lead us to exercise caution and await definitive data before using this for our patients.

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