

Unusual Imaging Manifestations of Severe Bilateral Cochlear Otosclerosis: A Case Report

► *Katie Taylor; DO: Alberto Iaia, MD; Michael Teixido, MD; Francesco Agnello, MD; Gianvincenzo Sparacia, MD*

Abstract

Otosclerosis is characterized by abnormal bony growth at the otic capsule of the petrous bone, which can lead to profound hearing loss. Crosssectional imaging plays an important role in confirming the suspicion of otosclerosis and determining disease extent. Here the authors present some unusual imaging manifestations of a severe case of bilateral cochlear otosclerosis in a 68-year-old female presenting with gradual sensorineural hearing loss.

Introduction Otosclerosis or otospongiosis, is defined as the abnormal growth of bone at the otic capsule, which typically produces a fixation of the stapes resulting in inhibition of normal sound transduction through the conductive hearing system into the cochlea.¹ With an estimated prevalence of 1 percent, otosclerosis demonstrates inheritance through an autosomal dominant gene transmission with incomplete penetrance, but may also occur sporadically.² Caucasians in their second to third decades of life are most commonly affected, with a female predilection of 2:1.³ Patients typically present with progressive bilateral conductive hearing loss. Clinical presentation widely varies depending on the extent of the disease and the exact anatomic location of involvement, in addition to conductive hearing loss, tinnitus, sensorineural hearing loss and balance instability are also frequently observed.^{1,2,4,5} The otic capsule is comprised of three layers: a thin endosteal layer, a middle layer of combined endochondral and intrachondral bone, and an outer periosteal layer. The pathophysiology of this disease may affect specific histologic components of the otic capsule. Otosclerosis occurs within the middle layer and progresses as a cycle of three coexisting pathologic phases. The first stage, known as the acute phase, is characterized by active deposition of islets of osteoid tissue. This is subsequently followed by a subacute phase, which consists of an increase in resorptive osteoclastic activity, resulting in the spongiotic remodeling of bone. Within the later chronic-sclerotic phase, osteoblastic activity is more prominent, producing new bone with irregular features.⁵ This pathologic process may be visualized by the clinician on otologic examination by the presence of a discolored, vascular hue along involved areas of the cochlear promontory, visible through the tympanic membrane (also known as Schwartze's sign).⁵

Otosclerosis is described by the anatomic locations of involvement, either as fenestral or cochlear (also known as

retrofenestral). Fenestral otosclerosis is the most common and is found in 85 percent of patients, while 15 percent present with the cochlear type. Typically, disease progression originates at the fissula ante fenestram and then spreads posteriorly to involve both the oval window and margins of the round window. Active disease then may continue to spread to the otic capsule. When disease involvement is limited to the fenestra, the stapes footplate may become fixed to the oval window niche producing a characteristic pattern of conductive hearing loss. Cochlear otosclerosis is characterized as involvement of the inner ear and is more likely to produce a sensorineural hearing loss by interfering with the transmission of electrical stimulation from the cochlear inner hair cells to the spiral ganglion neurons.^{1,5} Rarely, cochlear otosclerosis will present without involvement of the perifenestral region.⁶ Treatment of hearing loss associated with otosclerosis includes stapedectomy and placement of a stapes prosthesis for fenestral otosclerosis, hearing aids for sensorineural loss, cochlear implantation to improve hearing in patients with severe cochlear involvement, or with bisphosphonate treatment to help arrest progression of cochlear disease.

Cross sectional imaging plays an important role in confirming the suspicion of otosclerosis and determining disease extent. There are key findings on temporal bone computed tomography (CT) imaging that may indicate the success of surgical therapy. Obliteration of the round window may predict a poor outcome after stapes prosthesis placement, since a fixed round window will be unable to undulate in coordination with the oval window during sound transduction. Additionally, cochlear implantation through the round window may likewise become more challenging with this presentation. It is also important to identify the height of the oval window niche, as narrow distances (< 1.4 mm on coronal CT reformats) may result in a more challenging surgery.⁵

CASE REPORT

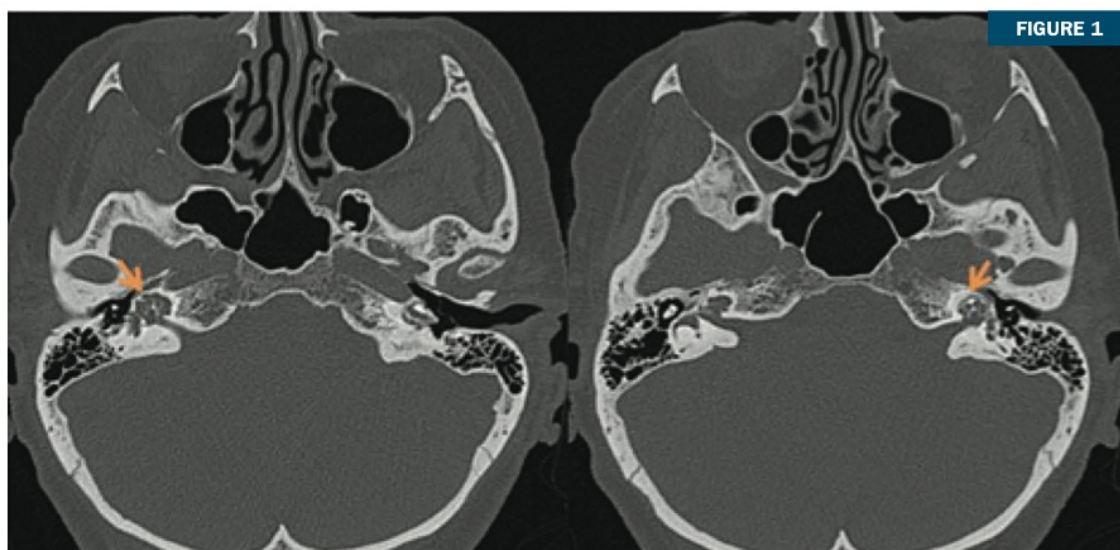


FIGURE 1

Axial HRCT images show bilateral and asymmetric pericochlear ring-like hypoattenuation (arrows), more extensive in the right middle ear.

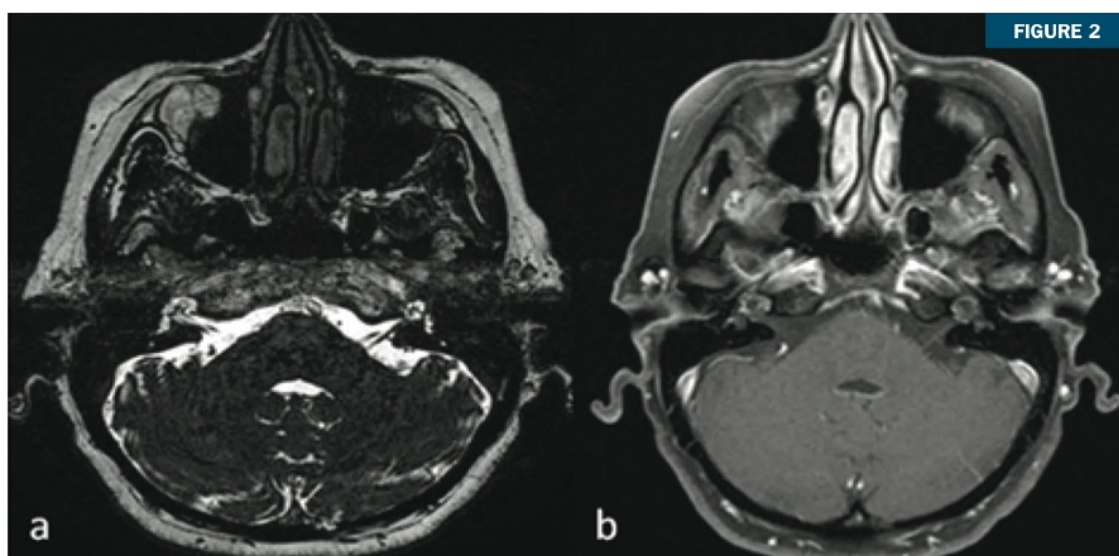


FIGURE 2

Axial MR images of the skull base show strong pericochlear hypointensity on heavily T2-weighted images and mild enhancement on gadolinium-enhanced images.

CLINICAL PRESENTATION

Here the authors present a 68-year-old female with a long-standing history of bilateral cochlear otosclerosis. She initially underwent stapedectomy on the right ear at age 16 followed by left sided stapedectomy at age 18. Despite these surgeries, the patient's hearing continued to deteriorate, eventually resulting in incomplete right-sided hearing loss by age 62. Word

discrimination in her left ear declined to 40 percent by age 66, which made her hearing aid only marginally useful. Hearing discrimination for loud speech (70 dB) with a hearing aid was 0 percent in the right ear while using sentences in quiet was 53 percent in the left ear. In 2015, she underwent cochlear implantation of a nucleus 512 device in her non-hearing right ear. A stiff styletted electrode was used to overcome obstructions in the cochlea. Although only partial electrode insertion was possible due to the extent of her disease, she did experience a

significant improvement in hearing on this side and it became her better hearing ear. In 2016, the patient's left ear was then implanted with complete electrode insertion. She experienced a hearing improvement on the left as well, and currently possesses functional hearing on both sides with 92 percent word recognition in sentences in quiet. Prior to the 2015 cochlear implantation in the right ear, the patient underwent a spiral-CT temporal bone study utilizing 0.625 mm slice thickness. These images demonstrated extensive lucency in the immediate vicinity of both the cochlea and vestibule, consistent with the patient's history of otosclerosis (**Figure 1**). Two months following the CT scan, the patient underwent an MRI study of the temporal bones. These images were acquired utilizing a 1.5T MR scanner, and included thin section images through the temporal bones. A thin-section 3D, heavily T2 weighted GRE Constructive Interference in the Steady State sequence (CISS), demonstrated marked signal abnormality surrounding the cochlea. This area of abnormal signal corresponded to the area of abnormal perichochlear lucency demonstrated on the CT scan. Remarkably intense pathologic enhancement was identified in both the perichochlear and perilyabyrinthine distributions, corresponding to the signal abnormality on MRI and abnormal lucency on the CT scan. Pathologic enhancement is very seldom observed on MR imaging of patients with this disorder and, when present, it might suggest an active or aggressive phase of the pathologic processes leading to otosclerosis (**Figure 2**).

DISCUSSION

Cross-sectional imaging plays an important role in the diagnosis and management of otosclerosis. Due to the typical histologic changes of the middle and inner ear structures during disease progression, imaging allows for the confirmation of otosclerosis in patients with documented hearing loss. It additionally may aid in diagnosis by eliminating other potential causes for hearing loss including acoustic neuromas, cholesteatomas, a congenital malformations, or infection.

This case illustrates the critical role that imaging plays in confirming the diagnosis of cochlear otosclerosis in a patient being evaluated for sensorineural hearing loss. Although conductive hearing loss occurs primarily in fenestral otosclerosis, the patient may also present with sensorineural or a mixed type hearing loss if the osseous abnormality extends to the cochlea. The patient presented herein experienced sensorineural hearing loss secondary to cochlear otosclerosis, with almost complete destruction of both otic capsules, as shown by her imaging studies.

The preferred imaging protocol for evaluating otosclerosis is a non-enhanced spiral CT of the temporal bone, secondary to its ability to detect small abnormalities of the bony labyrinth with fine anatomic detail. CT should be also used to facilitate preoperative surgical planning, as well as aid in the prediction of successful surgical outcomes. It is necessary for the CT protocol to be obtained with a slice thickness of 0.5-0.625 mm with an increment of 0.3 mm to allow for the ability to obtain high quality multiplanar reformatted images.⁷ If MR imaging is requested, a T1 contrast enhanced sequence should be obtained in order to identify enhancing foci from the acute phases of otosclerosis. A high resolution T2 MR imaging alone may miss otosclerosis.⁵

CONTRIBUTING AUTHORS

■ **KATIE TAYLOR, DO** is a fourth year diagnostic radiology resident at Christiana Care Health System in Newark, Del.

■ **ALBERTO IAIA, MD** is Chief of Neuroradiology at Christiana Care Health System in Newark, Del.

■ **MICHAEL TEIXIDO, MD** is an Otolaryngologist at Christiana Care Health System in Newark, Del. and Thomas Jefferson University in Philadelphia, Penn.

■ **FRANCESCO AGNELLO, MD** is a diagnostic radiology resident at the University of Palermo in Palermo, Italy.

■ **GIANVINCENZO SPARACIA, MD** is on the diagnostic radiology faculty at the University of Palermo in Palermo, Italy.

REFERENCES

1. Juliano AF, Ginat DT, Moonis G. Imaging Review of the Temporal Bone: Part II. Traumatic, Postoperative, and Noninflammatory Nonneoplastic Conditions. *Radiology*. 2015;276(3):655-72. doi:10.1148/radiol.2015140800. PMID: 26302389
2. Mousoulidis I, Axon P, Baguley D, et al. A review on the genetics of otosclerosis. *ClinOtolaryngol* 2007;32:239-47
3. Whetstone J, Nguyen A, Nguyen-Huynh A, Hamilton BE. Surgical and clinical confirmation of temporal bone CT findings in patients with otosclerosis with failed stapes surgery. *AJNR Am J Neuroradiol* 2014;35(6):1195-1201
4. Declau F, van Spaendonck M, Timmermans JP, Michaels L, Liang J, Qiu JP, van de Heyning P. Prevalence of histologic otosclerosis: an unbiased temporal bone study in Caucasians. *Adv Otorhinolaryngol*. 2007;65:6-16. PMID: 17245017
5. Swartz NG, Berger AS. Fenestral and cochlear otosclerosis: computed tomographic evaluation. *Am J Otol*. 1985 Nov;6(6):476-81. PMID: 4073255.
6. Lee TC, Aviv RI, Chen JM, Nedzelski JM, Fox AJ, Symons SP. CT Grading of otosclerosis. *AJNR Am J Neuroradiol*. 2009; 30:1435-1439
7. Curtin HD. Imaging of Conductive Hearing Loss With a Normal Tympanic Membrane. *AJR Am J Roentgenol*. 2016;206(1):49-56. doi: 10.2214/AJR.15.15060. PMID: 26491893.

Natural Disaster Planning: Expect the Worst, Leave Nothing to Chance

► *Kathleen Kennelly*

The best way to physically and financially survive a natural disaster is to be prepared in advance with a plan that considers the worst-case scenarios and leaves little to chance.

Over the past decade, thousands of U.S. property owners have experienced the devastating consequences of natural catastrophes, such as flood, fire, earthquake, wild fires, or windstorm. Combined, these catastrophic events have increased in severity and frequency, causing billions of dollars in losses and hundreds of fatalities and injuries in recent years.

Indeed, the period from 2008 to 2015 is on record as the most expensive for insured losses from convective events, which include severe thunderstorms, tornado, hail, lightning, and flash floods. Windstorm losses continue to cause the most catastrophe damage; fire remains the most frequent cause for displacement; and tornadoes' share of U.S. catastrophe losses is rising rapidly.

In addition, recent winter storm and wildfire losses, driven by El Niño, warm ocean current associated with severe weather conditions, have been unusually costly and erratic. For example, in 2015, the number of acres burned in wildfires in the United States was approximately 10.1 million acres, compared to 3.8 million the previous year. Meanwhile, winter storms caused an estimated \$3.5 billion in insured losses in 2015, up from \$2.6 billion in 2014.

Despite these troubling trends, many homeowners are not adequately prepared for natural disasters and often place their



properties and financial well-being at substantial risk.

At USI Insurance Services, the Personal Risk team has been advising clients on ways to better prepare for natural disasters. Based on its vast experience handling natural disaster claims, the team has compiled a comprehensive disaster preparedness checklist, best practices, and key resources to help clients better manage natural disaster risks. Following are a few must-do items.

SAFEGUARD IMPORTANT DOCUMENTS

Creating a hard copy file of key documents such as insurance declaration pages for home, auto, and other personal assets, health care coverage cards or list of emergency contacts for medical, financial, legal, insurance, and contractors are an essential part of disaster planning.

The cost of labor and materials and additional living expenses while waiting for restoration of property could range from tens of thousands of dollars to more than \$100,000, depending on the extent of damage and property values.

For example, while conducting a risk assessment for a client whose home was susceptible to flooding from storms, the USI Personal Risk team recommended the creation of hard copies with key contacts and documents be kept at a separate work location.

USI's advice proved critical when the client's home was flooded after a severe thunderstorm. Although the client and his family were forced to evacuate their home, access to the emergency files at work made it possible to quickly contact USI and carriers to get the claim started. As a result, the restoration of the client's home was completed well ahead of others in the neighborhood with similar losses,

