Clinical Evaluation of the Patient with Otosclerosis

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INTRODUCTION

Otosclerosis, an autosomal dominant condition involving the otic capsule, is histologically characterized by abnormal resorption and reformation of labyrinthine bone. Otosclerosis most commonly manifests clinically as a conductive hearing loss. However, because of variable penetrance, a mixed (conductive-sensorineural) hearing loss and purely sensorineural hearing loss can occur.1–5

The condition is most common in the Caucasian population affecting approximately 1%. An average of 10% of Caucasians have been found to have histologic evidence of otosclerosis in 2 large cadaveric studies; however, only 12% of those with histologic findings exhibited clinical signs and symptoms of otosclerosis.6,7 Japanese and South

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KEYWORDS

• Otosclerosis • Conductive hearing loss • Mixed hearing loss • Carhartt’s notch

KEY POINTS

• Otosclerosis classically presents in an adult with progressive unilateral conductive or mixed hearing loss, absent stapedial reflexes and normal otoscopy.
• An examination using a 512 Hz tuning fork is essential to diagnosis; if the examination does not correlate with the audiogram, repeat the audiogram.
• Radiologic evaluation is not essential in diagnosis but can identify other etiologies of conductive hearing loss in the setting of normal otoscopy, such as superior semicircular canal dehiscence or enlarged vestibular aqueduct.
• A computed tomography (CT) scan of the temporal bone should be performed to identify superior semicircular canal dehiscence in patients with a conductive hearing loss and symptoms of a third window phenomenon.
• A masking dilemma in bilateral far advanced otosclerosis creates a difficult clinical picture, so each case should be carefully assessed by the audiology team prior to surgical intervention.
American populations exhibit an incidence of 0.5%, and the African American population have even fewer cases. Average prevalence is 0.3%. Despite race, when 1 ear is effected the contralateral ear will show histologic signs of otosclerosis 80% of the time.

Average age of presentation is 15 to 45 years. Otosclerosis advances more rapidly in females than males. Hormonal factors have been implicated in progression of otosclerosis. Females have reported onset of hearing loss or worsening of symptoms during pregnancy. Estrogen receptors have been found on otosclerosis plaques. Despite this, Stankovic and colleagues minimized the association between pregnancy and progression of otosclerosis. This correlation remains controversial.

Clayton showed a statistically significant likelihood of women with otosclerosis to have osteoporosis when compared to their counterparts with presbycusis only.

Approximately 60% of patients with clinical otosclerosis report a family history of the condition. The remaining 40% are thought to represent autosomal-dominant inherited cases with failure of penetrance in other family members, new mutations, those with environmental etiology, and rare cases of alternate inheritance (ie, autosomal recessive).

A patient’s clinical presentation is directly affected by the location and extent of the sclerotic lesion. A lesion originating from the fissula ante fenestrum and advancing across the annular ligament of the stapes footplate will result in stapes footplate fixation and conductive hearing loss. Less commonly, the lesion progresses medially into the endosteum of the cochlea and results in a sensorineural hearing loss.

**CLINICAL PRESENTATION – HISTORY**

The classic presentation of otosclerosis is an adult-onset, progressive conductive hearing loss. Patients may describe improved hearing clarity in noisy environments. This phenomenon is known as Paracusis of Willis, wherein the conductive hearing loss subdues the background noise such that it improves the signal-to-noise ratio for the patient.

Vestibular symptoms have been reported in up to 40% of patients with otosclerosis. It is important to tease out the specifics of the vestibular complaint while obtaining the history, as misdiagnosis can have significant implications on treatment outcomes. For example, in the setting of Meniere disease, saccular distention due to endolymphatic hydrops can put the saccular membrane in contact with the underside of the stapes footplate. A stapedotomy here can lead to injury of the membrane and profound sensorineural hearing loss (SNHL), thus making Meniere disease is a relative contraindication to a stapedotomy. Furthermore, Mikulec reported 8 patients with unilateral conductive hearing loss, presumed otosclerosis, who failed to improve following an uncomplicated stapes procedure. These patients were found to have superior semicircular canal dehiscence (SSCD). It is important to screen for third window symptoms such as autophony, pulsatile tinnitus, or pressure-induced vertigo in the setting of conductive hearing loss.

**CLINICAL PRESENTATION – PHYSICAL EXAMINATION**

A complete head and neck and otological examination is performed. Otosclerosis most commonly presents with normal otoscopy. A reddish blush may be noted on the promontory. In 1873, Schwartze described a reddish hue on the cochlear promontory observed through an intact tympanic membrane. Highly vascular areas of otospongiosis (early phase otosclerosis) have a reddish hue under otoscopic or microscopic examination. This finding is aptly named the Schwartze sign.
**Tuning Fork Examination**

This examination confirms the audiometric findings and determines if the patient would benefit from surgical intervention.

The Weber test is performed by placing a tuning fork on the patient’s forehead, bridge of nose, or upper incisors. The patient will perceive sound in the ear with a conductive hearing loss, and the ear with a greater conductive loss in the setting of bilateral disease. The test is sensitive to a 5 dB difference between ears.

The Rinne test evaluates the patient’s perceived loudness of air conduction compared with bone conduction. A 256 or 512 Hz tuning fork is first held 2 to 3 cm away from the external auditory canal (air conduction); then the base is placed firmly over the mastoid bone (bone conduction).

Bone conduction perceived louder than air conduction is termed a negative Rinne test. This is diagnostic for a conductive hearing loss. 256 Hz tuning fork is indicative of a 10 to 15 dB air bone gap. 512 Hz tuning fork reveals a 20 to 25 dB air bone gap. A patient with a conductive hearing loss is considered a surgical candidate for stapedotomy when he or she shows a negative Rinne test at 512 Hz. Air conduction perceived louder that bone conduction is termed a positive Rinne test.

**Audiometric Testing**

A standard audiometric battery should be performed including pure tone thresholds with air conduction and bone conduction, speech reception thresholds, word recognition scores, and immittance testing (tympanometry and acoustic reflexes).

On an audiogram, otosclerosis is seen as a unilateral or bilateral air bone gap, usually greater in the low frequencies. Speech recognition scores are typically as expected for the degree of hearing loss noted. Bone conduction may show a depression at 2000 Hz without a coinciding depression in air conduction, narrowing the air bone gap at that frequency. This is known as a Carhart notch and is common but not exclusive to otosclerosis (Fig. 1). This apparent depression of bone conduction is a result of an impedance mismatch of the otic capsule from stapes fixation. Coinciding SNHL is

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**Fig. 1.** Audiogram showing normal hearing of the left ear (X), a moderate conductive hearing loss in the right ear (O), and classic Carhartt’s Notch at 2K Hz. Acoustic reflexes are absent ipsilateral when testing the right ear, and absent contralateral when testing the left.
variable. In a retrospective study of 290 patients with over 10 years of follow-up, Ishai and colleagues showed approximately one-third of patients with otosclerosis will have clinically significant progression of SNHL greater than age-matched controls with normal ears.\textsuperscript{17}

Immitance testing of acoustic (stapedial) reflexes is an integral component of the audiometric workup. Ipsilateral acoustic reflexes are characteristically absent in the setting of stapedial fixation due to otosclerosis (see Fig. 1; Fig. 2). The on-off effect is indicative of early stapes fixation, shown by decreased impedance at the onset and the end of the stimulus. Intact acoustic reflexes can be indicative of early otosclerosis or direct the examiner toward another etiology of the conductive hearing loss such as SSCD or enlarged vestibular aqueduct (EVA). A recent retrospective study showed that of patients with conductive hearing loss and at least 1 detectable acoustic reflex, a nonossicular etiology was present in 52% of ears. They found that screening for third window symptoms (autophony, pulsatile tinnitus, or pressure-induced vertigo) in addition to acoustic reflexes carries a 94% positive predictive value of correctly diagnosing an ossicular etiology.\textsuperscript{18,19} A patient with conductive hearing loss and suprathreshold bone conduction and/or an acoustic reflex in at least 1 frequency with or without complaints of third window symptoms should have a CT of the temporal bones to rule out SSCD or EVA prior to middle ear exploration (Fig. 3).

**Radiologic Evaluation**

Although the diagnosis of otosclerosis is primarily a diagnosis obtained by history and audiometric testing, there are certain cases where radiology can help clarify the clinical picture. A CT scan can confirm the diagnosis of otosclerosis. An active otosclerotic focus will project as a focus of hypolucency at the fissula ante fenestrum (Fig. 4), or lucency can completely encircle the cochlea in more advanced otosclerosis - this is termed a halo sign. (Fig. 5). A CT scan is especially helpful to identify superior canal dehiscence in a patient with a conductive hearing loss and symptoms of a third window phenomena (Fig. 6). Although a CT scan is useful to confirm the diagnosis of

![Fig. 2. Audiogram showing a bilateral profound mixed hearing loss and absent acoustic reflexes bilaterally.](image-url)
otosclerosis or direct the clinician toward other pathology, a normal CT scan of the temporal bones does not rule out otosclerosis.

Finally, the role of stapedotomy is unclear in far-advanced otosclerosis with significant sensorineural deficit and a superimposed conductive component (see Fig. 3). The masking dilemma in this situation makes a reliable audiometric diagnosis difficult; thus discussion of the case with the audiology team proves valuable. The role of stapedotomy in far advanced otosclerosis should be considered on a case-by-case basis.

In summary, the clinical evaluation of otosclerosis relies on a careful history and audiometric testing to guide the practitioner to a treatment plan beneficial for the patient (Fig. 7).

Fig. 3. Audiogram showing normal hearing of the left ear (X) and a moderate low-frequency conductive hearing loss in the right ear (O). Suprathreshold right-sided bone conduction is suggestive of superior semicircular canal dehiscence. Acoustic reflexes are present bilaterally.

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Fig. 4. Axial CT of the right temporal bone. Foci of lucency at the fissula ante fenestrum (arrow).
Fig. 5. Axial CT of the left temporal bone. Multiple foci of active otosclerosis surrounding the cochlea and labyrinth (arrows). Halo Sign.

Fig. 6. CT right temporal bone without contrast showing superior semicircular canal dehiscence in 3 planes. (A) Coronal view. (B) Pöschl view. Pöschl images are cut in a plane perpendicular to the long axis of the temporal bone. This plane is 45° offset from both the coronal and sagittal plane. The superior semicircular canal will appear as a ring. (C) Stenver view; Stenver images are cut perpendicular to the Pöschl images and show a cross section of the superior bony cortex of the superior semicircular canal.
Fig. 7. Flow chart describing the clinical workup for conductive or mixed hearing loss.
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