Clinical Evaluation of the Patient with Otosclerosis

Michael F. Foster, DO*, Douglas D. Backous, MD

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.

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

Disclosure Statement: D.D. Backous has received honoraria for consulting with Medtronic, and Stryker. M.F. Foster has nothing to disclose.
Center for Hearing and Skull Base Surgery, Swedish Neuroscience Institute, 550 17th Avenue, Suite 540, Seattle, WA 98122, USA
* Corresponding author.
E-mail address: michaelfoster5@gmail.com

Otolaryngol Clin N Am ■ (2017) ■–■
https://doi.org/10.1016/j.otc.2017.11.004
0030-6665/17/

American populations exhibit an incidence of 0.5%, and the African American population have even fewer cases. Average prevalence is 0.3%.[8] 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.[9] 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.[10]

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).[11]

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).[12–14] 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.[1]