Impact of Imaging in Management of Otosclerosis

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KEYWORDS

• Otosclerosis • Computerized tomography • MRI

KEY POINTS

- Imaging is a useful adjunct to the clinical and audiometric information and is often critical to confirm the correct diagnosis and prevent potential complications.
- High-resolution computed tomography (HRCT) without contrast is the modality of choice for the demonstration of fenestral and retro-fenestral (cochlear) spongiotic lesions.
- Both HRCT and MRI are recommended before cochlear implantation surgery in patients with a far-advanced otosclerosis.

INTRODUCTION

Traditionally, imaging was not considered a requirement for the diagnosis of otosclerosis-related hearing loss (ORHL). Histologic changes typical to otosclerosis were described more than 100 years ago,[1–5] although were not found to correlate with the severity of conductive or sensorineural hearing loss[6]; the radiographic findings for otosclerosis were described more than 50 years ago.[7] Nevertheless, controversy exists regarding the correlation between imaging and the degree of hearing loss in otosclerosis.[8–18] Typical otosclerosis-related imaging is useful when evaluating patients with ORHL before primary as well as revision stapes surgery. Imaging is considered the standard of care in far-advanced otosclerosis before cochlear implantation.[8,11,14]

DISEASE OVERVIEW

The pathophysiologic hallmark of the fenestral subtype of otosclerosis is remodeling of the temporal bone that is primarily taking place in the area of the oval window,

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specifically in its anterior part, the fissula ante fenestram, which is a groove between the oval window and the cochleariform process. During the active (otospongiotic) stage of the disease, hypodense foci of bone can be identified in this area.[8] These foci will be replaced later by sclerotic bone in the nonactive (otosclerotic) stage of the disease that can progressively involve the footplate resulting in its thickening and fixation. This stage of the disease is manifested by progressive conductive hearing loss.[9] In 1% to 10% of the cases, a retro-fenestral subtype of the disease occurs with the disease involving the otic capsule, which might become demineralized, leading to far-advanced otosclerosis (defined by House and Sheehy[19] as longstanding hearing loss secondary to otosclerosis with an air conduction [AC] pure tone average of 85 dB or greater and no measurable bone conduction [BC]).[9,19] Diagnosis of the disease is typically based on history, physical examination, and characteristic audiometric findings.[8,9] Occasionally the course of otosclerosis might deviate from the classic presentation, especially in the retro-fenestral subtypes of the disease when mixed or even pure sensorineural hearing loss might occur.[8] The role of imaging in these challenging situations becomes more significant and is further discussed later in this article.

FENESTRAL AND RETRO-FENESTRAL OTOSCLEROSIS: IMAGING DIAGNOSIS

The most common manifestation of fenestral otosclerosis, especially in its spongiotic active stage, is demineralization of the fissula ante fenestram, anterior to the oval window (**Fig. 1**). In the nonactive otosclerosis, this area becomes sclerotic and in advanced stages of the disease this sclerosis can thicken and obliterate the oval window. The former stage of the disease is usually easier to detect, whereas the later becomes detectable only when significant otosclerotic bone causes irregularity and significant thickening of the otic capsule.[10] These lesions can be demonstrated on axial and coronal sections of high-resolution computed tomography (HRCT) without contrast, with a window level at 300 to 400 Hounsfield units (HU) and a width of 2000 to 3000 HU, the modality of choice for otosclerosis, with sensitivity varying from 34% to 91%.[10] A recent study demonstrated sensitivity higher than 90% in

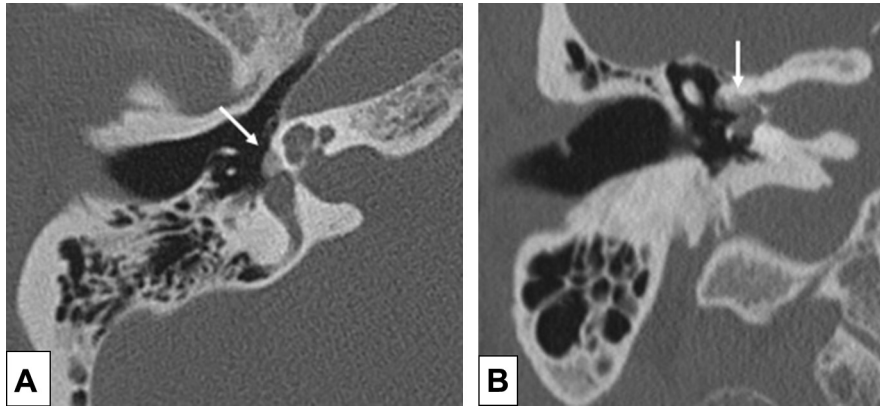

Fig. 1. Fenestral otosclerosis. Axial (*A*) and coronal (*B*) HRCT images of active, spongiotic stage of otosclerosis (*arrow*) with demineralization of the fissula ante fenestram, anterior to the oval window. (*Courtesy of* Simon Angeli, MD, Rita Ghose Bhatia, MD, University of Miami, Miami, FL)