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Update on the imaging diagnosis of otosclerosis☆

J. Gredilla Molinero[a,*], M. Mancheño Losa[b], N. Santamaría Guinea[a], N. Arévalo Galeano[a], M. Grande Bárez[a]

[a] *Unidad Central de Radiodiagnóstico, Hospital Universitario Infanta Leonor, Madrid, Spain*
[b] *Servicio de Otorrinolaringología, Hospital Universitario Infanta Leonor, Madrid, Spain*

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Abstract Otosclerosis is a primary osteodystrophy of the temporal bone that causes progressive conductive hearing loss. The diagnosis is generally clinical, but multidetector CT (MDCT), the imaging technique of choice, is sometimes necessary. The objective of this article is to systematically review the usefulness of imaging techniques for the diagnosis and postsurgical assessment of otosclerosis, fundamentally the role of MDCT, to decrease the surgical risk.

PALABRAS CLAVE
Otosclerosis;
Tomografía computarizada multidetector;
Resonancia magnética;
Prótesis de estribo

Actualización en el diagnóstico radiológico de la otosclerosis

Resumen La otosclerosis es una otodistrofia primaria del hueso temporal que produce una hipoacusia de transmisión progresiva. El diagnóstico es generalmente clínico, pero en ocasiones es necesaria la realización de una tomografía computarizada multidetector (TCMD), que es la técnica de imagen de elección. El objetivo de este artículo es realizar una actualización sistemática de la utilidad de las técnicas de imagen en el diagnóstico y la valoración posquirúrgica de la otosclerosis, fundamentalmente del papel de la TCMD, con el fin de disminuir el riesgo quirúrgico.

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* Corresponding author.
E-mail address: jgredmol@yahoo.es (J. Gredilla Molinero).

Introduction

Otosclerosis is a primary otodystrophy characterized by abnormalities in the otic capsule osseous remodeling of the temporal bone.[1–3] The otic capsule is made up of three layers of different ossification; among them, the intermedial layer typically shows endochondral ossification. Due to unknown causes, in patients with otosclerosis the endochondral bone is replaced by disorganized spongy bone, less dense and with a greater vascular component, which makes up the otospongiosis stage or active stage. As the disease evolves sometimes the pathologic spongy bone re-calcifies while showing a greater sclerosis component giving rise to otosclerosis or inactive stage.[1,4–6]

It is a genetic disease that is inherited by a dominant autosomal pattern, with incomplete penetrance and a variable clinical expression.[2,3,5,6] Damage is bilateral in 80–85 per cent of the patients, shows female predominance (2:1) and its incidence peak is between the second and fourth decades of life.[1–3,5,7] It is more frequent among caucasians,[5] with a prevalence of around 0.3–0.4 per cent.[2] Although there is no clear consensus, some studies[8] confirm that the location and extension of the otosclerotic foci condition the diagnosis, and in particular the type of hearing loss of these patients.

The main symptom of the disease is a progressive hearing loss, usually bilateral that can be associated or not with tinnitus.[1,3,7] Sometimes it can also occur as a mixed, or exceptionally, a pure sensorineural hearing loss.

Diagnosis has been classically based on the presence of a compatible clinical diagnosis in a young adult patient, with normal otoscopy and a familial history of otosclerosis (Table 1).[4]

To many ENT specialists there is no need to perform imaging modalities in typical cases[1,9,10] and confirmation diagnosis is performed during surgery, when the otosclerotic foci are observed in the oval window with fixation of the stirrup.[4,7] In atypical cases (Table 1) imaging modalities are necessary to establish preoperative confirmation diagnosis.[10,11]

Otosclerosis treatment is mainly surgical, through stapedotomy, whether partial or complete with the subsequent placement of stirrups prosthesis. In highly evolved cases, placing a cochlear implant is indicated.[12]

In this article we will assess the role of radiology in diagnosing otosclerosis, as well as its utility in diminishing surgical risk and in the diagnosis of postsurgical complications.

Table 1 Types of clinical presentation.

Typical symptoms	Atypical symptoms
Progressive hearing loss	Mixed or sensorineural hearing loss
Bilateral damage	Unilateral damage
Young adult	Children or advanced age
No stapedius reflex	Two-stage stapedius reflex
Familial history of otosclerosis	No familial history of otosclerosis

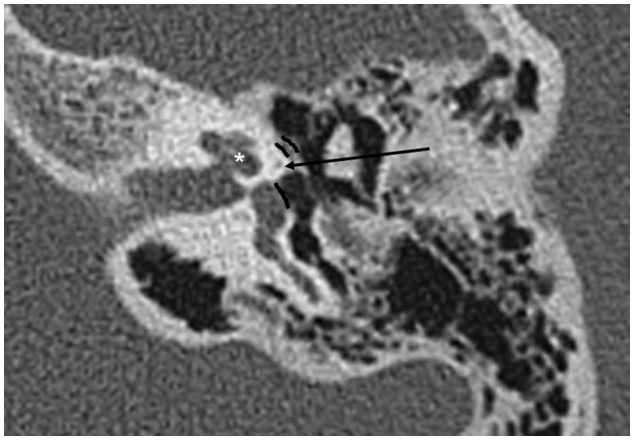

Figure 1 CT scanning of the petrous bone-basic anatomy on an axial plane with location of fissula ante fenestram (long, black arrow) and oval window (black line). Cochlea (*), horizontal segment of facial nerve (black parallel lines).

Radiographic findings. The role of the multidetector computed tomography

The multidetector computed tomography (MDCT) is the modality of choice for the radiologic diagnosis of otosclerosis.[4,11–15] Its sensitivity goes from 80 per cent to 95 per cent[4,8,11,12,14,16,17] in the most recent studies, thanks to the technological improvements of these machines.

False negatives are due to foci smaller than 1 mm and some lesions in sclerotic stages, their attenuation is similar to that of the normal adjacent bone.[1,4,11,18]

The preoperative MDCT allows us first to confirm diagnosis, by performing differential diagnosis with other conditions that also onset with conductive or mixed hearing losses, such as fixation of the non-otosclerotic ossicular chain, ossicular disconnection, superior semicircular canal dehiscence, dilated vestibular aqueduct, etc. It also allows us to identify factors of poor prognosis and anatomic abnormalities that can make surgery difficult, such as a dehiscent facial nerve or an obliterating footplate.

For its optimal characterization it is necessary to standardize image collection. Axial reconstructions perpendicular to the plane of the lateral semicircular canal are performed.

Axial slices are the most useful ones for the diagnosis of otosclerosis since the oval window, the *fissula ante fenestram* and the branches of the stirrup are better observed on this plane, and since the sensitivity for finding lesions is greater in fenestral forms[13,14,19] (Fig. 1).

Coronal slices allow us to assess a possible dehiscence of the horizontal segment of the facial nerve, the height of the oval window, the thickness of the stirrup footplate or the presence of superior semicircular canal dehiscence (Fig. 2).

Radiographic findings

Otosclerosis is divided radiologically into two types: fenestral otosclerosis and cochlear otosclerosis. In general, cochlear forms encompass the fenestral form so at present