

Update of cochlear densitometry as a radiological diagnosis of otosclerosis

Actualización de la densitometría coclear como diagnóstico radiológico de la otosclerosis

David Bejarano[1]
Juan Rolla[1]
Nebil Larrañaga[2]
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Summary
Otosclerosis is a disease of unknown etiology, frequently causing hearing loss as a result of osteodystrophy of the labyrinthine capsule that generates hearing loss due to fixation of the stapes, or to formation of foci of bone sclerosis at the endocochlear level. Diagnosis is generally clinical; however high-resolution computed tomography of the temporal bone is the imaging method of choice. The technique used is cochlear densitometry, which consists of the quantitative measurement of tissue density of specific areas in the affected osseous structures of the inner ear, by placing regions of interest (ROI).

Resumen
La otosclerosis es una enfermedad de etiología desconocida, que frecuentemente causa hipoacusia producto de una osteodistrofia de la cápsula laberíntica que genera la pérdida de audición por fijación del estribo o por formación de focos de esclerosis ósea endococlear. El diagnóstico es generalmente clínico; sin embargo, la tomografía computarizada de alta resolución del hueso temporal es el método de estudio imagenológico de elección. Además de la evaluación morfológica detallada de las estructuras óseas de la cápsula ótica la técnica utilizada es la densitometría coclear, la cual consiste en la medición cuantitativa de la densidad tisular de áreas específicas en las estructuras óseas del oído interno afectadas mediante la selección de regiones de interés (ROI).

Key words (MeSH)
Densitometry
Otosclerosis
Multidetector computed tomography
Hearing loss

Palabras clave (DeCS)
Densitometría
Otosclerosis
Tomografía computarizada multidetector
Pérdida auditiva

Introduction

Otosclerosis is an otodystrophy of inflammatory etiology that has not yet been fully elucidated, characterized by an alteration in the bone remodeling of the otic capsule of the temporal bone. It is classified as fenestral (85%) and cochlear (15%). It is an autosomal dominant genetic disease with incomplete penetrance and variable clinical expression (1, 2). The condition is bilateral in 80-85% of patients, with a predominance in females (2:1), and its peak incidence is between the second and fourth decades of life (1, 3). The main symptom of the disease is conductive or progressive transmission hearing loss, usually bilateral, which may or may not be associated with tinnitus. It can also present as mixed hearing loss or, exceptionally, pure sensorineural hearing loss if the cochlea is affected (3, 4).

Multidetector computed tomography (MDCT) is the radiological method of choice for evaluating the labyrinthine windows and the otic capsule (3). In addition to detailed morphological evaluation of the bony structures of the otic capsule, the technique used is cochlear densitometry, which consists of quantitative measurement of tissue density in specific areas of the affected bony structures of the inner ear, by defining regions of interest (ROI) (Figure 1), in order to diagnose otosclerosis early, subclassify it (fenestral and cochlear), evaluate some of its differential diagnoses, and enable a correct assessment useful for the surgical technique to be implemented, as well as helping to establish the postoperative prognosis and monitor the disease. The treatment of otosclerosis is purely surgical, involving partial or complete stapedectomy and the subsequent placement of a stapes prosthesis. In very severe cases, a cochlear implant may be indicated (1, 3).

Discussion

Although its origin is still unknown, explanatory models for the disease have been proposed. The onset of this condition is thought to be caused by foci of bone resorption generated by regional osteoclasts activated via inflammatory mediators. Secondly, there is a recruitment of lymphocytes, plasma cells, fibroblasts, and histiocytes. A relationship with type II collagen fibers, present in the remaining cartilaginous tissue of the rock, has also been suggested, which would cause an autoimmune reaction responsible for bone tissue resorption. Histopathologically, there are three phases: acute (osteoid tissue), subacute (osteoclastic remodeling), and chronic (sclerosis) (1, 3).

As mentioned, 85% of cases are of the fenestral type (Figures 2 to 6), which manifests as conductive hearing loss, while the remaining 15% are of the cochlear type (Figures 7 to 9), which will present with sensorineural

[1]Diagnostic imaging resident. Diagnostic Imaging Department, Centro de Educación Médica e Investigaciones Clínicas "Norberto Quirno" (CEMIC). Buenos Aires, Argentina.

[2]Diagnostic imaging specialist. Diagnostic Imaging Department, Centro de Educación Médica e Investigaciones Clínicas "Norberto Quirno" (CEMIC). Buenos Aires, Argentina.

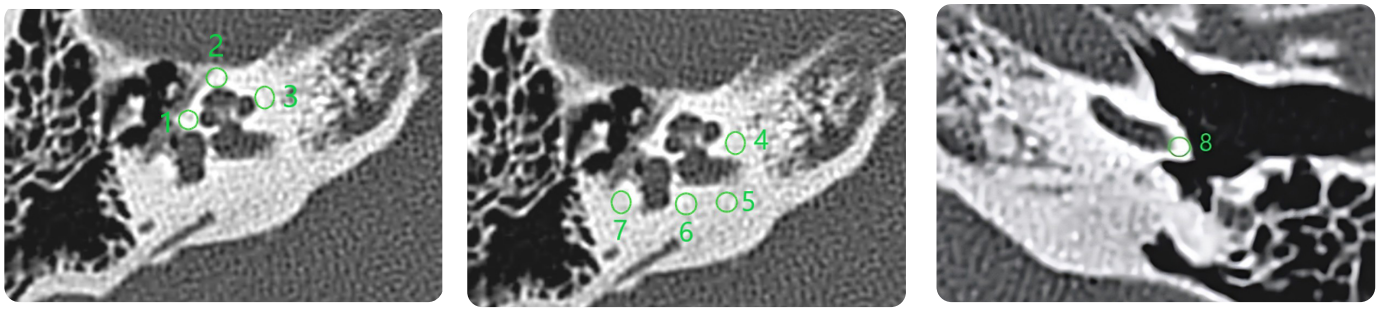


Figure 1. CT scan of the right ear, axial section. ROIs located in the structures of the inner ear that must be taken into account when performing the cochlear densitometry technique.

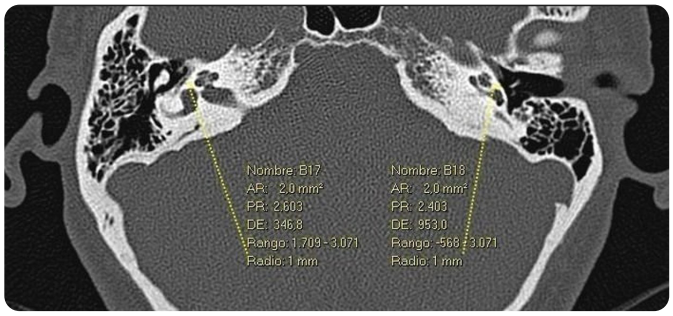


Figure 2. Patient without otosclerosis. Control patient: 2,314 and 2,584 HU.

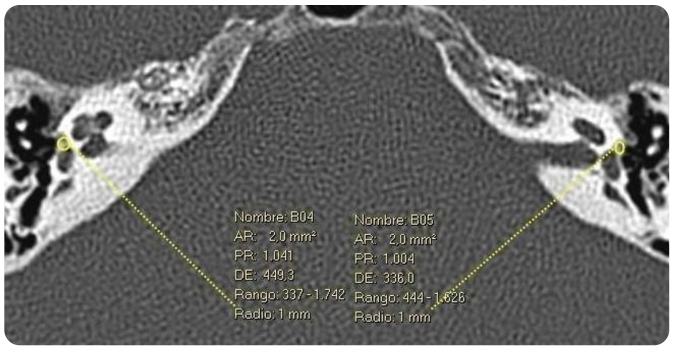


Figure 3. Patient with bilateral otosclerosis. Examples of measurements taken in a patient with otosclerosis compared to the control patient in Figure 2. Patient with bilateral fenestral otosclerosis: 1,041 and 1,004 HU.

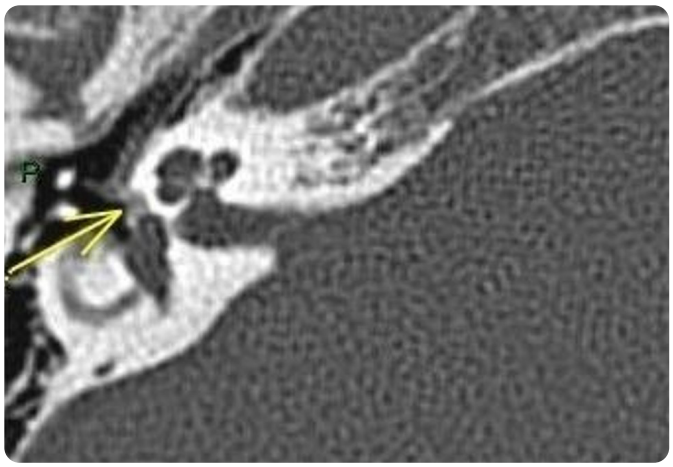


Figure 4. Patient with right otosclerosis. Focal image of low bone density in the right fissula ante fenestram (arrow).

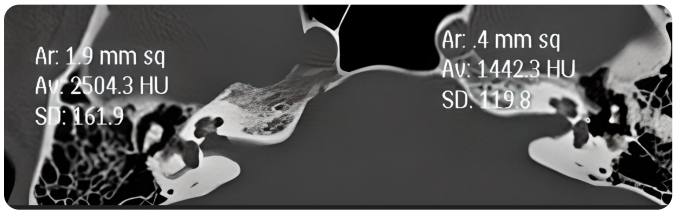


Figure 5. Patient with left otosclerosis. Focal image of low bone density in the left fissula ante fenestram.

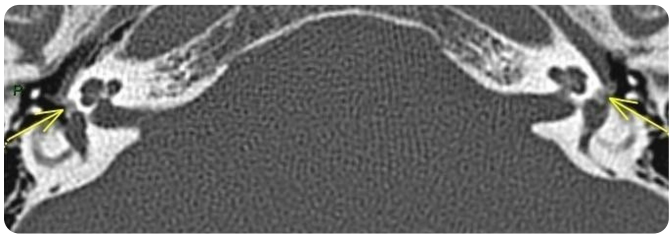


Figure 6. Patient with bilateral otosclerosis. Focal image of low bone density in both fissula ante fenestram (arrows).

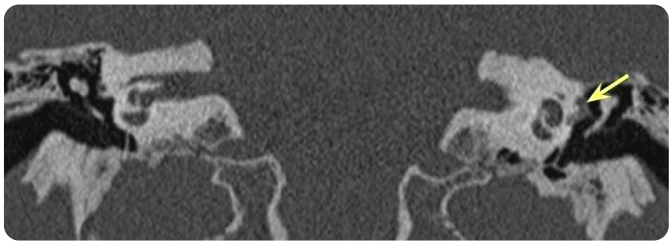


Figure 7. Patient with left otosclerosis. Focal image of low bone density in the left pericochlear region (arrow).

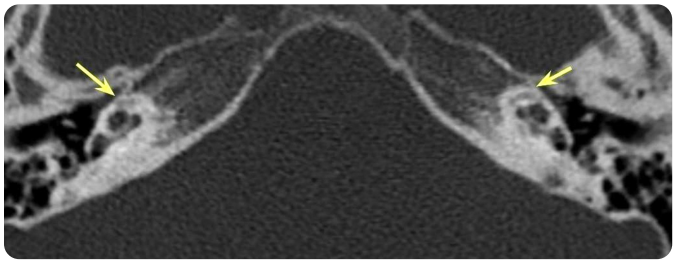


Figure 8. Patient with bilateral otosclerosis. Focal image of low bone density in the bilateral pericochlear region (arrows).

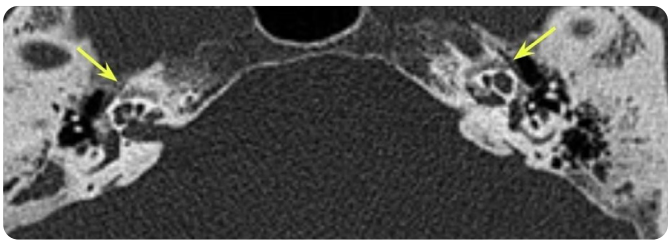


Figure 9. Patient with bilateral otosclerosis. Focal low bone density in the bilateral pericochlear region (arrows).

hearing loss. In the fenestral presentation, the final result of the alterations determines an abnormal fixation between the stapes and the oval window with calcification of the annular ligament, which can even lead to ankylosis of both structures (5). It should be noted that in 15% of cases of fenestral involvement, there is concomitant retrofenestral involvement. In the cochlear presentation, the toxic effect caused by lysosomal proteolytic-osteolytic enzymes on the cochlear structures, particularly on the spiral ligament, determines the onset of sensorineural hearing loss (5).

The method of choice for studying otosclerosis is high-resolution CT scanning of the temporal bone, and the primary objective is to demonstrate the existence and distribution of low-density otosclerosis foci (OSF) on CT scans at an early stage of the disease, findings that were not visible on CT scans. the primary objective is to demonstrate the existence and distribution of low-density otosclerosis foci (OSF) on CT at an early stage of the disease, findings that were not visible on conventional or simple helical CT (1, 2, 5, 8). Currently, the use of magnetic resonance imaging (MRI) for the diagnosis of otosclerosis is limited and generally occurs in the context of the first imaging study in patients with sensorineural hearing loss. The imaging findings described include areas of enhancement near the oval and round windows in the fenestral form, increased signal in T2-weighted sequences, and subtle enhancement in the pericochlear and perilabyrinthine region of the otic capsule in the retrofenestral form. However, the greatest utility of MRI in this disease lies in its ability to rule out differential diagnoses (4, 5).

Densitometry is a test that measures calcium and other minerals in the bones. It shows bone density, known as bone mass or bone density. The value of densitometry in otosclerosis consists of three points: diagnosis and therapy, research, and post-surgical evaluation (1, 4). The study of choice for the diagnosis of cochlear otosclerosis (5), which is based on the detection of changes in the contour of the capsule, is high-resolution CT with densitometric measurements, which allow alterations in the mineralization of the capsule to be recognized. It should be noted that there are similar changes that can be caused by other diseases, in addition to otosclerosis, such as its differential diagnoses: Paget's disease, osteogenesis imperfecta, and syphilis (3). The availability of a test that allows for the evaluation of the density and maturation of the otosclerotic process is extremely helpful in providing a reliable approach to the ongoing study of otosclerosis.

CT densitometry also helps to establish the progression of the disease by comparing the densitometric curves obtained before and after treatment. Based on this, it is possible to determine whether there has been a change in the extent of the disease and the maturation of the bone process (3, 4).

The technique consists of quantitatively measuring the tissue density of specific areas in the affected bone structures of the inner ear by placing eight regions of interest (ROI) with a diameter of 2 mm; although some authors argue that it is possible to use only the first five ROIs and calculate their average (PROMED) (6, 7) (Table 1).

Table 1. Normal densities in regions of interest

ROI	Densities	Standard deviation
1 Fissula ante fenestram (FAF)	2,170 UH	122.4
2 Cochlear apex	2,196 UH	145.9
3 Precochlear region 2.223	2,223 UH	135.4
4 Anterior margin of the IAC	2,132 UH	147.8
5 Posterior margin of the IAC	2,002 UH	135.3
6 Saccule	2,075 UH	123.7
7 Posterior semicircular canal	2,057 UH	145.6
8 Round window	(2,081 UH)	142.7
(PROMED) Average density recorded in the eight areas.	UH: 2,117	68.0

Source: adapted from Puiggrós et al. (6).

The parameters with the highest diagnostic sensitivity and specificity considered in the literature are the measurement of the mean density of the otic capsule (PROMED), fissula ante fenestram (ROI 1) (9) and precoclear region (ROI 3), and the stipulated threshold values that define the area under the curve for establishing the diagnosis are: PROMED: 1,980 HU (100% specificity) ROI 1: 1,750 HU (100% specificity) ROI 3: 2,114 HU (70% specificity). The images illustrate the densities obtained in a control patient.

Conclusion

Anatomical recognition of the structures involved in otosclerosis, the otic capsule of the temporal bone, and the technique for demonstrating them is essential. The study of these bone structures using the cochlear densitometry technique in CTD has a sensitivity of close to 90% for the radiological diagnosis of otosclerosis, which allows for pre-surgical planning and improves the postoperative prognosis.

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Correspondence

David Bejarano
Galván 4102, Código Postal C1431FWO
Ciudad Autónoma de Buenos Aires, Argentina
davidri_17@hotmail.com

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