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CT Densitometry of the Cochlear Capsule in Otosclerosis

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Otosclerosis is a primary focal disease of the labyrinthine capsule, which affects only humans. Otosclerosis develops in the enchondral layer of the otic capsule with destruction of the enchondral bone and formation of foci of lamellar bone [1]. These foci may progressively enlarge and extend to the endosteal and periosteal layers of the capsule. The site of predilection of the otosclerotic foci is the area just in front of the oval window (fissula ante fenestram). Hence, involvement of the adjacent footplate of the stapes occurs with consequent conductive hearing loss. Otosclerotic foci also occur in other areas of the labyrinthine capsule, particularly in the cochlea and at the fundus of the internal auditory canal. Involvement of the membranous labyrinth by direct invasion or by unknown factors is responsible for cell degeneration and consequent sensorineural or mixed hearing loss. Otosclerotic foci vary from a loose and irregular network of bony trabeculae containing large vascular lacunae, osteoblasts, and osteoclasts (immature or active foci) to a relatively avascular and acellular, much denser type of mature bone (inactive foci). The density of the normal otic capsule is fairly uniform, but its thickness varies. In the cochlea, the capsule is thicker along the medial aspect where the periosteal layer is wider and merges with the often sclerotic bone of the petrous pyramid.

In the past, multidirectional tomography has been used for the diagnosis in vivo of otosclerosis involving the cochlear capsule [2, 3]. With this technique changes in the outline of the capsule are more easily detected than changes in the actual density. As previously described [4], spongiotic foci appear as localized areas of thinning or interruption in the contour of the capsule. Large or confluent foci account for more extensive areas of demineralization where the contour of the capsule is partially or completely erased. Sclerotic foci are identified as areas of thickening and scalloping in outline of the capsule.

Comparison of multidirectional tomographic studies performed at various time intervals in order to evaluate the evolution of the disease is difficult and subjective due to the lack of densitometric measurements and too often dependent on extrinsic factors such as quality of the films. We report a new approach to the study of the cochlear capsule based on densitometric readings obtained in high-resolution CT images.

Cochlear otosclerosis can be recognized in most cases by high-definition axial and coronal CT images. However, routine technique does not allow a quantitative measurement of the changes. Reported here is a study of cochlear CT densitometry. The densitometric profile of the cochlear capsule was obtained in 10 ears with normal hearing and 50 ears in 27 patients with known clinical otosclerosis and progressive mixed-type hearing loss. Patients with hearing loss had decreased absorption of as much as 60% in comparison to the normal range. In 42 ears with abnormal densitometric profiles, the changes were visibly apparent on the CT images, whereas in eight others, the densitometric profile alone demonstrated decreased absorption.
Materials and Methods

The CT studies were performed on a GE CT/T 8800 unit using 1.5-mm collimation with 0.25 pixels, extended gray scale to 4000 H (+3000 to −1000), and the fast array processor (RDPCP 120), which permits a practical throughput for these studies. The scanner is recalibrated every 2 weeks to prevent a fluctuation of more than 1% in the system. The cochlea is identified and its edges localized in the scout digitized image. Six to eight sections at 1-mm increments are usually sufficient to cover the cochlear capsule. In order to shorten the examination time and prevent motion, we use fast-scanning rapid sequence (dynamic mode) and retrospective review targeting.

In the first examination, both axial (horizontal) and coronal sections are obtained to localize more precisely the site and extent of the involvement. The axial images are obtained with the scanner plane parallel to the canthomeatal line and no angulation of the gantry. The coronal sections are obtained with the patient supine or prone and the head hyperextended. The gantry is tilted until the scan plane is perpendicular to the canthomeatal plane.

Densitometric readings are taken in axial sections, and images in only this plane are obtained in follow-up studies. There are two reasons for using horizontal axial sections for densitometry: (1) This projection is the easiest to obtain and reproduce at a later date. A variation of less than 1 mm between comparable sections does not affect the results. (2) Histopathologic sections are usually obtained in this plane, and therefore it is easier to compare them with horizontal CT images.

Densitometric data are collected from two CT images corresponding to the histologic sections shown in figures 1 and 2. The lower section (fig. 1) passes through the basal turn of the cochlea and round window niche. The upper section (fig. 2) crosses the modiolus and exposes all three coils of the cochlea. The contour of the cochlear capsule shown in the two sections is scanned using the smallest cursor (0.25 × 0.25) for the lateral aspect of the cochlea and a larger cursor (0.25 × 0.50) for the thicker medial aspect of the capsule.

We start our measurements about 2.5 mm anterior to the round window, where five readings are obtained (fig. 3). After skipping the round window gap, measurements are obtained along the intercristal aspect of the basal turn. Fifteen readings are taken in the lower section. The upper or mid-modiolar section of the cochlea is then scanned starting medially above the internal auditory canal. From here the cursor is moved first anteriorly to the apex of the cochlea, then posteriorly along the lateral aspect of the cochlea to the anterior margin of the oval window (point 29), and finally along the anterior wall of the vestibule. Sixteen readings are obtained in this upper section, for a total of 31. A profile of the density of the capsule can be obtained by plotting the densitometric values vs. the 31 points where the readings are made.

Results

Normal Densitometric Profile

The densitometric profile of the normal cochlear capsules has been determined (fig. 4). Using the above described technique, readings were obtained in 10 ears with normal hearing. All 10 ears represent the uninvolved side of patients with unilateral chronic otitis media or acoustic neuroma. The age range of the subjects was 21–50 years.

The densitometric profiles of the 10 normal capsules were compared, and the mean value calculated for each of the 31 points from the 10 readings. A variation of ±10% was found from point 6 to point 27. From points 1 through 5 and 23 through 31, the variation was as high as ±15%. The increased variation in the region of the basal turn is certainly due to partial-volume averaging because the capsule is thinner in this area. The standard deviation for each point is indicated in figure 4.

The densitometric curve of the normal cochlear capsule shows a gradual decrease in density from 1600 H at the anterior aspect of the basal turn (point 1) to 1200 H at the margins of the round window (points 5 and 6). From this point, the density increases linearly to reach 2200 H, except for a deep notch at the anterior margin of the oval window or
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**Fig. 2.** A, Horizontal histologic section (reprinted from [1]). B, Corresponding axial CT section crossing modiolus and exposing all three coils of cochlea (arrow), vestibule (arrowhead), and footplate of stapes.

**Fig. 3.** A, Diagram showing 31 sites where densitometric data are obtained and recorded. RW = round window. B and C, CT sections showing actual sites of measurement.
in the area of the fissula ante fenestram (points 28 and 29), where the density dips to 1800 H.

Cochlear Otosclerosis

The term cochlear otosclerosis is used here to indicate involvement of the cochlear capsule by otosclerosis. No attempt was made to identify cases with so-called pure cochlear otosclerosis. A series of 50 ears from 27 patients with known clinical otosclerosis and progressive mixed type of hearing loss was reviewed.

In 42 ears, involvement of the cochlear capsule by otosclerosis was identified in the axial and coronal sections. The changes ranged from small and isolated foci of decreased density to diffuse demineralization of a large area of the capsule with complete dissolution of its contour [5–7].

A typical sign of cochlear otosclerosis was the formation of a double-ring effect due to confluent spongiotic foci within the thickness of the capsule. The band of intracapsular demineralization was in some cases limited to a segment of the capsule, but in others it followed almost the entire contour of the cochlea.

Densitometric profiles were obtained for the 50 ears of this series. Variations in density exceeding 15% from the normal in the basal turn and 10% in the rest of the cochlea were considered significant. In all 50 ears examined, the densitometric profile revealed changes of decreased absorption of as much as 60% in comparison to the normal range. In none of the cases were foci of increased density identified. This observation confirms the previously reported impression that sclerotic foci are not the result of increased density but rather of increased thickness due to apposition of mature otosclerotic bone [4]. In most of the cases with abnormal densitometric profile, the changes in the cochlear capsule were visible in the CT images. However, in eight cases the densitometric profile demonstrated areas of decreased absorption in the region of the apical coil that could not be recognized in the CT images, even retrospectively.

Representative Case Reports

Case 1

A 34-year-old man with a family history of otosclerosis complained of tinnitus and moderate bilateral hearing loss. The otologic diagnosis was clinical otosclerosis. The audiometric evaluation demonstrated a bilateral moderate mixed hearing loss. The conductive component was greater on the left. A multidirectional tomographic study revealed bilateral marginal thickening of the footplate of the stapes, but no evidence of capsular changes. A high-resolution CT horizontal study showed a normal cochlear capsule (figs. 5A and 5B). However, the densitometric profile indicated the presence of spongiotic foci of otosclerosis on the right in the region (points 20–30) of the middle and apical coils, as well as at the anterior aspect of the oval window (fig. 5C).

Case 2

A 50-year-old woman had progressive bilateral hearing loss from age 25. The audiometric evaluation demonstrated a moderate to severe mixed loss in both ears. Three multidirectional tomographic studies over a recent 8-year period demonstrated thickening of the footplate of both stapes and fairly severe spongiotic changes throughout both cochlear capsules. The high-resolution coronal (fig. 6A) and axial (figs. 6B and 6C) CT studies confirmed the presence of multiple foci of demineralization in the cochlear capsules. There was formation of a double-ring effect due to confluent spongiotic foci within the thickness of the capsule. The densitometric study of the left cochlear capsule (fig. 6D) revealed a diffuse demineralization most severe at points 15 to 22, along the medial aspect of the basal and middle coils. The promontory was only minimally involved.

Case 3

A 39-year-old man had bilateral severe hearing loss, worse on the
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Fig. 5.—Case 1. A and B, Axial CT sections of right ear demonstrate apparently normal cochlear capsule. C, Densitometric profile obtained at same level reveals presence of spongiotic foci in region of middle and apical coils. RW = round window.

left, and tinnitus. The audiogram confirmed a severe flat hearing loss with a definite conductive component. The hearing loss had been progressive for 20 years. The multidirectional tomographic and high-resolution axial (figs. 7A and 7B) and coronal (fig. 7C) CT studies demonstrated extensive demineralization of the cochlear capsules with complete dissolution of their contours. The densitometric profile of the right cochlear capsule (fig. 7D) showed a severe involvement of the promontory and a striking, almost flat demineralization of the rest of the cochlear capsule.

Case 4

A 50-year-old man had progressive mixed hearing loss in the right ear for 20 years. An early multidirectional tomographic study demonstrated thickening of the footplate of the right stapes and fairly severe spongiotic changes in the capsule of the basal turn of both cochleas. A right stapedectomy produced a good initial result. The otoscopic examination revealed bilateral reddening of the promontory seen through the transparent tympanic membranes (positive Schwartze sign). Tomograms 3 years later showed a wire prosthesis on the right in satisfactory position. The appearance of the cochlear capsules was unchanged. After another 12 years, the patient developed a recurrent conductive deficit in the right ear. In addition, the audiometric study revealed a high tone loss on the left side that was not present previously. Tomograms showed separation of the lateral end of the right prosthesis from the incus. There was considerable remineralization of the spongiotic foci in the cochlear capsules. High-resolution CT of the right ear with a densitometric profile (fig. 8) showed the presence of mixed foci in the region of the basal turn and spongiotic foci of otosclerosis in the middle and apical coils.
Discussion

The value of CT densitometry in otosclerosis consists of three factors: diagnosis, research, and posttreatment evaluation. High-resolution CT with densitometric measurements is the study of choice for the diagnosis of cochlear otosclerosis. The CT images are superior to the corresponding tomograms because of the improved contrast and, above all, by the lack of blurring, which is inherent to multidirectional tomography due to incomplete cancellation of structures outside the focal plane. With multidirectional tomography, the diagnosis of cochlear otosclerosis was based on the detection of changes in the outline of the capsule only; with CT, actual alterations in the mineralization of the capsule are recognizable. It should be pointed out, however, that similar changes may be caused by diseases other than otosclerosis, such as Paget disease, osteogenesis imperfecta, and syphilis.

The availability of a test that allows evaluation of the density and therefore the maturation of the otosclerotic process has opened a reliable approach to studying the evolution of otosclerosis in vivo. For this purpose, we have started a research project on otosclerosis that includes interval CT examinations in a large series of patients with combined stapes fixation and sensorineural hearing loss or pure sensorineural loss in which the diagnosis of cochlear otosclerosis was suspected on the basis of individual and family history, positive Schwartzte sign, and audiometric configuration.

CT densitometry constitutes an objective approach to establish the evolution of the disease after medical or surgical treatment. By comparing densitometric curves obtained before and after treatment, it will be possible to establish if any change has occurred in the extent of the disease and maturation of the process.

A software program aimed to simplify the collection and display of densitometric data is being developed by General Electric.
Fig. 7.—Case 3. Axial (A and B) and coronal (C) CT sections show severe and diffuse demineralization (arrowheads) of right cochlear capsule (C). Thickening of footplate of stapes (S), best seen in coronal image (C). D. Densitometric profile shows striking demineralization of cochlear capsule. RW = round window.

Fig. 8.—Case 4. Densitometric profile shows presence of mixed foci of otosclerosis in region of basal turn and of spongiotic foci in middle and apical coils.

REFERENCES