Cochlear Otosclerosis (Otospongiosis): CT Analysis with Audiometric Correlation

Cochlear otosclerosis (otospongiosis) is a progressive disorder characterized by sensorineural hearing loss (SNHL) (1). The diagnostic foci of demineralization within the otic capsule have been identified both with CT (2) and conventional multidirectional tomography (3, 4). We undertook to determine the extent of audiometric correlation with findings of CT.

MATERIALS AND METHODS

The CT scans of 60 patients (many of whom had SNHL) who had computed-tomographically confirmed fenestral otosclerosis (5) and those of an additional 30 individuals under 50 years of age who had SNHL and who were clinically suspected of having pure cochlear otosclerosis underwent detailed scrutiny in search of foci of demineralization in the otic capsule (otospongiosis). Abnormal bony sclerosis was also sought. Audiograms of these patients or the reports of the audiograms were studied to determine the degree of sensorineural hearing loss and the specific frequency levels involved. If multiple audiograms were available, they were evaluated to determine if the process was progressive.

All patients underwent scanning using the same General Electric 8800 CT/T machine: 1.5 mm thick coronal and axial images were obtained. Only a few of these patients could not tolerate the coronal position. Each image was targeted for maximal bony detail (ReView, General Electric).

RESULTS

Twelve patients were found to have one or more foci of demineralization within the otic capsule (otospongiosis) (Figure 1–4) (TABLE I). Eight of these were bilateral. All of these eight were highly symmetrical. There were five women and seven men who ranged in age from 21 to 74 years. All had predominantly sensorineural mixed hearing loss in the abnormal ears. Serial audiograms revealed progressive disease in 14 ears (eight patients). The audiometric frequency involved corresponded to the cochlear region which was described as normal in all cases. The apical turn was involved in eight of 20 ears, the middle turn in 14, and the basilar turn in 15. The internal auditory canal, an unusual site outside the otic capsule described by several previous authors, was involved in two (6–8). The semicircular canals were spared in all patients. All but one patient with foci of demineralization demonstrated by CT had fenestral disease as well.

Abnormal sclerosis of the cochlea is much more difficult to evaluate; we believe that there is striking variability in the thickness of the cochlear turns in the normal population. The normally thin basilar turn bordering the air-filled middle ear was the easiest to evaluate in this regard. This manifested as abnormal irregularity or generalized thickness (Fig. 5). Twelve patients (four who had bilateral disease) were diagnosed as being in the remineralized (healed) phase of the disorder using these criteria. All predictably had static high frequency loss. Two of these also definitely had abnormal generalized patchy sclerosis of the entire cochlea and internal auditory canal. Each of
This 38-year-old woman has bilateral progressive SNHL in all frequencies. Left middle ear exploration revealed fenestral disease. There was a positive Schwartz sign bilaterally. There is remarkable demineralization around the entire cochlea (long-stemmed arrows). The basilar turn and internal auditory canal are also involved. This was a symmetrical process. Figure 1a and 1b are of the left ear, and Figure 1c and 1d are of the right ear. A large round window hyperdense plaque is incidentally demonstrated (single arrowhead, Fig. 1d).

**DISCUSSION**

The inner ear, which is contained within the petrous portion of the temporal bone, consists of an osseous labyrinth (otic capsule) encasing a membranous labyrinth. The osseous labyrinth is subdivided into the vestibule, the semicircular canals and the cochlea. The membranous labyrinth contains an endolymphatic portion and a perilymphatic portion. The membranous portion of the cochlea consists of three tubular canals that run parallel to one another: the scala vestibuli, the cochlear duct (scala media) and the scala tympani. Therefore, the membranous labyrinth contains an endolymphatic space surrounded by a perilymphatic space. They are coiled together to form two and one-half helical turns approximately 32 mm in length. The two complete turns are referred to as the apical and middle while the remainder is the basilar turn (promontory) (9-11).

This 47-year-old woman has progressive SNHL in the higher frequencies, worse on the left side. A well-defined focus of demineralization is identified at the junction between the middle and basilar turn on both axial and coronal views (long-stemmed arrow, Fig. 2a and 2b) on the left side. A smaller focus in a similar location is noted on the right (long-stemmed arrow, Fig. 2c).
The scala vestibuli begins at the oval window and winds to the apical turn where it communicates via the helicotrema with the scala tympani, which subsequently terminates at the round window. They are filled with potassium-rich perilymph. The interposed cochlear duct (scala media) contains potassium-rich endolymph (9, 10). Reissner's membrane separates the cochlear duct from the scala vestibuli while the basilar membrane forms its boundary with the scala tympani. It is this latter membrane that bears the sensory apparatus for the transduction of sound energy, the organ of Corti (9, 11).

The basilar membrane widens progressively as it approaches the apex. In accordance with the principles of resonance there is maximal amplitude of vibration of high tones near the basal turn, for medium tones in the middle turn, and for low tones in the apical turn (11, 12).

A cochlear map is illustrated to show the approximate positions of different frequencies within the cochlea (Fig. 6). The highest audible frequency of approximately 20,000 Hz is associated with the extreme broad end of the membrane behind the round window. The middle point of the membrane subtends a frequency of roughly 2,000 Hz while the apical end at the helicotrema corresponds to a frequency of just 60 Hz (10).

Otosclerosis is considered by many to be a misnomer for what is more appropriately termed "otospongiosis progressiva insidiosa" or more simply, "otospongiosis" (5–7). It is a slowly progressive focal disorder of the otic capsule characterized by resorption of the endochondral (middle) layer. This disease is bilateral in 80%. Audiometric symmetry and a certain tendency toward histologic symmetry have been described (7, 8). These areas of resorption have been demonstrated to occur along the vascular channels within this layer. Eventually spongy vascular new

### Table I: Demineralization of the Otic Capsule

<table>
<thead>
<tr>
<th>Patient Age and Sex</th>
<th>Laterality</th>
<th>Turns Involved*</th>
<th>Frequencies</th>
</tr>
</thead>
<tbody>
<tr>
<td>38 F</td>
<td>Bilateral</td>
<td>A, M, B, IAC</td>
<td>All</td>
</tr>
<tr>
<td>50 F</td>
<td>Bilateral</td>
<td>M, B, IAC</td>
<td>Middle and high</td>
</tr>
<tr>
<td>47 F</td>
<td>Bilateral</td>
<td>M, B, (L &gt; R)</td>
<td>High &gt; middle</td>
</tr>
<tr>
<td>26 M</td>
<td>Bilateral</td>
<td>A, M, B</td>
<td>Sawtooth</td>
</tr>
<tr>
<td>37 M</td>
<td>Left</td>
<td>B &gt; M</td>
<td>Middle and high</td>
</tr>
<tr>
<td>52 M</td>
<td>Bilateral</td>
<td>A, B</td>
<td>Low &gt; high</td>
</tr>
<tr>
<td>64 M</td>
<td>Left</td>
<td>distal M</td>
<td>High</td>
</tr>
<tr>
<td>73 F</td>
<td>Bilateral</td>
<td>A</td>
<td>Low and high</td>
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<tr>
<td>74 M</td>
<td>Left</td>
<td>B</td>
<td>High</td>
</tr>
<tr>
<td>80 M</td>
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<td>M, B</td>
<td>Middle and high</td>
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<tr>
<td>56 M</td>
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</tr>
<tr>
<td>21 F</td>
<td>Bilateral</td>
<td>M &gt; B</td>
<td>Middle and high</td>
</tr>
</tbody>
</table>

* A = apical turn; M = middle turn; B = basilar turn; IAC = internal auditory canal.

**Figure 3**

- **a** and **b**. This 26-year-old man has progressive SNHL in a sawtooth pattern, *i.e.*, the involvement was at scattered frequencies ranging from low to high. Symmetrical foci of demineralization are identified in the apical and middle turns bilaterally on these sections (long-stemmed arrows). The basal turn was also involved (not illustrated).

**Figure 4**

- **a**, **b**, and **c**. This 52-year-old man has progressive low to middle frequency SNHL. There is well-defined demineralization involving the distal portion of the apical turn adjacent to the processus cochleariformis (long-stemmed arrows). This process is remarkably symmetrical (coronal views, Fig. 4a and 4b). Figure 4c is an axial section.
This 50-year-old woman has progressive SNHL in the lower frequencies. In the higher frequencies her loss is marked and static. Foci of demineralization were noted in the middle and basal turns bilaterally (not illustrated). This coronal section illustrates abnormal thickening of the basal turn (arrowheads), which presumably represents the healed phase of this disorder. The oval window is remarkably narrow.

CONCLUSION

The finding of one or more foci of demineralization in the otic capsule indicates progressive disease. These foci may be subtle and should be carefully sought.

The frequency at which hearing loss occurs audiometrically correlates with the level of cochlear involvement.

Areas of remineralization (healed phase) are difficult to diagnose except perhaps in the basilar turn.

CT provides an easily reproducible means of diagnosis, which may be of considerable value if sodium fluoride treatment is a possible treatment modality.

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References