Otosclerosis: Incidence of Positive Findings on High-Resolution Computed Tomography and Their Correlation to Audiological Test Data

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Objectives: Computed tomographic (CT) scanning with slices of 1 mm or more has not been sufficient to demonstrate otosclerotic foci in most cases to date.

Methods: We investigated the validity of CT scans with a 0.5-mm cubical scan technique, with and without planar reconstruction, and correlated these findings with audiological data. Forty-four temporal bone CT scans from 30 patients with conductive or mixed hearing loss were evaluated.

Results: Otosclerotic foci were visualized in 74% of the cases. With reconstruction at the workstation, the sensitivity increased to 85%. Whereas in fenestral otosclerosis a correlation was found between the size of the focus and the air-bone gap, no correlation was seen between the size of the focus and bone conduction thresholds with cochlear involvement. Otosclerotic foci in patients treated with sodium fluoride were smaller than those in patients without treatment. This finding may indicate a beneficial effect of sodium fluoride on otosclerotic growth.

Conclusions: High-resolution CT scans are a valid tool that can be used to confirm, localize, and determine the size of clinically suspected otosclerotic foci.

Key Words: audiometry, computed tomography, otosclerosis.

INTRODUCTION

Although otosclerosis has an estimated prevalence of 10% in the Caucasian population and was described histopathologically nearly 250 years ago, its diagnosis is, to this day, made clinically. A suspicion of otosclerosis is evoked by a combination of the history and audiometric findings (conductive hearing loss, absent stapedial reflexes, Carhart’s notch, and type A tympanogram). Confirmation of the diagnosis typically requires surgical middle ear exploration.

Fine-cut computed tomographic (CT) images have been proven to demonstrate otosclerotic foci around the oval and round windows, as well as the otic capsule. So far, the clinical role of CT scanning in otosclerosis remains controversial. The differential diagnosis of otosclerosis is extensive and encompasses entities from ossicular discontinuity to congenital fixation. High-resolution CT (HRCT) scanning may be helpful in differentiating these middle ear disorders and confirming the diagnosis of otosclerosis before operative treatment, thereby helping in surgical planning and patient counseling.

This study was intended to determine the accuracy of fine-cut CT scanning in identifying otosclerotic foci, and also to determine whether there is a correlation between the extent of otosclerosis and the degree of hearing loss (conductive and sensorineural).

METHODS

Between May 2001 and May 2003, we collected data on consecutive patients with normal findings on otoscopic examination and long-standing progressive unilateral or bilateral hearing loss suggestive of otosclerosis. Both genders and all age groups were included in the study after informed consent was obtained. Patients with congenital malformations, a history of chronic ear infections, previous surgery, or sudden hearing loss of the affected ear were excluded.

The initial workup for hearing loss included otoscopy, audiometry (pure tone and speech), a tympanogram, and stapedial reflex testing. The preoperative air and bone conduction thresholds for the speech frequencies of 500, 1,000, 2,000, and 4,000 Hz were recorded, and the air-bone gap (preoperative values) was calculated. All tests, including the HRCT scanning, were timed closely together. A total of 44 HRCT scans, with each ear counted as a single scan, from 30 patients were studied; because of the above-men-
tioned exclusion criteria, only 44 HRCT scans, instead of 60, were analyzed. The CT scans were obtained with identical protocols in all patients, and the images were read by the same experienced neuroradiologist in a single-blinded study design.

Eighteen of the 30 patients, who entered the study from November 2002 onward, had their HRCT scans read at an imaging computer workstation, as well as by printed films. The direct visualization of all three dimensions, as well as the free rotation of the image at the time of the reading, allowed a more accurate measurement of the extent of the otosclerotic foci. The results of the readings of the printed images and of the workstation-based images were compared. When abnormalities other than otosclerosis were detected on HRCT scanning, patients were excluded from the study. The HRCT scanner model used was an MX 8000 (Picker/Marconi, Cleveland, Ohio) 4-detector row helical HRCT scanner with a 2 × 0.5-mm collimation. Thin-section images were created in any desired plane from the initial axial cut on a computer workstation. Images were produced with a high-resolution bone algorithm and read with a window width of 4,000 Hounsfield units.

Initially, all 44 temporal bone HRCT scans from the 30 patients were read from printed films. Patients with positive results were divided into 3 groups depending on the size of the otosclerotic focus. In fenestral otosclerosis, group 1 included patients with small foci or visible otospongiosis limited to the fissula ante fenestram. The foci of the group 2 patients reached at least half of the diameter of the oval window niche and/or the cochleariform process. The foci of the group 3 patients reached at least half of the diameter of the oval window niche extending over the entire diameter of the oval window niche (Fig 1).
For cochlear involvement, the otosclerotic foci were divided in a similar manner. Group 1 patients had a singular spongiotic focus not exceeding the diameter of one cochlear turn. Group 3 patients presented with a spongiotic involvement of the entire otic capsule, and group 2 patients had an involvement of the cochlea with an extent between those of groups 1 and 3 (Fig 2).

Twenty-seven scans from 18 patients underwent additional reading and measurements on a computer workstation. The obtained results were compared with those of the printed films (Fig 3).

Sodium fluoride therapy was offered as a medical treatment option to patients with a sensorineural hearing loss component. Eleven patients had received 40 mg of sodium fluoride (Ossin, Grünenthal, Aachen, Germany) for at least 6 months before our investigation.

For statistical analysis, we used nonparametric tests, including Mann-Whitney, Kruskal-Wallis, Pearson, and Spearman \( \rho \) correlation tests.

RESULTS

Radiologic Findings. Of the 30 patients included in this study, 20 were female and 10 male. The mean age, independent of gender, was 49 years (range, 9 to 72 years).

A well-defined focus of otosclerosis was seen radiologically on 70.5% of the printed HRCT films (31 of 44); no otosclerotic foci were identified in the other 29.5% (13 of 44). The Table shows the classification...
of the otosclerotic foci and the corresponding pure tone averages. There was a significant correlation ($p = .005$) between the size of the fenestral otosclerotic focus and the air-bone gap (Figs 1, 4, and 5), but no correlation ($p > .1$) between the extent of cochlear involvement and the bone or air conduction levels (Figs 2 and 6). The size of the fenestral otosclerotic focus had no influence on the bone conduction levels ($p = .5$).

To evaluate the reliability of the printed images in comparison to the reconstructed data in the computer system, we compared 27 HRCT scans of the 18 patients from whom images were also available in a digitized format. Digital data were not available from all patients. By excluding the older HRCT scans (non-digitized), we decreased the number of ears analyzed from 44 to 27. The positive findings on the CT films with fenestral otosclerosis (20 of 27; 74%) and the additional cochlear involvements (9 of 27; 33%) were confirmed at the workstation, with the exception of 1 patient. This patient did not present with radio-

**CORRELATION BETWEEN EXTENT OF OTOSCLEROSIS AND HEARING**

<table>
<thead>
<tr>
<th>Group</th>
<th>No. of Ears (n = 44)</th>
<th>Mean Air Conduction (dB)</th>
<th>Mean Bone Conduction (dB)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fenestral otosclerosis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No focus</td>
<td>13</td>
<td>29</td>
<td>22</td>
</tr>
<tr>
<td>1</td>
<td>13</td>
<td>50</td>
<td>29</td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td>47</td>
<td>27</td>
</tr>
<tr>
<td>3</td>
<td>6</td>
<td>71</td>
<td>39</td>
</tr>
<tr>
<td>Cochlear involvement</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No focus</td>
<td>30</td>
<td>42</td>
<td>21</td>
</tr>
<tr>
<td>1</td>
<td>7</td>
<td>47</td>
<td>30</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>51</td>
<td>39</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>78</td>
<td>50</td>
</tr>
</tbody>
</table>

logically visible cochlear involvement, but received an initial diagnosis of mild audiological cochlear involvement (group 1). Another 3 of 7 cases, previously read as negative from the printed film, did show a fenestral otosclerotic focus at the workstation. Three of 18 HRCT scans negative for cochlear involvement were shown to have visible cochlear involvement. All patients with cochlear involvement had fenestral otosclerotic foci. The detection rate for otosclerotic changes increased by 11% (from 74% to 85% for fenestral involvement and from 33% to 44% for cochlear involvement) when a computer-aided analysis was used instead of manual reading of the film. The measurements at the workstation showed 0.5-mm to 10-mm foci in all patients, whereas 16 cases had foci as large as 2.5 mm and 5 cases as large as 3.5 mm (Fig 3). There was a very high correlation ($p = 1.5E-6$) between the subjective grouping on the CT films (groups 1 to 3) and the digital measurements of focus size. The size of the focus was independent of the age of the patient ($p > .2$).

**Audiological Findings.** In general, the size of the focus correlated with the corresponding air-bone gap (Fig 5), but there was no correlation between the size of the focus and the degree of sensorineural hearing loss. However, when the cochlea was involved, the mean bone conduction thresholds tended to be worse ($p = .059$). In conclusion, it seems that a longer fenestral otosclerotic focus has greater likelihood of involving the cochlea.

**Fluoride Therapy.** Eleven of the 18 patients with inner ear impairment had received sodium fluoride (Ossin) for at least 6 months before our HRCT investigation. The otosclerotic foci in the fluoride group were found to be significantly smaller ($p = .01$) than
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Fig 4. Air and bone conduction thresholds in fenestral otosclerosis (corresponding to Fig 1). A) Group 1. B) Group 2. C) Group 3.

those of the patients who were not medically treated. However, the audiological results (mean bone conduction threshold, 23 versus 24 dB; mean air conduction threshold, 46 versus 49 dB) of the groups were equal.

DISCUSSION

Radiologic Findings. The radiologic detection of an otosclerotic focus depends upon the method used: manual reading or computer-aided analysis of the HRCT scans. On manual reading of the HRCT scans, fenestral otosclerosis was found in 70% to 74% of cases. There was a 70.5% detection rate on the printed films in 44 ears of 30 patients, and otosclerotic foci were found in 74% of the printed films in 27 ears of the 18 patients who had CT scans stored digitally. This value is comparable to those obtained in other studies; however, it is clinically unsatisfying consid-

Fig 5. Mean air-bone gap for different sizes of fenestral otosclerotic foci. Note clear correlation of size of focus and air-bone gap.
Fig 6. Air and bone conduction thresholds in cochlear otosclerosis (corresponding to Fig 2). Note that there is no correlation between size of focus and bone conduction thresholds. A) Group 1. B) Group 2. C, D) Group 3.

Two studies by Mafee et al. showed that correct positioning of the temporal bone is important for evaluation of otosclerotic changes along the long axis of the oval window. The CT resolution used by Mafee et al. did not allow for detection of an otosclerotic focus of less than 2 to 3 mm on axial CT scanning. In our series, otosclerotic foci as small as 1 mm became visible, because 0.5-mm cuts and reconstruction were used.

An HRCT scan can also give false-negative results. During operation, Swartz et al. found in 2 patients an otosclerotic plaque that was not visible on CT scans. Two of our patients showed dense otosclerotic changes of the anterior oval window at surgery in the presence of negative radiologic findings. A re-evaluation of the CT scans at the workstation showed a prominent cochleariform process but no otosclerotic lesions. The false-negative radiologic image may be due to the fact that on CT scanning, the inactive form
of otosclerotic lesions has the same density as the surrounding bone and therefore remains undetected, whereas lesions from the active otospongiotic form are more visible because their density is lower than that of the otic capsule. In addition, there are other disorders, such as ankylosis or fibrous changes of the annular ligament, as described by Cherukupally et al.\(^5\) that can cause significant air-bone gaps without being detected on CT scans. In a study by Grayeli et al.\(^6\) measurements of bone density were significantly lower in 10 patients with otosclerosis than in controls. Four of 20 ears (20\%) did not show hypodensities in the area of the fissula ante fenestram. This may explain why 15\% of our otosclerosis patients had a normal-appearing CT scan. All of these patients had the smallest of the air-bone gaps.

High-resolution CT scanning with multiplanar reconstruction is, in the authors' opinion, valuable for a precise preoperative evaluation and may be used to exclude patients who have a high surgical risk.

**Audiological Findings.** There is still controversy regarding the correlation between the level of hearing and cochlear involvement in otosclerosis. Swartz et al.\(^7\) and Güneri et al.\(^8\) found a clear correlation between the location of the otosclerotic focus around the cochlea and the degree of sensorineural hearing loss. Schuknecht and Barber\(^9\) and Derks et al.\(^10\) reported, on the contrary, that the size and location of the focus did not influence the bone conduction threshold. Kimomizu et al.\(^11\) investigated the correlation between CT scanning and audiometry in Japanese patients with otosclerosis. They showed, by using a 1-mm scan, that the size of the fenestral focus is proportional to the air-bone gap and that the extent of cochlear involvement correlates with the bone conduction levels. Racial differences were implicated in the lower detection rate for otosclerotic foci of only 54\% in their patients. In the present study, a direct relationship was found between the size of the fenestral focus and the air-bone gap, but not between cochlear involvement and sensorineural hearing loss. The theory that otosclerotic foci extending into the cochlear endosteum cause significant sensorineural hearing loss was reviewed by Nelson and Hinojosa\(^12\); there was no clear correlation. Four patients of our study were found to have at least moderate cochlear involvement with radiologically close proximity to the endosteum without sensorineural hearing loss. However, in comparing the two CT scans of patients with severe cochlear involvement (group 3; Fig 2C,D) and their corresponding audiograms (Fig 6C,D), the hypothesis of endosteal involvement and sensorineural hearing loss may be supported, but clearly reaches the limitations of the method used.

**Fluoride Therapy.** Fluoride therapy has been proposed as a possible treatment of the cochlear involvement of otosclerosis. Vartiainen and Vartiainen\(^13,14\) showed a beneficial effect of fluoride on the natural course of hearing, although the prevalence of otosclerosis was not affected; this discrepancy supports a genetic origin for otosclerosis. Van Den Bogaert et al.\(^15\) further evaluated the genetic background of otosclerosis; two new genes, loci OTSC1 and OTSC2, were linked to the disease.

Fluoride therapy was used in 11 patients with sensorineural hearing loss. Whereas the audiological results (mean bone conduction threshold, 23 versus 24 dB; mean air conduction threshold, 46 versus 49 dB) were equal with and without treatment, the otosclerotic foci in the fluoride group were found to be significantly smaller (\(p = .01\)). These findings may indicate that patients with extensive cochlear involvement do benefit from fluoride therapy, in the sense that a progression of sensorineural hearing loss was prevented. A similar conclusion was reached by Derks et al.\(^10\)

**CONCLUSIONS**

1. High-resolution CT scanning with 0.5-mm slices permits visualization, localization, and quantification of fenestral otosclerotic foci in 74\% of cases in which otosclerosis is clinically suspected. Foci of less than 1 mm can be visualized as long as they have an active, spongiform appearance.

2. Multiplanar reconstruction increases the rate of detection of otosclerotic foci by 11\% (from 74\% to 85\%).

3. The present study shows that there is a correlation between the size of the otosclerotic focus and the extent of conductive hearing loss, but no correlation between otosclerotic cochlear involvement and the extent of sensorineural hearing loss.

4. It seems that fluoride treatment may limit the growth of active, spongiotic foci in otosclerosis.

5. The indication for 0.5-mm HRCT scanning in clinical otosclerosis supplemented by workstation reading of the images should be rather liberal, because the determination of the size of the focus, the involvement of the oval window, and the precise definition of the cochlear involvement have a great influence on the surgical planning and preoperative counseling of the patient.

6. One should be aware that significant cochlear involvement by otosclerosis may remain undetected if only patients with a sensorineural hearing loss are subjected to a radiologic investigation.
REFERENCES


