Significance of High Resolution CT Scan in Otosclerosis

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Abstract

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

Methods: We investigated the validity of CT scans with a 0.75-1mm cubical scan technique, with and without planar reconstruction, and correlated these findings with audiologicai data and to determine its usefulness to predict hearing deterioration and surgical difficulties. Fifty one CT scans from 29 patients with conductive or mixed hearing loss were evaluated.

Results: Otosclerotic foci were visualized in 68.9% of the cases. Whereas in fenestral otosclerosis a correlation was found between the size of the focus and the airbone gap, no correlation was seen between the size of the focus and bone conduction thresholds with cochlear involvement. Among 31 operative ears, 19 were classified as positive for otosclerosis and this was confirmed during surgery, 7 were classified as negative for otosclerosis and during surgery, 3 of them (42.8%) proved to be cases of otosclerosis and they were could infra-radiologic cases. The remaining 5 cases were doubtful according to preoperative CT scans. Four of them (80%) were found to have otosclerotic foci during surgery.

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. Doubtful and negative preoperative cases should alert surgeons to the possibility of intraoperative complications and possibility of postoperative sensorineural hearing loss.

Key Words: Otosclerosis – High resolution CT.

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. Otosclerosis is a primary focal osteodystrophy of the human otic capsule with an initial clinical manifestation of slowly progressive conductive or mixed hearing loss, tinnitus and vestibular symptoms. As the disease progresses, inner ear symptoms, such as sensorineural hearing loss and tinnitus, may manifest themselves, constituting a dynamic clinical feature [1]. Due to the close anatomic relationship between the vestibule and the stapes footplate, vertiginous symptoms can occur in patients with otosclerosis [3].

Diagnosis of otosclerosis depends on medical history, physical examination and audiological tests (such as conductive hearing loss, absent stapedial reflexes, Carhart’s notch, and type A tympanogram). Previously, audiological measurements such as air-bone gap (ABG) or types of conductive hearing loss have been used to reveal the disease progression [3,4]. Since its introduction into otology, the computed tomographic (CT) scan has been shown to be a useful instrument for diagnosis of otosclerosis [5]. Many studies found a reliability of more than 90% in the diagnosis of otosclerosis [6,7]. This reliability was limited in cases of isolated otosclerotic foci or by superficial foci too small to be visualized on the CT scan [8]. 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 [8]. Otosclerosis can be divided into 2 types, fenestral and retro-fenestral, according to the topography of the lesions. The first is characterized by lesions preferentially located in the lateral wall of the otic capsule, i.e. the regions of the round and oval windows, promontory, and tympanic segment of the fallopian canal. The second is presence of otosclerotic foci in other regions of the labyrinthine capsule, including the pericochlear region, the semicircular canals, internal acoustic meatus, vestibule, and
cochlear and vestibular aqueducts [9]. The area called the fossula ante fenestram, located just before the oval window, is the most common site involved with otosclerotic foci.

The main goal of this study was to specify the reliability of high-resolution CT scans (HRCT scans) in the diagnosis of otosclerosis based on comparing radiologic and surgical findings. In addition, to discuss the role of preoperative CT scans in preventing intraoperative incidents and their importance in localizing hypodensities for hearing prognosis.

### Material and Methods

Between June 2009 and September 2010, 29 patients (51 ears) with clinical diagnosis of otosclerosis were collected at Fayoum University Hospital, Fayoum Egypt. Both genders and all age groups were included. Patients with congenital malformations, a history of chronic ear infections, previous ipsilateral ear surgery, or sudden hearing loss of the affected ear were excluded. All of the patients were presented with progressive unilateral or bilateral hearing loss with or without tinnitus, no history of ear discharge or trauma. Seven of them had done stapedial surgery in one ear sometime ago and consequently, these operated ears were excluded from the study.

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.

The enrolled patients were preoperatively scanned with high resolution computer tomography (HRCT) at private radiological centers counting each ear as a single scan. All CT images were obtained using a ten-detector-row helical CT scanner (Semens, SOMATOM Sensation 10) with 0.75-mm. slice thickness. The scan conditions were 120kV, 180mA, 0.75mm/s, 512-512 matrix, and the field of view (FOV) was 22cm. The window width was 4000 Hounsfield units (HU) and the window level was 700 HU. According to classification of otosclerosis into fenestral and retrofenestral, certain regions of interest were under focus during performing HRCT scanning. These regions include: Anterior to intersection of anterior stapes arch and stapes footplate, posterior to intersection of posterior stapes arch and stapes footplate, lateral to the middle turn of the cochlea, lateral to the area between the apex of the cochlea and its middle turn, apex of the cochlea, anteromedial to the middle turn of the cochlea, anteromedial to the basal turn of the cochlea, internal auditory meatus and the round window. Computed tomographic scan results with respect to the presence of otosclerosis were classified as positive, negative, or doubtful: Positive when hypodense foci were seen around the otic capsule (Fig. 1); negative in the absence of any hypodensity or other temporal bone finding, including tympanosclerosis, ossicular malformation, and superior semicircular canal dehiscence (Fig. 2); and doubtful in cases of minimal hypodense foci such as an isolated thickening of the anterior part of the footplate or a triangular widening of the anterior branch of the stapes (Fig. 3). 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. Group 3 contained patients with large otosclerotic foci extending over the entire diameter of the oval window niche (Fig. 6). 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 group 1 and group 3. The direct visualization of all three dimensions, as well as the free rotation of the image at the time of the scanning, allowed a more accurate measurement of the extent of the otosclerotic foci at the imaging computer workstation.

Pure tone audiometry was done for every patient preoperatively and 2 months after surgery. Air conduction was recorded at 0.125, 0.25, 0.5, 1, 2, 4 and 8 kHz and the maximum air conduction values for each frequency were 85, 105, 110, 110, 110, 110 and 105 decibels hearing level (dB HL), respectively. Bone conduction was recorded at 0.25, 0.5, 1, 2 and 4 kHz and the maximum bone conduction values for each frequency were 35, 50, 65, 65 and 60 dB HL, respectively. The mean air conduction threshold and the mean bone conduction threshold were computed as the averages of the thresholds at the speech frequencies of 0.5, 1, 2, 4 kHz. The mean air-bone gap was defined as the mean air conduction threshold minus the mean bone conduction threshold. Conductive hearing loss (CHL) was diagnosed if the mean air-bone gap was more than 15 dB HL and the mean bone
conduction threshold was less than 25 dB HL. If the mean air-bone gap was less than 15 dB HL and the mean bone conduction threshold was more than 25 dB HL, sensorineural hearing loss (SNHL) was diagnosed. When the mean air-bone gap was more than 15 dB HL and the mean bone conduction threshold was more than 25 dB HL, the diagnosis was mixed hearing loss (MHL).

Results

Radiologic findings: Of the 29 patients included in this study, 20 were female and 9 male. The mean age, independent of gender, was 31.7 years (range, 25 to 53 years). A well-defined focus of otosclerosis was seen radiologically in 68.9% of the printed HRCT films (35 of 51); no otosclerotic foci were identified in the other 24.4% (12 of 51) and doubtful result in the remaining 6.7% (4 of 51).

Regarding the relation between the otosclerotic focus and the preoperative pure tone average, there was a significant correlation ($p=0.05$) between the size of the fenestral otosclerotic focus and the air-bone gap, but no correlation ($p>0.1$) between the
extent of cochlear involvement and the bone or air conduction levels. The size of the fenestral otosclerotic focus was found statistically to have no influence on the bone conduction levels.

The measurement of otosclerotic foci at the workstation showed 0.5-mm to 10-mm foci in all positive patients, whereas 17 cases had foci as large as 2.5mm and 3 cases as large as 3.5mm. There was a very high correlation 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 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 ($p=.039$). In conclusion, it seems that a longer fenestral otosclerotic focus has greater likelihood of involving the cochlea.

Table (1): Correlation between the extent of otosclerosis and hearing.

<table>
<thead>
<tr>
<th>Group</th>
<th>Number of ears n=51</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>21</td>
<td>29</td>
<td>22</td>
</tr>
<tr>
<td>1</td>
<td>20</td>
<td>50</td>
<td>29</td>
</tr>
<tr>
<td>2</td>
<td>9</td>
<td>47</td>
<td>27</td>
</tr>
<tr>
<td>3</td>
<td>1</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>18</td>
<td>47</td>
<td>30</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>51</td>
<td>39</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
<td>78</td>
<td>50</td>
</tr>
</tbody>
</table>

Surgical findings: The decision to perform a surgical middle ear exploration was based upon the history of the pathology, clinical reports, and the needs of the patient in terms of hearing. Not all patients agreed the surgical treatment. Among 51 scanned ears, only 31 ears were operated. Nineteen ears of them were classified preoperatively as positive, seven as negative and the remaining five as doubtful.

All ears with positive CT scan results had a positive diagnosis of otosclerosis during surgery. Among 5 ears with doubtful hypodensity, during surgery, we found 4 with otosclerotic foci (80%) and in 1 a minor malformation of the anterior branch of the stapes. Among 7 negative CT scans, we found 4 cases with variations of the ossicular chain or incidents of the stapes. These included narrow oval window niche, incus malformation and fracture of the footplate. The remaining 3 cases (42.8%) were positively diagnosed as otosclerosis during surgery and were defined as infraradiologic forms (i.e., a patient with negative CT scan presenting a positive surgical otosclerosis).

Table (2): Locations of otosclerotic hypodensities across all CT scans.

<table>
<thead>
<tr>
<th>Location of the otosclerotic focus</th>
<th>% of CT scans</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated or associated AFH</td>
<td>68.5%</td>
</tr>
<tr>
<td>Isolated thickening footplate</td>
<td>1.9%</td>
</tr>
<tr>
<td>Hypodensity extended to endosteum</td>
<td>2.4%</td>
</tr>
<tr>
<td>Isolated pericochlear hypodensity</td>
<td>2.6%</td>
</tr>
<tr>
<td>IACH</td>
<td>0%</td>
</tr>
<tr>
<td>Ossification of round window</td>
<td>0.1%</td>
</tr>
<tr>
<td>Hypodensity of vestibule</td>
<td>0%</td>
</tr>
<tr>
<td>No hypodensities</td>
<td>24.4%</td>
</tr>
</tbody>
</table>

AFH: Anterior fenestral hypodensity.
IACH: Internal auditory canal hypodensity.

Discussion

Otosclerosis is a disease involving bone dystrophy. Histologically, bone is absorbed by osteoclastic activity, and new bone is deposited by osteocytes. In the active stage, the lesions contain disorganized bone rich in osteocytes with low mineralization and highly vascularized connective tissue [10]. The lesions appear demineralized, and thus they are theoretically visible in CT because their density is lower than that of the surrounding bone. Demineralized lesions may present anywhere from small and isolated foci to diffuse demineralization. The small and isolated foci are difficult to detect by visual analysis of CT results.

Demineralized lesions can be found using CT only when obvious diffuse lesions such as “double-ring sign” appear, but such cases account for only a small proportion of otosclerosis. Most otosclerosis patients show no pathological findings in CT. The location and extent of demineralized lesions vary. Some lesions appear as sclerotic lesions which are less vascular and more solid, with only a few small marrow spaces present and a considerable reduction in cellular activity [9]. It is difficult to
find sclerotic lesions both because of their small size and because their density is close to that of normal bone [11].

On manual reading of the HRCT scans, fenestral otosclerosis was found in 68.9%. This value is comparable to those obtained in other studies; Swartz et al. [9], found that 26 of 35 patients (74%) had fenestral otosclerosis on CT scans analyzed in 2 planes: Axial and coronal (both non-cuboidal). Reconstruction was considered unnecessary. Nearly 20 years later. Shin et al. [12], found a fenestral otosclerotic focus on axial CT scans in 119 of 153 patients with otosclerosis (78%). Two studies by Mafee et al. [11,13] 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 3mm on axial CT scanning. In this series, otosclerotic foci as small as 1mm became visible, because 0.75-1mm cuts and reconstruction were used.

An HRCT scan can also give false-negative results. During operation, Swartz et al. [9] 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 reevaluation 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.

Swartz et al. [9] and Guneri et al. [14] found a clear correlation between the location of the otosclerotic focus around the cochlea and the degree of sensorineural hearing loss. Schuknecht and Barber [3] and Derks et al. [15] reported on the contrary that, the size and location of the focus did not influence the bone conduction threshold.

Kiyomizu et al. [16] investigated the correlation between CT scanning and audiometry in Japanese patients with otosclerosis. They showed, by using a 1mm 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 [17] but there was no clear correlation. Four patients of this study were found to have at least moderate cochlear involvement with radiologically close proximity to the endosteum without sensorineural hearing loss. Fluoride therapy has been proposed as a possible treatment of the cochlear involvement of otosclerosis. Vartiainen and Vartiainen [18] showed a beneficial effect of fluoride on the natural course of hearing. Naumann et al. [8], used the fluoride therapy in 11 otosclerotic patients with sensorineural hearing loss and in comparison with other patients suffering from the same condition but not subjected to fluoride therapy, they found that the otosclerotic foci in the fluoride group were significantly smaller ($p=.01$). Unfortunately, fluoride therapy is not applied in our country.

Incidents involving the stapes footplate were significantly more frequent when the CT scan was doubtful or negative. In a previous study, Shin et al. [6] found the same results with a significant risk ($p=0.04$) of complication of the stapes footplate in cases of infraradiologic forms. Infraradiologic forms correspond to superficial histopathologic otosclerotic foci usually associated with reduced stapes fixation and a higher risk of fragile footplate. Thus, in cases of negative or doubtful HRCT scan otosclerosis, the surgeons have to be alerted to the higher risk of encountering a stapes footplate problem.

Conclusion:

High-resolution CT scanning with 0.75-1mm slices permits visualization and localization of fenestral otosclerotic foci in 68.9% of cases in which otosclerosis is clinically suspected. 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. Preoperative HRCT of clinically suspected cases of otosclerosis allows foresee anatomic difficulties (i.e., narrow oval window, facial dehiscence) and look for other associated diagnoses (i.e., malleus fixation, ossicular malformation), identify a risk of mobilized stapes footplate in cases of negative or doubtful images, and inform patients of the possible risk of postoperative sensorineural hearing loss in cases of per-
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icochlear, internal auditory canal, or round window involvement.

References


