Superior semicircular canal dehiscence: prevalence in a population with clinical suspected otosclerosis-type hearing loss

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Abstract. Superior semicircular canal dehiscence: prevalence in a population with clinical suspected otosclerosis-type hearing loss. Superior semicircular canal dehiscence (SSCD) can present with a variety of symptoms that can be predominantly auditory, predominantly vestibular or both. It can mimic a wide range of otological disorders, in particular otosclerosis-like stapes fixation. Our study revealed that, in 5.3% of our patients with clinically suspected otosclerosis, SSCD was detected in high-resolution multi-detector computed tomography (HRMDCT) of the temporal bone. We therefore emphasise the value of HRMDCT with reconstructions in the plane of the superior semicircular canal and perpendicular to the superior semicircular canal in the diagnostic work-up of each patient with a tentative diagnosis of otosclerosis-type stapes fixation. Where there are doubts, VEMP testing should be performed. We believe that a number of unexplained complications after an uneventful stapedotomy procedure might be explained by a pre-operatively undetected dehiscent superior semicircular canal and therefore unwarranted surgery. The typical feature of the conductive hearing loss due to SSCD compared to otosclerosis is an air-bone gap which is much more prominent at low frequencies than at high frequencies.

Introduction

Superior semicircular canal dehiscence (SSCD) was first described by Minor et al.1 in 1998 as a clinical and radiological entity which typically consists of vestibular symptoms induced by sound or changes in the middle ear or intracranial pressure. It is therefore also known as Minor’s syndrome. As clinicians’ experience with SSCD accrued, it became clear that SSCD may not only induce vestibular symptoms, but that it can also lead to a variety of symptoms that can be either predominantly auditory, predominantly vestibular or both. More specifically, a subgroup of patients was described that presented with an apparent conductive hearing loss in the absence of major vestibular symptoms, therefore convincingly mimicking a condition of ossicular-fixation-like otosclerosis.2,3 If SSCD remains undetected, many of these patients are liable to undergo middle ear surgery without benefit, or worse.

In the current paper, our primary objective is to assess the prevalence of SSCD in a population of patients with a tentative diagnosis of stapes fixation based on clinical and audiological examination. Our secondary goal is to investigate whether there are significant differences between the pure-tone audiograms of the surgically confirmed otosclerosis population (with a real air-bone gap) and the SSCD population with a virtual air-bone gap in order to identify possible differential diagnostic criteria.

Methods

A retrospective review was conducted of all patients attending an otological consultation at our department between March 2005 and June 2008. On the basis of case history, clinical examination and audiological testing, 114 patients (150 ears) were tentatively diagnosed with stapes fixation due to otosclerosis. The audiological work-up included pure-tone audiometry (air and bone conduction), tympanometry and acoustic reflex (stapedial reflex) measurement. All patients had at least two pure-tone audiometric examinations prior to surgery.
In our department all patients with suspected otosclerosis undergo high-resolution, multi-detector computed tomography of the temporal bone (HRMDCT), with specific attention being paid to SSCD prior to surgery. The raw data are also reformatted in the plane of, and perpendicular to, the superior semicircular canal. All tomographies were evaluated by the same neuroradiologist (FG). The diagnosis of radiological SSCD was only withheld when the absence of bone was obvious in both planes.

Vestibular evoked myogenic potential (VEMP) testing was also performed on all subjects diagnosed with SSCD.

Results

In our population of 114 patients (150 ears), six patients (5.3%) were found to have probable SSCD based on the imaging studies (MDCT). Three of these patients have a bilateral dehiscence, so we have a total of 9 SSCD ears out of these 150 ears (6.0%).

Figure 1 depicts a typical image of SSCD as found in one of these patients.

The typical symptoms and clinical findings of our patients with SSCD are summarised in Table 1. As these patients were suspected of having otosclerosis-type stapes fixation, the most common complaint was hearing loss, which was present in all cases. Two out of the six patients complained of a ‘blocked ear’ (aural fullness). These two patients also mentioned tinnitus. When asked specifically about vestibular complaints, only one patient had a history of mild disequilibrium and imbalance.

Micro-otoscopy was normal in all but one ear, which showed a retraction of Shrapnell’s membrane. Table 2 presents an overview of the results of the audiological examination.

Pure tone audiometry revealed a conductive hearing loss with a significant air-bone gap (ABG) in all 6 patients (9 ears) (Figure 2). Table 3a summarises the mean air-bone gap at 250 Hz, 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz in the ears with SSCD versus the ears with proven otosclerosis-type stapes fixation. The average air-bone gap in the lower frequencies (250, 500 and 1000 Hz) was 23 dB, and 10 dB in the high-frequency range (2000 and 4000 Hz).
Superior semicircular canal dehiscence

Table 3b summarises the bone conduction thresholds at 250 Hz, 500 Hz and 1000 Hz.

Tympanometry was normal (type A according to Jerger & Jerger) in all but one ear (the one with retraction of Shrapnell’s membrane), which showed a type C diagram.

Acoustical (stapedial) reflexes were absent in all but two ears (77.8%).

All patients without SSCD on HRMD CT underwent a laser-assisted stapedotomy with interposition of a Kurz àWengen titanium clip prosthesis™. All patients were operated by the same surgeon (GF). The results of this series will be discussed in a forthcoming paper. The averages of the pre-operative air-bone gaps in this group are also given in Table 3a. The pre-operative bone conduction thresholds are listed in Table 3b. The average of the ABG in the lower frequencies (250, 500 and 1000 Hz) was 36 dB; in the high frequencies the average ABG was 19 dB.

In patients with diagnosed SSCD, a more extensive audiological evaluation was performed. This revealed that Hennebert’s sign (vertigo and nystagmus induced by pressure in a blocked meatus acusticus externus) was present in one patient (17%). VEMP testing was performed in all ears. All had positive findings, meaning that the VEMP was present with a normal or high amplitude.

Discussion

As experience with SSCD accrues, it is emerging that it is not such a rare condition. In a microscopic study performed on 1000 temporal bones at the Johns Hopkins University in 2000,
SSCD was found in 0.7% of the examined specimens. However, because of the variability of its presenting symptoms, SSCD often remains a challenging diagnosis. Especially when it mimics a well-known otologic entity, such as otosclerosis, the possibility of SSCD is often overlooked, leading to unnecessary surgery and probably complications.

The present study reveals that, in our population of 150 ears (114 patients) with a clinical tentative diagnosis of otosclerosis-type stapes fixation, 9 ears (6%) had in fact SSCD as demonstrated with HRMDCT.

Pure tone audiometry indicated, in all 9 cases, a conductive/pseudo-conductive hearing loss with a significant air-bone gap and a distinct Carhart’s notch at 2000 Hz. Recent research supports the ‘third window theory’ to explain the virtual air-bone gap in this subgroup of patients. According to this theory, the dehiscence in the wall of the superior canal acts as a third mobile window, which shunts the air-conducted sound away from the cochlea – thus elevating the air conduction thresholds – and increases the difference in impedance between the scala tympani and scala vestibuli, therefore improving thresholds for bone-conducted sound. However, it remains unclear why some patients with SSCD predominantly have auditory symptoms, whereas others present with only vestibular complaints and a third group with both auditory and vestibular symptoms. Further research in this area should be performed to elucidate the pathophysiology of SSCD further. Van Spauwen et al. have reported that a posterior semicircular canal dehiscence can present...
with similar symptoms. They too stress the importance of CT and MRI imaging and VEMP testing.

In the 5 ears diagnosed with SSCD, tympanometry revealed a Type A diagram (Jerger & Jerger) in all but one ear, which showed a Type C. Stapedial reflexes were absent in all but two cases. Zhou et al.\(^7\) found in their series of 26 patients with SSCD that acoustic reflexes were present in 89% of the cases. Mickulec et al.\(^2\) also noted that acoustic reflexes were also present in 3 out of 4 ears with SSCD that had not undergone stapes surgery.

So seven out of our nine ears with radiologically apparent SSCD had audiological test results that were fully compatible with stapes fixation due to otosclerosis. The presence of a possible SSCD is suggested by radiological findings. The most suitable imaging technique for detecting SSCD is high-resolution multidetector computed tomography (HRMDCT) of the temporal bone with 0.6 mm collimation (or less) in the axial plane. Images should subsequently be reformatted to include cuts in the plane of the superior semicircular canal (Pöschl’s incidence) and perpendicular to this plane (Stenver’s incidence).\(^4,10\) The radiological diagnosis of probable SSCD is retained only when the dehiscence is apparent in both reconstruction series. HRMDCT alone, however, is not sufficient to establish a positive diagnosis of SSCD since there might be a very thin bony layer covering the membranous superior semicircular canal (SSC) that is too thin to be detected by HRMDCT. Radiologists should therefore describe a possible or probable dehiscent SSC, to be confirmed by clinical evidence, unless – of course – the superior semicircular canal is blatantly uncovered, as is the case in our subjects.

As our study revealed that, in 5.3% of the patients with a tentative diagnosis of otosclerosis-type stapes fixation based on clinical and audiological examination, a manifestly dehiscent superior canal was diagnosed on HRMDCT, it is clear that this is a subgroup of patients that should not be overlooked. It is our considered opinion that undetected SSCD might explain a certain number of “dead ears” after an uneventful procedure (0.5-1% of all stapedotomy cases according to literature). As Van Spauwen et al.\(^7\) point out, the unexpected dead ear might be attributed to a perilymph “gusher”, similar to those cases where a direct connection exists between the internal auditory canal and the scala tympani of the basal turn of the cochlea. Indeed, the “third window” allows for direct transmission of pressure changes in the cavum cranii to the perilymphatic space, causing oozing of perilymph or even a “gusher”. Ruptures of inner ear membranes might therefore occur as well, leading to irreversible damage. We therefore emphasise the importance of pre-operative HRMDCT of the temporal bone with reformating as described earlier in the diagnostic work-up of each patient with a tentative diagnosis of otosclerosis-type stapes fixation before surgery is considered.

In their paper on revision stapedectomy, Battista et al.\(^11\) state that “patients with persistent conductive hearing loss after stapedectomy may also have an unrecognized superior semicircular canal dehiscence” and that “if an SSCD is found on CT, revision stapedectomy should not be performed”. A course of events such as that described by Battista et al.\(^11\) is to be avoided and stresses our point that HRMDCT of the temporal bone should be

### Table 3a

Mean Air-Bone Gap at 250, 500, 1000, 2000 and 4000 Hz in ears with SSCD versus ears with otosclerosis-type stapes fixation

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>SSCD (n = 9)</th>
<th>Otosclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>250</td>
<td>35 dB (SD 7.1)</td>
<td>43 dB (SD 9.7)</td>
</tr>
<tr>
<td>500</td>
<td>21 dB (SD 8.1)</td>
<td>35 dB (SD 11.1)</td>
</tr>
<tr>
<td>1000</td>
<td>14 dB (SD 7.8)</td>
<td>30 dB (SD 11.7)</td>
</tr>
<tr>
<td>2000</td>
<td>1 dB (SD 2.2)</td>
<td>10 dB (SD 11.4)</td>
</tr>
<tr>
<td>4000</td>
<td>12 dB (SD 8.3)</td>
<td>28 dB (SD 14.7)</td>
</tr>
</tbody>
</table>

### Table 3b

Mean Bone Conduction Thresholds at 250, 500 and 1000 Hz in ears with SSCD versus ears with otosclerosis-type stapes fixation

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>SSCD (n = 9)</th>
<th>Otosclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>250</td>
<td>8 dB (SD 5.8)</td>
<td>16 dB (SD 10.6)</td>
</tr>
<tr>
<td>500</td>
<td>17 dB (SD 10.8)</td>
<td>28 dB (SD 12.6)</td>
</tr>
<tr>
<td>1000</td>
<td>12 dB (SD 9.2)</td>
<td>32 dB (SD 13.5)</td>
</tr>
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</table>
performed prior to primary surgery and not afterwards.

In all ears with obvious SSCD diagnosed on HRMDC, a VEMP was performed. In all these ears, a normal VEMP with normal or even high amplitude was recorded. This illustrates the value of VEMP testing in the diagnostic work-up, as positive VEMP testing excludes important middle-ear pathology such as otosclerosis-type stapes fixation. Consequently, we have adopted VEMP testing in the diagnostic work-up of otosclerosis, especially when the radiological findings indicate a possible SSCD.

According to contemporary literature, the typical features of the “conductive” hearing loss in ears with SSCD are: bone conduction thresholds that are better than 0 dB in the lower frequencies and a more prominent low-frequency air-bone gap. In our study, this was clearly not the case: the bone conduction thresholds were not better than 0 dB in the lower frequencies in any of our cases. The mean air-bone gap was larger in the lower frequencies (250, 500 and 1000 Hz) compared to the mean air-bone gap in the higher frequencies (2000 and 4000 Hz) in both the SSCD group and the otosclerosis group. However, the relative difference between the air-bone gaps in the lower and the higher frequencies was more prominent in the group with SSCD. The latter characteristic might therefore be considered as an indication of possible SSCD.

White et al. suggest that vibration-induced nystagmus may be a sensitive screening test in the office setting for detecting SSCD. We have no experience with this test, but we plan to add it to our testing protocol.

Conclusions

Superior semicircular canal dehiscence (SSCD) can present with a variety of symptoms that can be either predominantly auditory, predominantly vestibular or both. It can mimic a wide range of otological disorders, in particular otosclerosis-like, stapes fixation. Our study revealed that, in 5.3% of our patients with clinically suspected otosclerosis, SSCD was detected on HRMDC of the temporal bone. We therefore emphasize the value of HRMDC with reconstructions in the plane of the superior semicircular canal and perpendicular to the superior semicircular canal in the diagnostic work-up of each patient with a tentative diagnosis of otosclerosis-type stapes fixation. Where there is doubt, VEMP testing should be performed. We believe that a number of unexplained complications after stapedotomy procedure might be explained by a pre-operatively undetected dehiscent superior semicircular canal and therefore unwarranted surgery.

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References


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