Pre- and postoperative CT appearance of superior semicircular canal dehiscence syndrome

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KEYWORDS
Ear; Vertigo; Deafness

Abstract We report here three cases of patients with superior semicircular canal dehiscence syndrome, prospectively monitored pre- and postoperatively. The computed tomography (CT) examination was performed in 0.6 mm slices with multiplanar reconstructions in the plane of the superior semicircular canal. All the patients also had an audiogram and a vestibular evoked myogenic potentials (VEMP) test. Preoperatively, all the patients had dehiscence of more than 3 mm. Conductive hearing loss for frequencies of less than 1000 Hz was present in all cases. The VEMP test showed stimulation thresholds lowered to less than 80 dB on the affected side. One case of bilateral involvement in the CT image corresponded to unilateral functional impairment. After plugging surgery, all the patients’ auditory and/or vestibular systems were improved. In one case, the reconstruction could not be visualised in CT owing to the radiotransparency of certain materials used in the surgical procedure (wax, fascia, cartilage).

Abbreviations

SSCDS superior semicircular canal dehiscence syndrome
SSC superior semicircular canal
CT computed tomography
MPR multiplanar reconstruction
VEMP vestibular evoked myogenic potential
RE right ear
LE left ear

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Pre- and postoperative CT appearance of SSCDS

Introduction

SSCDS was described for the first time in 1998 by Lloyd Minor [1]. This rare entity, which has an estimated prevalence of 0.5%, denotes a set of clinical, vestibular and cochlear symptoms associated with the presence of dehiscence, a covering defect in the roof of the bony wall of the SSC. The presence of this bone defect is the cause of the formation of a ‘third window’ in the inner ear, the physiopathological consequences of which are dissipation of the acoustic energy of sound waves, abnormal mobility of the endolymph, and reduction in the threshold of bone conduction [2]. These phenomena are seen clinically as pseudo-conductive hearing loss, vertigo and autophony, symptoms which make this condition a differential diagnosis for otospongiosis [3–5]. The exact aetiology is unknown [1—5]. In CT, the dehiscence on the roof of the SSC can be seen correctly (Fig. 1) using thin slices centred on the temporal bones, with oblique coronal MPR in the plane of the SSC, known as the plane of Pöschl [6]. The condition can be treated with surgery when the symptoms are severe and interfere greatly with patients’ everyday life [7]. The principle of surgery is to plug the dehiscence [8], aiming to block the extra window with wax, and then cover the operative site with muscular fascia, powdered bone, a chip of bone, or auricular cartilage (Fig. 2). We report three cases of patients with SSCDS who underwent surgery, showing the pre- and postoperative CT appearances and comparing them with the audiograms and VEMP data [9,10].

Case 1

Mrs C., aged 52, had had sensations of vertigo for the past 15 years, which occurred particularly when coughing. Questioning found that there had been a head injury a few days before the symptoms started to appear. The patient also complained of hearing loss and a feeling that the right ear was ‘full’. Otoscopic examination of the tympana was normal. The audiogram showed right unilateral conductive deafness for frequencies below 1000 Hz, and a Carhart notch at 2000 Hz. A stapedius reflex test could not be carried out as it triggered the onset of vertigo. A CT scan showed no otospongiosis but the presence of a large 3.5 mm dehiscence in the roof of the right SSC (Fig. 3a). The left SSC was normal. It was decided to surgically plug the dehiscence. The result was postoperative reduction in vertigo and closure of the air-bone gap on the audiogram (Table 1). The stapedius reflexes were present, with no vertigo. The postoperative CT verified reconstruction of the bone (Fig. 3b).

Table 1 Pre- and postoperative audiograms and vestibular evoked myogenic potential (VEMP) tests of cases 1, 2 and 3.

<table>
<thead>
<tr>
<th>Case</th>
<th>Preoperative gap (dB)</th>
<th>Postoperative gap (dB)</th>
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<td></td>
<td>RE</td>
<td>LE</td>
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<tr>
<td>Case 1</td>
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<td>Case 3</td>
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Values in decibels of the pre- and postoperative air-bone gaps on the audiograms. RE: right ear; LE: left ear.

Figure 1. Oblique coronal MPR of a superior semicircular canal (SSC) in the plane of Pöschl: a: axial CT slice through the SSC. The white lines indicate the oblique coronal plane to be reconstructed; b: CT slice in the Pöschl plane. A large dehiscence is visible (black arrow).

Figure 2. Expected postoperative appearance after plugging a superior semicircular canal (SSC) dehiscence. Plugging wax closes the dehiscence. The roof of the SSC is then covered by a layer of muscular fascia, a shaving of auricular cartilage and/or a shaving of the temporal bone obtained in situ.
Case 2

Mrs D., aged 60, had complained for the past year of vertigo triggered by coughing and sneezing. During examination, horizontal rotatory nystagmus upwards and towards the right was found during the Valsalva manoeuvre (the Tullio phenomenon). The tympana were normal. The audiogram showed unilateral right conductive deafness (Table 1) for frequencies lower than 1000 Hz, a Carhart notch at 4000 Hz and an increase in bone conduction thresholds of 10 dB. The stapedius reflexes were normal, whilst inducing them triggered vertigo on the right. The CT scan showed wide bilateral dehiscence in the roofs of the SSCs, of 4 mm on the right and 3 mm on the left (Fig. 4a and b). VEMP changes occurred only on the right side (Table 2). Plugging surgery was performed on the right side only. Postoperatively the sensations of vertigo were reduced. The audiogram showed closure of the air-bone gap on the right, whilst the amplitude of the VEMP electrical responses had decreased (Tables 1 and 2). The postoperative CT scan confirmed bone coverage of the canal treated (Fig. 4c).

Case 3

Mrs P., aged 43, had been complaining for 2 years of vertigo triggered by low frequency sounds, for example by the noise of a passing train. On questioning it was found that she had fallen down a staircase just before the symptoms started to appear. She also had autophony, with the impression of hearing her own footsteps resonate, and bilateral tinnitus. Otoscopic examination of the tympana was normal. The audiogram showed bilateral conductive deafness (Table 1) at frequencies below 1000 Hz. The stapedius reflexes were present, but triggered an episode of vertigo. A CT scan showed wide bilateral dehiscence of the SSCs assessed to be 4.5 mm on both the right and left sides (Fig. 5a and b). The amplitude of the N23 and P13 wave responses was increased bilaterally on testing the VEMP, with bilateral reduction in the stimulation thresholds to below 80 dB (Table 2). It was decided to plug the dehiscences, first the right side, then the left. The patient reported a marked improvement in her symptoms following these two surgical procedures. Postoperative audiograms showed bilateral closure of the air-bone gap and normal stapedius reflexes with no sensation of vertigo. The amplitude and response thresholds were normal on both the right and left on the VEMP tests (Tables 1 and 2). On the other hand, the reconstruction could not be visualised in the postoperative CT scan (Fig. 5c and d). All the materials used — wax, muscular fascia, covering cartilage — were radiotransparent.

### Table 2 Pre- and postoperative audiograms and vestibular evoked myogenic potential (VEMP) tests of cases 1, 2 and 3.

<table>
<thead>
<tr>
<th>Air-bone gap (dB)</th>
<th>Preoperative</th>
<th>Postoperative</th>
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<tr>
<td></td>
<td>RE</td>
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<td>Case 3</td>
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Values in decibels of the stimulation thresholds on the VEMP tests.
RE: right ear; LE: left ear.

### Discussion

The role of CT in the management of patients with SSCDS is central. During diagnosis it helps associate clinical symptoms with an anatomical lesion by objectifying the presence of dehiscence in the roof of the SSC. The main differential diagnosis for this condition is otospongiosis [1,3,4], which has a similar clinical picture. In both these conditions, the audiogram shows conductive hearing loss, while with imaging they can be correctly differentiated from each other [4,5]. A CT scan can also be used to measure the extent of the exposed area, which is a criterion for operability. Dehiscences of more than 3 mm are considered to be operable [8],
Figure 4.  CT appearance in the Pöschl plane of the superior semicircular canals (SSCs) of case 2: a and b: preoperative CT of the right (a) and left (b) SSCs. There is wide, bilateral CT dehiscence (black arrows). Only the right ear is clinically affected; c: postoperative CT of the right SSC. Bone powder (black arrow) is visible.

Figure 5.  CT appearance in the Pöschl plane of the superior semicircular canals (SSCs) of case 3: a and b: preoperative CT of the right (a) and left (b) SSCs. There is wide, bilateral CT dehiscence (black arrows). Both ears are clinically affected; c and d: postoperative CT of the right (c) and left (d) SSCs. The plugging wax, muscular fascia and covering cartilage are not visible (white arrows).
as was the case in our three observations. In the postoperative phase, CT is of use in looking for complications such as a fistula or pneumolabyrinth. In the first two of our observations, it allowed us to visualise the surgical reconstruction.

However, CT over-estimates the real prevalence of SSCDS due to its lack of sufficient spatial resolution [11]. In our study, we used thin 0.6 mm CT slices with steps of 0.2 mm in all the patients. Imaging assesses the prevalence as 4% [11,12], compared with 0.5% by optical microscopy in autopsies, because of the existence of bony walls thinner than 0.2 mm [12]. This high rate of false-positives is a problem, especially when faced with visualisation of bilateral CT dehiscence in a symptomatic patient: this was the situation in cases 2 and 3. In these two patients, it was important to determine which side or sides should be treated surgically, and it was not possible to establish this clearly based on the imaging data alone. Postoperatively, case 3 illustrates another limitation, arising from the radiotransparent character of certain materials used for surgical reconstructions, such as the plugging wax, covering muscular fascia or cartilage. From this patient’s CT scan, it was not possible to determine whether the third window had been correctly closed. These limitations of imaging mean that it is essential to undertake additional functional tests. The audiogram tests cochlear function, and, if there is SSCDS, shows conductive deafness, which predominates at frequencies below 1000 Hz, and, unlike in most cases of otospongiosis, that the stapedius reflexes are preserved, with an increase in bone conduction thresholds [1–5]. This was the case in our three observations on the side affected. Postoperatively, the air-bone gap was closed in all our patients. The VEMP tests assesses vestibular function. In the event of dysfunction, there is an increase in the amplitude of responses between the P13 and N23 waves, and a reduction in the stimulation thresholds below 80 dB [8,9]. In case 2, testing the VEMP allowed us to determine the side affected (unilateral right) whereas CT showed bilateral dehiscence. On the other hand, case 3 was an example of bilateral involvement both on the CT scan and in the functional tests. Postoperatively, normalisation of the VEMP confirmed closure of the third window in cases 2 and 3, which, for case 3, was not possible with imaging alone.

Combining CT, an audiogram and VEMP testing in our observations made it possible for us to define a population of patients with SSCDS who could benefit from SSC reconstruction surgery, by improving the overall specificity of the tests performed. This combination objectified the lesions, made it clear whether they were operable or not and the side or sides for surgery. In postoperative follow-up, this same combination confirmed the closure of the third window, even when the reconstruction materials were not visible by CT.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References