

Computed Tomography–Based Workup of Conductive Hearing Loss with Normal Middle Ear: Don't Forget Superior Semicircular Canal Dehiscence

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Abstract: Patients with superior semicircular canal dehiscence (SSCD) may have conductive hearing loss (CHL) with normal middle ear (ME). A patient with CHL and normal tympanic membrane at otoscopy underwent tympanotomy for presumed otosclerosis but had negative intraoperative findings. A high-resolution computed tomography (CT) scan of the temporal bone performed a few years later showed the absence of middle-ear disorder (especially the absence of imaging signs of otosclerosis) and revealed a bony dehiscence of the superior semicircular canal. High-resolution CT scan is the test of choice for diagnosing SSCD; it mimics otosclerosis by manifesting itself as CHL with a normal ME. The aim of this study is to remind radiologists and otolaryngologists that SSCD should be systematically considered among the etiologies of CHL with normal ME.

Key Words: computed tomography; conductive hearing loss; otosclerosis; superior semicircular canal dehiscence; temporal bone

Superior semicircular canal dehiscence (SSCD) was first recognized and described in 1998 [1]. Dehiscence of the bone overlying the superior semicircular canal can result in a syndrome of vertigo and oscillopsia induced by loud noises or by maneuvers that change middle-ear or intracranial pressure [1,2]. Patients with this disorder can also have conductive hearing loss (CHL) or hyperacusis to bone-conducted sounds in the presence of normal middle-ear function, with such symptoms as autophony, or hearing one's own eye movements or pulse. Thus, SSCD syndrome gives an explanation for both (1) a specific type of dizziness previously considered to be of unknown cause and (2) a particular type of CHL, previously either deemed idiopathic or wrongly attributed to otosclerosis [2–7].

However, the aforementioned auditory symptoms and signs can occur without vestibular manifestations [3–5,7,8]. This study aims to heighten awareness of the entity of SSCD in the imaging of patients investigated for CHL in the presence of normal middle-ear function, either with—or especially when without—associated vestibular manifestations.

PATIENT STUDY

A 54-year-old man was referred to our center at the Fondation Adolphe de Rothschild, Paris, France, owing to an ongoing complaint of hearing impairment and his rejection of hearing aids. He presented with a long history of left-sided CHL and negative exploratory tympanotomy results from examination in another institution 10 years earlier for presumed otosclerosis. He had no history of vertigo, disequilibrium, or otitis media. His records showed no subjective or objective postoperative changes in his hearing. Hearing remained the same

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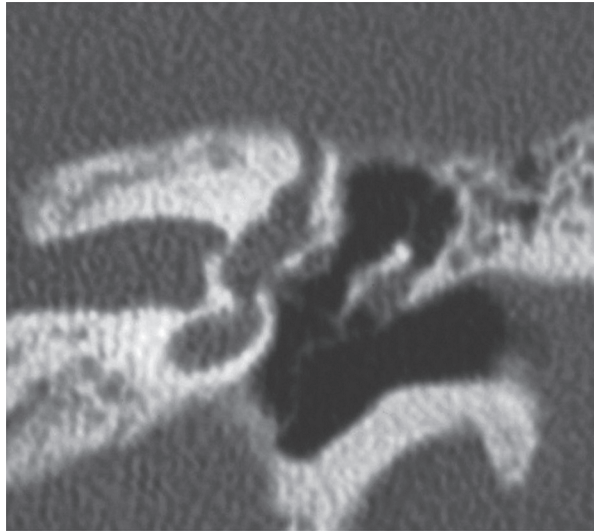


Figure 1. Standard coronal 0.6-mm-thick reformatted computed tomography image perpendicular to the plane of the lateral semicircular canal, showing a dehiscence of the bony roof of the superior semicircular canal.

throughout the years that followed and at the time of our workup. The old records and his post-referral examinations with tuning forks showed consistency with a left-sided CHL, whereas a series of pre- and postoperative audiograms and new audiograms in our center showed a stable 45-dB hearing loss (pure-tone average) and a 25-dB low-frequency air-bone gap in the left ear. Furthermore, we could not detect any eye movements or vestibular symptoms that were evoked by loud tones, Valsalva maneuvers, or pressure in the external auditory canal. Acoustic reflexes had been inconsistently recorded from the left ear (i.e., they were elicited at times and absent on other occasions).

We performed high-resolution computed tomography (CT) with a Siemens Sensation 16 CT scanner, with helical slices with 0.6-mm collimation of the x-ray beam and 0.3-mm incremental reconstructions in the axial and coronal plane (Fig. 1), yielding a pixel dimension of 0.137 mm^2 and a voxel dimension of 0.0822 mm^3 . The images were reformatted in a plane parallel to the superior semicircular canal (SSC; Pöschl plane) so that the entire dome of the canal could be visible on a single slice (Fig. 2). These images demonstrated a dehiscence 3 mm long in the bone overlying the left SSC.

According to clinical examination, hearing tests results, normal middle-ear surgical exploration, and evidence of a bony dehiscence of the roof of the SSC, the diagnosis of SSCD syndrome was given to the patient. We advised him that no surgical way to improve his hearing was available. We offered him a new trial with a hearing aid, but he declined.

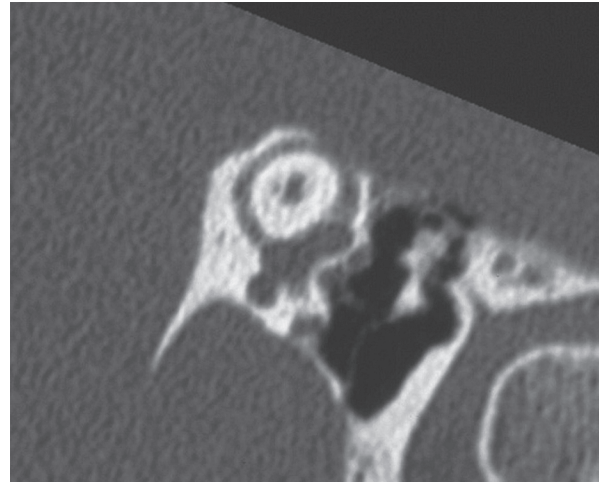


Figure 2. Oblique coronal 0.6-mm-thick reformatted computed tomography image in the plane of the superior semicircular canal, depicting more precisely the extent of the bony dehiscence located on the superomedial aspect of the canal.

DISCUSSION

Accurate CT-based diagnosis depends on the technological ability to detect the dehiscence of bone overlying the SSC. The presence of even a very thin sliver of bone is sufficient to close the canal and prevent the problem. Helical CT with 0.5-mm collimation of the x-ray beam allows visualization of layers of bone as thin as 0.1 mm [9].

The SSC lies in a plane approximately 45 degrees divergent from both the sagittal and coronal planes, being oblique to the routine transverse and coronal planes used in CT. Therefore, when current technology is pushed to the very limits of resolution in an effort to increase diagnostic accuracy, thin-section CT technique may be combined with off-axis oblique reformations [10]. The plane used in CT imaging, being 45 degrees from both the sagittal and coronal planes, is called the *Pöschl transverse pyramidal plane*. It sections the petrous bone in a plane perpendicular to its axis and along the axis of the cochlear modiolus, optimally showing the vestibular aqueduct in longitudinal section and displaying the SSC as a ring with the entire arc of its outer wall displayed on one image (see Fig. 1). The Stenvers plane is also 45 degrees oblique from the sagittal and coronal planes but rotated 90 degrees from the Pöschl plane; it shows the turn of the cochlea and displays the superior cortex of the SSC in a perfect cross-section. However, a more recent contention asserted that coronal reformations from multi-detector-row CT of the temporal bone are sufficient for the evaluation of SSCD, because the oblique reformations of Pöschl and Stenvers do not change the radiographic diagnosis and may be reserved for equivocal or confusing cases [11].

A further attempt to improve accuracy involves striving to overcome the inconveniences of the partial volume effect by examining the actual attenuation numbers within individual voxels, even though the image does not display the structure as the expected white line of bone. With such techniques, high-resolution temporal-bone CT scans may reach a sensitivity of 100%, specificity of 99%, positive predictive value of 93%, and negative predictive value of 100% in the identification of SSCD [12].

SSCD patients with symptomatology restricted to the auditory domain are a minority in comparison with those with vestibular manifestations and those with both types of symptoms. A meta-analytic count of the literature that includes patients of the former subgroup allows one to tally 27 patients who had CT-confirmed SSCD and only auditory and no vestibular symptoms [2–6,8,13]. The clinical presentation of SSCD in many of these patients so closely mimicked what is seen in ossicular abnormalities, such as otosclerosis, that surgical exploration and even stapedectomies and ossiculoplasties were performed without benefit to hearing [2,4–6]. Two similar cases have recently been recognized at our institution, one already published [7] and the other being presented here. Recognition and characterization of the CHL in SSCD combined with a heightened awareness of its diagnostic possibility should serve to reduce the number of patients who undergo unnecessary surgery.

SSCD syndrome is not the only entity that produces a CHL in the presence of a normal-appearing tympanic membrane. The differential diagnosis reveals two types of pathologies in this respect. On the one hand are abnormalities of the middle-ear conductive mechanism, such as ossicular discontinuity, tympanosclerosis, malleus head fixation, osteogenesis imperfecta, and Paget's disease. These can be detected radiographically but also present tympanometric and acoustic reflex abnormalities. On the other hand, a group of "inner-ear conductive hearing loss" pathologies present as an apparent CHL in the absence of any abnormality of the middle-ear conductive mechanism—with preserved acoustic reflexes and normal tympanometry. These include SSCD, the enlarged vestibular aqueduct syndrome, and other inner-ear malformations (of which the X-linked stapes gusher syndrome is most important). The role of CT in their detection is crucial, as only imaging can recognize them when investigating patients with air-bone gaps on audiometry and normal middle ears, so as to prevent unnecessary and deleterious ossicular replacement surgery. The existence of two simultaneous pathologies in the same patient is a rare instance, but it should also be kept in mind (i.e., concomitant otosclerosis and SSCD) [14].

Surgical treatment of SSCD (plugging or resurfacing SSC via a middle fossa approach) is indicated in the

case of disabling vertigo. In the case of CHL without vertigo, surgery is not recommended because of the risks of sensorineural complications [8]. In such cases, a hearing aid represents the safest method by which to improve hearing.

CONCLUSION

Otolaryngologists and neuroradiologists should be attentive to and systematically seek SSCD (among other causes of audiometric air-bone gap) on CT scans that are carried out in the workup of CHL in the absence of apparent tympanic abnormality, because not all such patients have otosclerosis.

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