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Clinical Record

Mondini-like malformation mimicking otosclerosis and superior semicircular canal dehiscence

M Karlberg, MD, PhD, M Annertz, MD, PhD*, M Magnusson, MD, PhD

Abstract
In 2003, it was reported that superior semicircular canal dehiscence can mimic otosclerosis because of low-frequency bone conduction hearing gain and dissipation of air-conducted acoustic energy through the dehiscence. We report the case of a 17-year-old girl with left-sided combined hearing loss thought to be due to otosclerosis. Bone conduction thresholds were −10 dB at 250 and 500 Hz and she had a 40 dB air–bone gap at 250 Hz. When a tuning fork was placed at her ankle she heard it in her left ear. Acoustic reflexes and vestibular evoked myogenic potentials could be elicited bilaterally. Imaging of the temporal bones showed no otosclerosis, superior semicircular canal dehiscence or large vestibular aqueduct, but a left-sided, Mondini-like dysplasia of the cochlea with a modiolar deficiency could be seen. Mondini-like cochlear dysplasia should be added to the causes of inner-ear conductive hearing loss.

Key words: Cochlear Diseases; Hearing Loss, Conductive; Vestibular Function Tests

Introduction

A hearing impairment with a consistent conductive element but without evidence of tympanic membrane or ossicular problems is termed inner-ear conductive hearing loss.[1,2] In 2003, two independent reports showed that dehiscence of bone overlying the superior semicircular canal (sSCC) can cause an inner-ear conductive hearing loss because of low-frequency bone conduction hearing gain and dissipation of air-conducted acoustic energy through the abnormal 'third window'.[3,4] Others have confirmed these findings,[5] and animal experiments support the third window hypothesis.[6] Disease entities other than sSCC dehiscence that open a third window into the inner ear might produce similar audio-vestibular effects, and this was recently shown in a patient with a venous malformation in the temporal bone[7] and also in a patient with dehiscence of bone between the cochlear basal turn and the carotid canal.[8]

Here, we report the case of a young woman with long-standing unilateral combined hearing loss thought to be due to otosclerosis. The audio-vestibular findings included low-frequency bone conduction hyperacusis and normal vestibular evoked myogenic potentials (VEMPs) and acoustic reflexes, despite an apparent low-frequency conductive hearing loss. Because of these findings, an sSCC dehiscence was suspected.[3,4] A computed tomography (CT) scan of the patient's temporal bones showed no otosclerosis or sSCC dehiscence, but a left-sided, Mondini-like dysplasia of the cochlea was observed.

Case report

A 17-year-old woman was referred to our balance clinic in March 2004 because of attacks of syncope and dizziness characterized as unsteadiness. In 1994, at seven years of age, she had been diagnosed with a left-sided combined hearing loss, with low-frequency conductive and high-frequency sensorineural loss. The hearing loss had been considered to be due to otosclerosis, and she had been followed with yearly audiometric tests, which showed unchanged hearing. In 1996, she had been referred for oto-neurological evaluation because of short attacks of vertigo during bouts of common cold. On that examination, she had shown no spontaneous, gaze-evoked, head-shaking or positional nystagmus and no deficiency of the vestibulo-ocular reflex on head impulse testing. Electro-oculography with tests of saccadic and smooth pursuit eye movements and optokinetic nystagmus and rotatory chair tests had been all normal. Her attacks of vertigo had been hypothesized to be caused by her 'otosclerosis'. In 2004, three weeks before she was seen at our balance clinic, she had had two attacks of syncope and had been observed at a paediatric clinic. She had also complained of headache and a continuous feeling of unsteadiness but no sensation of rotatory vertigo.

On examination, there was no spontaneous or gaze-evoked nystagmus and no deficiency of the vestibulo-ocular reflex on head impulse testing. Using infra-red videonystagmoscopy, there was no spontaneous, head-shaking, vibration-induced or positional nystagmus. The Hennebert test was negative for both ears. Nasal or glottal Valsalva manoeuvre and jugular compression produced no eye movements or vertigo.

Oto-microscopy showed normal auditory canals and tympanic membranes, with air-filled middle ears. The Weber test (using tuning forks of 128 and 512 Hz) lateralized to the left. In her left ear, the patient could also hear both the 128 Hz and the 512 Hz tuning forks when they were placed on her right ankle.

From the Departments of Otorhinolaryngology – Head & Neck Surgery, and *Diagnostic Radiology, Lund University Hospital, Lund, Sweden.
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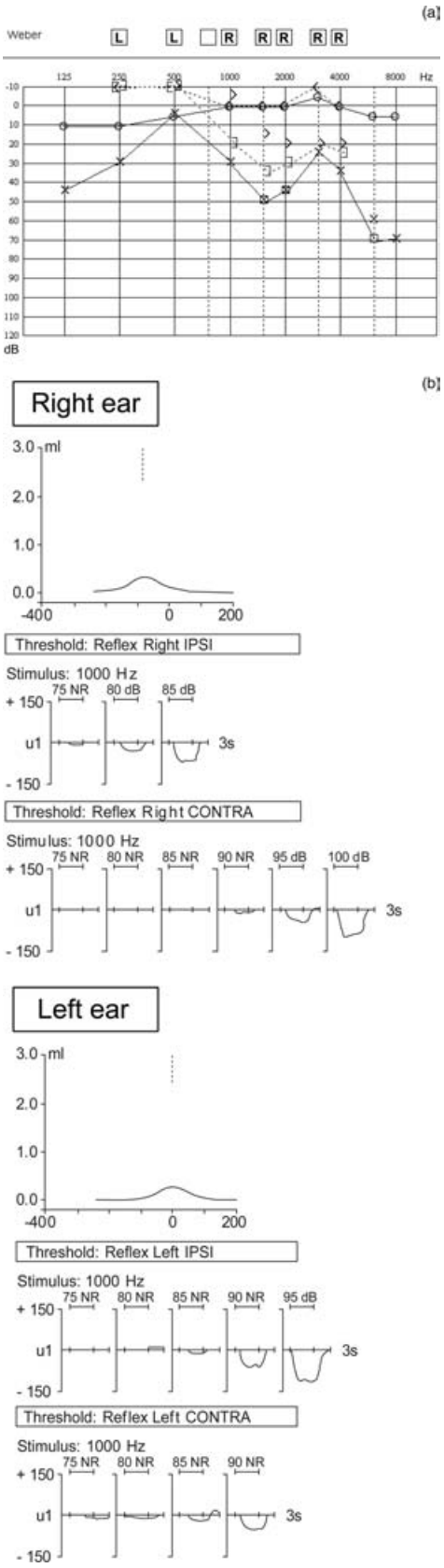

FIG. 1

(a) Pure tone audiometry shows a left-sided combined hearing loss with a low-frequency conductive component. The Weber test lateralized to the left ear at 250 and 500 Hz, and there was bone conduction hyperacusis (−10 dB) at 250 and 500 Hz. (b) Tympanometry and stapedial reflexes were elicited from both ears. IPSI = ipsilateral; CONTRA = contralateral

The audiogram showed a combined hearing loss in the patient's left ear, with a 40 dB air–bone gap at 250 Hz; hearing in her right ear was normal except for a 20 dB air–bone gap at 250 Hz (Figure 1a). The bone conduction

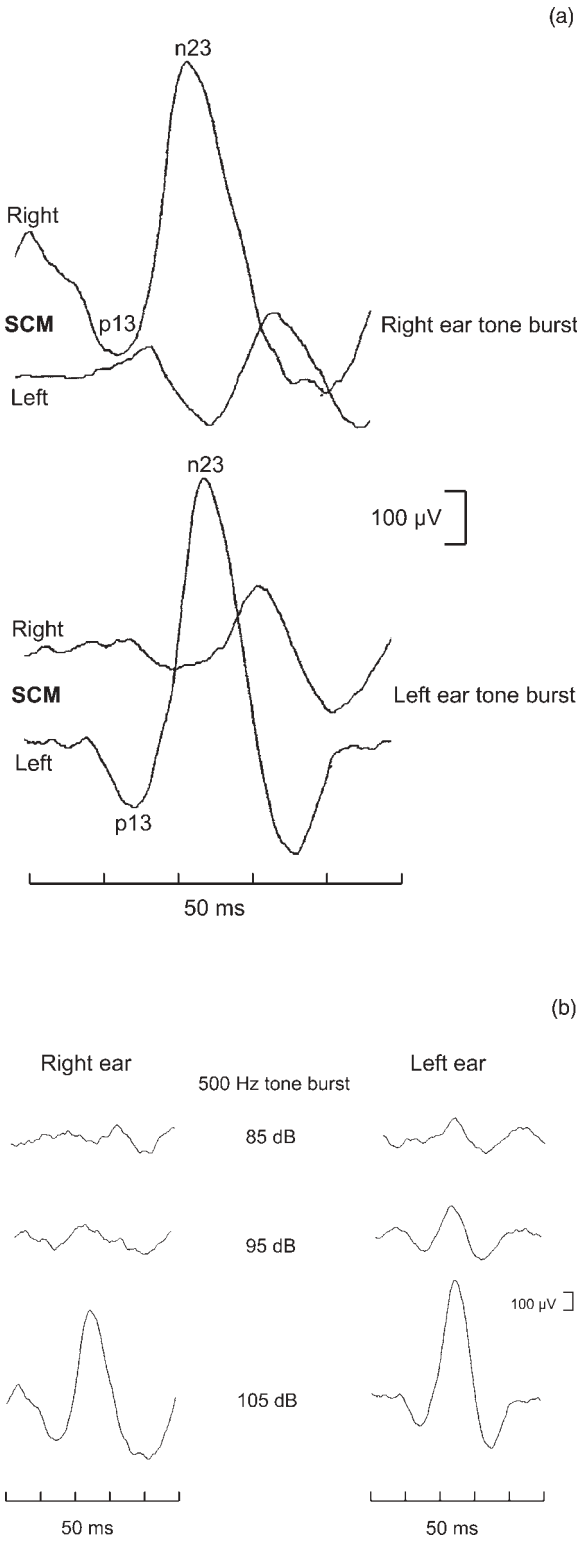

FIG. 2

(a) Large vestibular evoked myogenic potentials (VEMPs) could be recorded from both sternocleidomastoid (SCM) muscles in response to 105 dB tone burst stimulations of 500 Hz. (b) The VEMP threshold was 85 dB in the left ear and 95 dB in the right ear.

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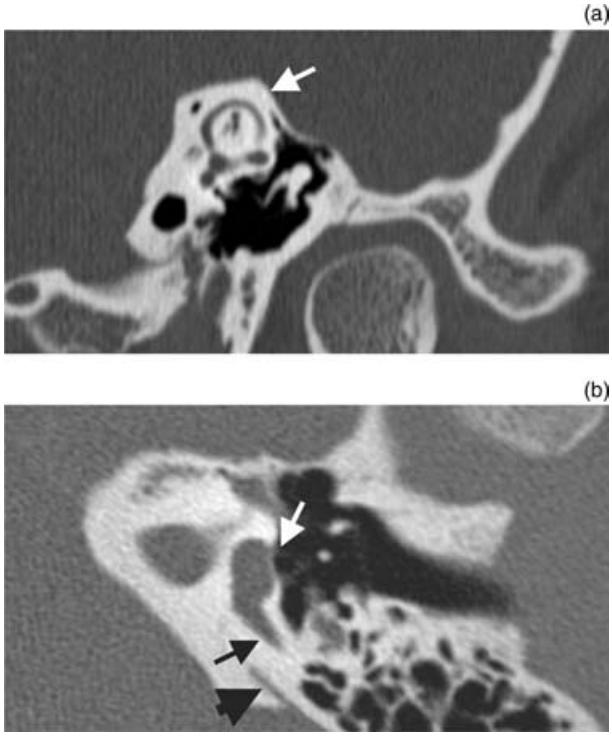

FIG. 3

(a) A computed tomography (CT) scan in the plane of the left superior semicircular canal shows normal thickness of bone overlying the canal (white arrow). (b) A CT scan in the plane of the left stapes (white arrow) shows no signs of otosclerotic foci. The vestibular aqueduct (large black arrowhead) is of a smaller diameter than the posterior semicircular canal (small black arrow).

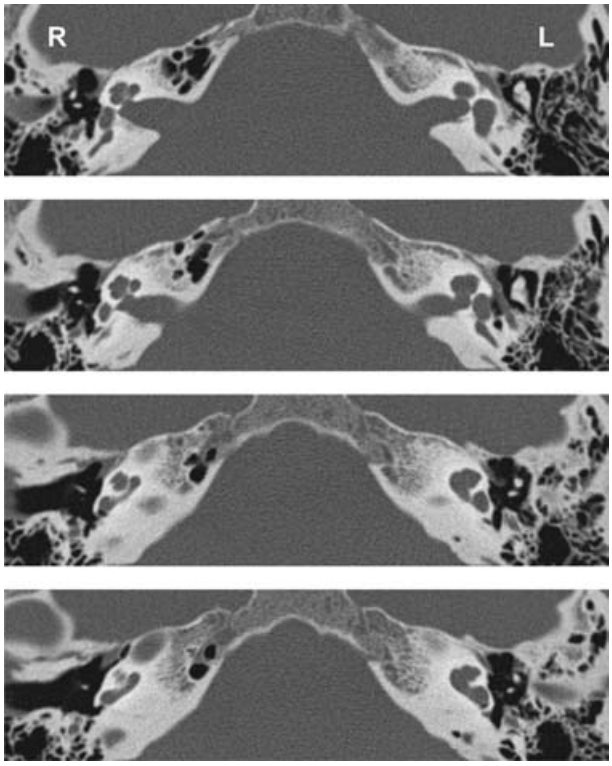

FIG. 4

Consecutive axial computed tomography scan slices show that the left cochlea is shorter and has a deficient modiolus as compared with the normal right cochlea.

thresholds at 250 and 500 Hz were −10 dB for both ears. The acoustic reflexes were present in both ears (Figure 1B).

Vestibular evoked myogenic potentials in response to 105 dB tone bursts at 500 Hz were present on both sides, despite the left conductive hearing loss (Figure 2a). The ipsilateral p13-n23 amplitude was 632 μV for the right ear and right sternocleidomastoid muscle and 710 μV for the left ear and left sternocleidomastoid muscle. The VEMP threshold was 85 dB in the left ear and 95 dB in the right ear (Figure 2b). The caloric test was normal.

The audiometric and vestibular findings suggested that the patient had an sSCC dehiscence in her left ear, and a high-resolution, multi-slice CT scan of the temporal bones was performed. The CT scan showed no signs of sSCC dehiscence, otosclerosis or large vestibular aqueduct (Figure 3). However, the scan showed dysplasia of the left cochlea, with a shortened cochlea and deficient modiolus (Figure 4). This might represent a Mondini-like dysplasia type B with communication between the basal turn of the cochlea and the internal auditory canal, as described by Zheng *et al.*[9]

Discussion

The original malformation described by Mondini consisted of a cochlea with 1.5 turns and a cystic apex, an enlarged vestibule with normal semicircular canals, and an enlarged vestibular aqueduct containing a dilated endolymphatic sac.[10] The Mondini malformation belongs to the group of congenital inner-ear malformations that result from arrested development during gestational weeks four to eight. Zheng *et al.*[9] have presented a classification based on the histopathologic cochlear findings: 1, common cavity cochlear dysplasia with severe dysplasia of the cochlea, without a complete basal turn; 2, Mondini dysplasia with 1.5 turns of the cochlea, a complete basal turn, complete bone at the base of the modiolus, and incomplete or absent interscalar septum; 3A, Mondini-like dysplasia type A, with two turns of the cochlea, normal interscalar septum and complete bone at the base of the modiolus; and 3B, Mondini-like dysplasia type B with 1.5 to two turns of the cochlea, a normal basal turn, and hypoplasia or absence of bone at the base of the modiolus, either with or without communication between the cochlea and the internal auditory canal. Based on the CT and audiovestibular findings in our patient, we suggest that she had a Mondini-like dysplasia type B, with communication between the cochlea and the internal auditory canal, as such a malformation would create a third inner-ear window.[9,11]

Any pathology that opens up a third window into the inner ear, be it a dehiscence of sSCC bone,[3–5] a large vestibular aqueduct,[12,13] a venous malformation,[7] a dehiscence of bone between the cochlear basal turn and the carotid canal,[8] or a communication between the basal turn of the cochlea and the internal auditory canal,[9,11] seems to be accompanied by the same, if somewhat variable, combination of findings: intact acoustic reflexes and VEMPs despite an apparent low-frequency conductive hearing loss, sometimes with low-frequency bone conduction hyperacusis. Nystagmus or ocular torsion in response to loud sounds (the Tullio phenomenon), pressure applied in the external auditory canal (the Hennebert sign), jugular compression or the Valsalva manoeuvre can be additional findings, but only when the size and location of the third window admit excitation or inhibition of an SCC cupula with intact function.[4,6]

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The message of this report is that any patient with an apparent conductive hearing loss but with preserved acoustic reflexes, preserved VEMPs and low-frequency bone conduction hyperacusis in the audiogram or during tuning fork tests to the ankle should, even in the absence of vestibular signs or symptoms, have a high resolution CT scan of the temporal bones. In this CT scan, it is important not only to look for sSCC dehiscence but also for any pathology that might act as a third inner-ear window, for example, cochlear dysplasia, large vestibular aqueduct or vascular malformations. Such findings, explaining the apparent conductive hearing loss, will prevent the patient from undergoing unnecessary middle-ear surgery. Furthermore, patients with a third window to the inner ear might also have a higher risk of excessive perilymphatic flow ('gusher') during stapedotomy or cochleotomy,[14] which makes the surgery not only unwarranted but also potentially harmful.

Acknowledgements

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- **In this case report, a 17-year-old woman with apparent conductive deafness was shown radiologically to have a left-sided, Mondini-like cochlear dysplasia**
- **Any patient with an apparent conductive hearing loss suggestive of otosclerosis but with preserved acoustic reflexes should have a high resolution CT scan of the temporal bones**
- **Mondini-like dysplasia is a possible cause of inner-ear conductive deafness, in addition to dehiscence of the superior semicircular canal**

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Address for correspondence:
Dr Mikael Karlberg,
Department of Otorhinolaryngology – Head & Neck Surgery,
Lund University Hospital,
SE-221 85 Lund, Sweden.

Fax: +46 46 2110968
E-mail: mikael.karlberg@skane.se

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