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Case Report

The Sensitivity of the cVEMP Test in Detecting A Superior Semicircular Canal Dehiscence and the Influence of a Coexisting Incudal Lysis: A Case Report

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In this case report, the air-conducted cervical vestibular evoked myogenic potentials (AC cVEMP) test was only sensitive for the left superior semicircular canal dehiscence (SCD), even though the contralateral SCD was of equal length (2.5 mm). Furthermore, a lysis of the processus lenticularis incudis caused a real conductive hearing loss in the left ear. A diminished left AC cVEMP was thus expected, but the opposite was shown (increased corrected amplitude, lowered detection threshold). The patient only experienced hearing loss, so middle ear surgery was performed to repair the lysis. The postoperative AC cVEMP showed a further "uncovering" of the SCD with increased corrected amplitude on the left but no vestibular symptoms. The significance of an SCD should be interpreted with caution, even when the AC cVEMP and the imaging are significant. Furthermore, AC cVEMPs should not be considered as evidence for the absence or presence of conductive hearing loss.

KEYWORDS: Dehiscence, semicircular canals, cervical vestibular evoked myogenic potential, lysis, conductive hearing loss

INTRODUCTION

A superior semicircular canal dehiscence (SCD) can induce vestibular sensations in response to loud sounds or pressure changes in the ear and/or intracranially [1]. A pseudo-conductive hearing loss can be induced as the inner ear biomechanics are changed with an apparent air–bone gap (ABG) as a result [1,2]. A test to identify SCDs is the (air-conducted) cervical vestibular evoked myogenic potential ((AC) cVEMP) test [3]. In case of a real ABG, the ABG degrades the AC cVEMP, as the sound energy reaching the inner ear is reduced [2]. In case of an SCD, however, the AC cVEMP parameters are increased [3]. Real conductive hearing loss can coexist with an SCD [4-6]. Moshtaghi et al. [5] reported a case in which the SCD was "masked" by otosclerosis. An ossicular chain reconstruction resulted in the "uncovering" of the SCD, which became symptomatic. Yong et al. found similar results [6]. In other cases, the SCD was more prominent and dominated the conductive hearing loss [4]. The use of imaging and thorough anamneses in such cases is of crucial importance to define the appropriate treatment.

In the current case report, a bilateral SCD and a real left-sided conductive hearing loss were coexisting. The sensitivity of the cVEMP test was only increased in the ear that also had an actual conductive component and this despite a radiologically significant length of the dehiscence bilaterally.

CASE PRESENTATION

A 59-year-old woman came to our ear-nose-and-throat department after being diagnosed elsewhere with otosclerosis. The patient's only complaint was hearing loss on the left side.

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The clinical examination showed a slight retraction of the posterior part of the left tympanic membrane, while the right tympanic membrane did not show any abnormalities. The audiological assessment provided normal tympanograms with present ipsilateral stapedial reflexes in only the right ear. Audiometric tests demonstrated a left conductive hearing loss (Figure 1). To confirm the suspected conductive hearing loss and to define the extent of the possible damage to the middle and inner ear, cone beam computed tomography (CBCT) was performed.

Cone Beam Computed Tomography showed a bilateral dehiscence of the superior semicircular canals (each of 2.5 mm), but no signs of otosclerosis (Figure 2a, 2b). The SCDs were found to be radiologically significant as the sizes exceeded 2.0 mm [7]. Additionally, a lysis of the lenticular process of the left incus was found (Figure 2c).

The cVEMP was bilaterally present (Figure 3a), but only the detection threshold of the left cVEMP (105 decibel Sound Pressure Level, dB SPL) was lowered when compared with the contralateral side (120 dB SPL) (left-right threshold difference=15 dB). A left-right threshold difference of 10 dB or more is abnormal according to our unpublished normative data. The corrected amplitudes of the cVEMP-responses, obtained at the highest stimulation level (i.e. 130 dB SPL) and at threshold level, were symmetric. The parameter "corrected amplitude" obtained at 130 dB SPL was within normal limits (normal values

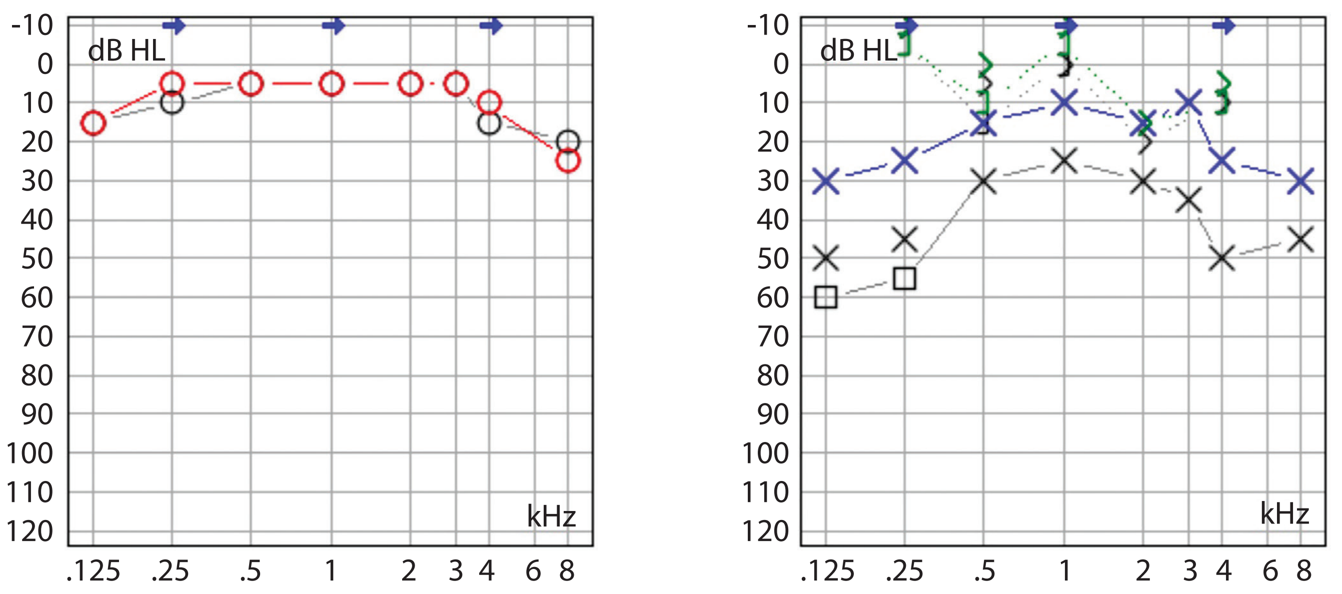

Figure 1. Pure tone audiometry measured pre- and postoperatively (black and colored lines, respectively). Preoperative pure tone averages (500, 1000, 2000 and 4000 Hz) were 8 decibel hearing level (dB HL) and 34 dB HL for the right and left ear, respectively. The left ear showed a conductive hearing loss with air–bone gaps (ABGs) of 60, 15, 25, 10, and 40 dB at 250, 500, 1000, 2000, and 4000 Hz, respectively. The postoperative measurement shows a partial closure of these ABGs, as the air-conduction threshold improved with an average of 17 dB. Arrows: Weber test indicating a conductive hearing loss in the left ear. x and o=left and right unmasked air-conducted hearing thresholds; ☐=masked left air-conduction hearing threshold; > and]=unmasked and masked left bone-conduction hearing thresholds.

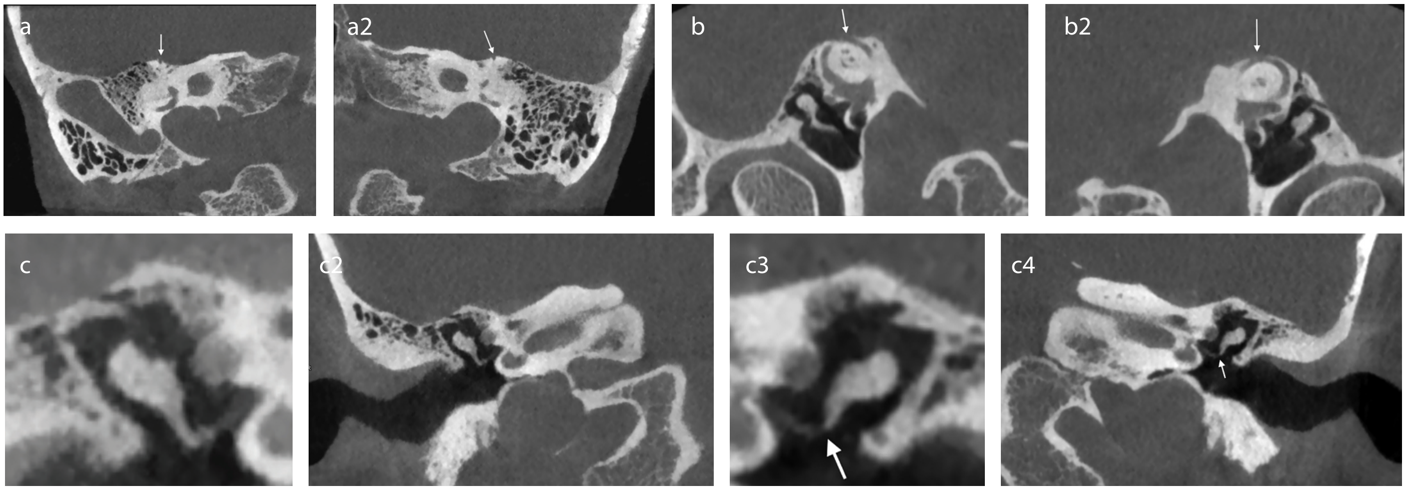

Figure 2 a-c. Cone beam computed tomography (CBCT) reformatted in the plane of Stenvers, perpendicular to the superior semicircular canal, showing the dehiscence of the bony roof of the right (left-hand panel) and left (right-hand panel) superior semicircular canal (arrows) (a). CBCT reformatted in the plane of Pöschl, perpendicular to the long axis of the temporal bone and in the plane of the superior semicircular canal, showing the entire extent of the defect (2.5 mm) (arrow) on both sides (b). Reformatted coronal CBCT and close-up of the incudostapedial joint showing a lysis of the lenticular process of the left incus (arrow). In the left-hand panel, the intact right-sided incudostapedial joint is presented with a close-up of the normal joint in the inset (c).

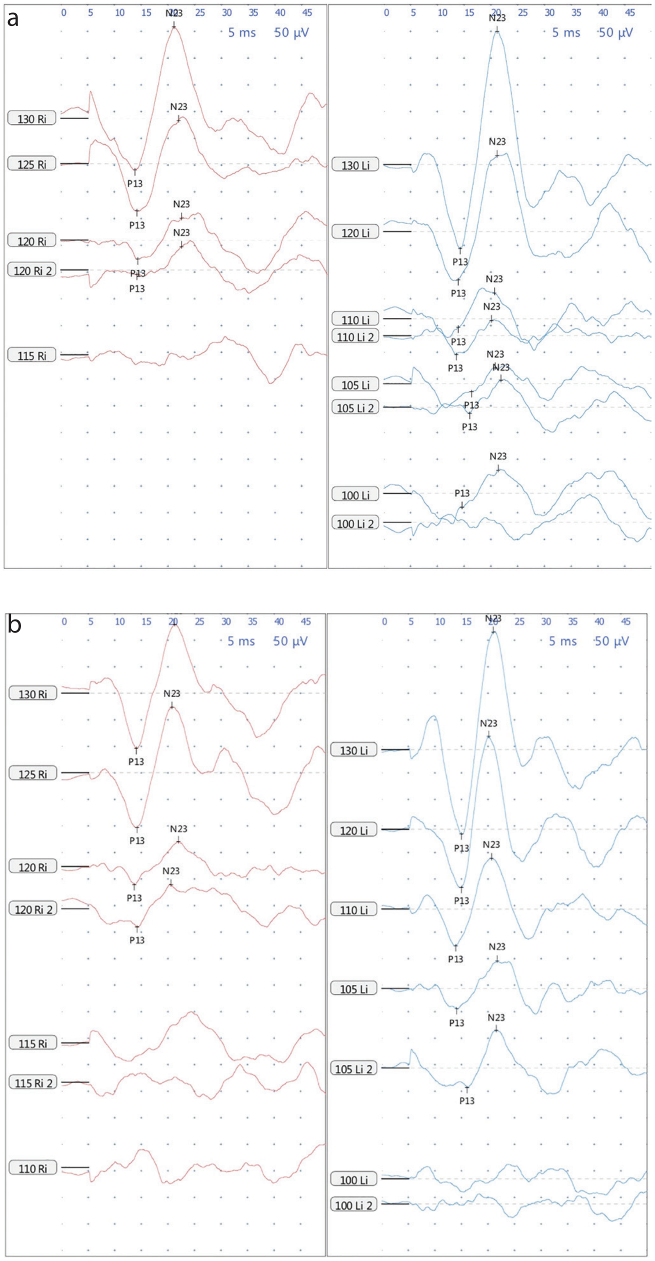

Figure 3. a, b. Air-conducted cervical vestibular evoked myogenic potentials (AC cVEMPs) obtained preoperatively (a). AC cVEMPs obtained postoperatively (b). Left-hand panels=right AC cVEMP, right-hand panels=left AC cVEMP. Li/Ri: left/right stimulation, ipsilateral recording (resp.). Values are expressed in decibel sound pressure level (dB SPL). X-axis: latency (milliseconds, ms).

ranging from 1.0 to 3.4; corrected amplitude left=2.5; corrected amplitude right=2.2) (Table 1). The latencies of the p13 and n23 peaks (Table 1) were also normal (normal p13 latency: 13.0-15.8 ms; normal n23 latency: 18.6-25.8 ms). No other vestibular tests were performed as there were no vestibular complaints.

In our department, an SCD is defined to be clinically significant when (1) the cVEMP test shows an increased p13-n23 amplitude and/or a lowered detection threshold, (2) the SCD-size is minimal 2 mm, and (3) the patient experiences SCD-mediated symptoms like autophony and pressure or sound-induced vertigo (Tullio phenomenon). For our patient, these criteria were only partially met as the patient did not report any vestibular symptoms and the cVEMP test only showed increased responsiveness on the left. Therefore, a transmeatal functional middle ear surgery was performed to minimize the hearing loss (i.e. the main complaint) by repairing the lysis with bone cement (OtoMimix®; Olympus Belgium, Berchem, Belgium).

Postoperatively, the left ABG closed partially, and the patient reported no complaints (Figure 1). The dehiscence might have caused the remaining small ABG. The cVEMP test was repeated and showed that the detection threshold remained stable for both sides, but for the left cVEMP, the corrected amplitude at threshold level increased from 0.4 to 0.8 (Figure 3b, Table 1). Furthermore, at the highest stimulation level of 130 dB SPL, the corrected amplitude was increased from 2.5 to 3.6 (Figure 3b, Table 1). The parameters "threshold" and "corrected amplitude" were now both indicative for the left SCD. The right AC cVEMP remained insensitive for the right SCD, despite its radiological significance.

DISCUSSION

In our patient, the clinical expression of two identical SCDs was different. The AC cVEMP was only capable of detecting the left SCD and this despite a coexisting incudal lysis. The AC cVEMP test did not detect the contralateral SCD despite the equal length. An additional saccular dysfunction may explain the right AC cVEMP, but this is not very likely due to the complete absence of vestibular symptoms. The counterintuitive AC cVEMP-results of our patient may be explained by contributing factors like ABG- and SCD-size [8, 9]. Pfamatter et al. [8] reported that SCDs of ≥2.5 mm predominantly result in combined auditory and vestibular symptoms. SCDs smaller than 2.5 mm result in isolated auditory or vestibular symptoms. Based on these findings, our patient should have experienced combined symptoms. This was not the case, but the coexisting incudal lysis may have influenced the overall experience of the symptoms. Another study [9] reported that the ABG-size at 250 Hz is significantly greater in patients with SCD who experience a greater subjective hearing loss. Although our patient had an ABG of only 15 dB at 500 Hz, the ABG at 250 Hz was 60 dB, which may have contributed to the patient's sole complaint of hearing loss. It is unclear how the incudal lysis influenced the possible vestibular sensations, especially when the postoperative absence of vestibular symptoms is considered. It is difficult to define strong and significant correlations between different contributing factors in isolated SCD cases [8,9], the interpretation of such interaction patterns in comorbid disorders is even more complex. Therefore, counseling is important, especially when surgical treatment (with a risk of uncovering the SCD) is considered [5].

In comorbid disorders, one disorder may dominate the other [4-6]. Consequently, there is a risk of underdiagnosing (and non-treatment) of secondary disorders. It is therefore important to combine anamnesis, clinical tests, and imaging.

CONCLUSION

Although the cVEMP test is considered the golden standard in identifying SCDs, it is not always capable of detecting them. In our patient, only the left sided SCD was detected, although the contralateral SCD was of equal length and radiologically significant. The left AC cVEMP did not show any signs of the incudal lysis but was highly suggestive

Table 1. Pre- and postoperative results of the air-conducted cervical vestibular evoked myogenic potential (AC cVEMP) test

	Stimulus intensity (130 dB SPL)	p13 latency (ms)	n23 latency (ms)	p13-n23 amplitude (raw, µV)	EMG-level (µV)	Corrected amplitude
Preoperative left cVEMP						
Highest stimulus intensity (dB SPL)	130	14.3	21.3	405.2	164.9	2.5
Threshold intensity (dB SPL)	105	16.0	22.0	60.6	164.3	0.4
Preoperative right cVEMP						
Highest stimulus intensity (dB SPL)	130	13.9	21.2	266.2	120.2	2.2
Threshold intensity (dB SPL)	120	14.3	22.5	51.4	100.4	0.5
Postoperative left cVEMP						
Highest stimulus intensity (dB SPL)	130	15.1	21.0	382.3	107.3	3.6
Threshold intensity (dB SPL)	105	16.1	21.6	107.1	126.9	0.8
Postoperative right cVEMP						
Highest stimulus intensity (dB SPL)	130	14.0	21.3	233.1	125.5	1.9
Threshold intensity (dB SPL)	120	14.2	20.6	79.5	151.4	0.5

dB SPL: decibel sound pressure level; ms: milliseconds; µV: microvolt; cVEMP; cervical vestibular evoked myogenic potentials

for the SCD. Therefore, AC cVEMPs cannot not be considered as evidence for the absence or presence of real conductive components. Furthermore, the AC cVEMP and imaging may indicate a significant SCD that cannot be clinically perceived.

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