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LETTER TO THE EDITORS

Idiopathic acute high-tone sensorineural hearing loss accompanied by vertigo: vestibulo-cochlear artery syndrome? Consideration based on VEMP and vHIT

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Dear Sirs,

A number of different pathological processes could result in acute sensorineural hearing loss (ASHL). It is widely believed that viral infection and vascular ischemia could be the most probable etiologies [1]. It is well-known that ASHL is frequently accompanied by vertigo [2].

In this case series, we studied patients with idiopathic acute high-tone sensorineural hearing loss (AHSHL) with vertigo using video head-impulse test (vHIT) and cervical vestibular evoked myogenic potential (cVEMP), discussing its etiology.

Among the patients in our database who underwent vHIT and cVEMP, we selected patients who fulfilled the following criteria as idiopathic AHSHL:

a. Unilateral sudden sensorineural hearing loss within 3 days at high frequencies (4000, and 8000 Hz) of $\geq$25 dB worse than the other side.
b. No elevation of hearing thresholds at low and middle frequencies (up to 1000 Hz).
c. Subjective feeling of vertigo
d. No other diagnosis than idiopathic sudden or acute sensorineural hearing loss

Eye-See-Cam system (Interacoustics, Denmark) or ICS Impulse system (GN Otometrics, Denmark) were used for vHIT. The methods were the same as in the previous study [3]. When a mean gain in vHIT of <0.7 for the vertical canals or <0.8 for the lateral canals with catch-up saccades were observed, the relevant canal was regarded as damaged.

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The Neuropack system (Nihon Kohden, Japan) was used to record cVEMP. Five hundred hertz air-conducted short-tone bursts (125 dB SPL) were used. The other recording methods were the same as in previous studies [3, 4]. Using the normalized amplitude of p13-n23, the asymmetry ratio (AR) was calculated [4]. The limit of the normal range of AR was set at 41.6 [4].

Consent concerning the use of clinical records was obtained from each patient in advance. This study was approved by the ethics committee of Teikyo University.

A total of three patients who fulfilled the criteria (a 40–44-year-old man, a 60–64-year-old man, and a 65–69-year-old woman) were enrolled. While patients #2 and #3 fulfilled diagnostic criteria of sudden sensorineural hearing loss [5], patient #1 had hearing loss only at 4000 and 8000 Hz. They did not have special risk factors for ischemia.

All the three patients showed decreased VOR gain and catch-up saccades on PSCC of the affected side (VOR gain = 0.30, 0.37, 0.33). However, they showed normal vHIT findings on LSCC and ASCC (Fig. 1). All the three patients showed normal cVEMP. ARs were 1.6, 2.6, and − 16.9.

In this case series, all the patients showed common features: (1) acute sensorineural hearing loss at high frequencies with normal hearing or mild hearing loss at low and middle frequencies; (2) severely damaged PSCC function with preserved function of LSCC and ASCC; and (3) normal saccule function [6].

Among the etiologies of ASHL, ischemia will explain these combinations of findings. According to Schuknecht [7], the inner ear has blood supply from the labyrinthine artery, which branches to the anterior vestibular artery and the common cochlear artery. The anterior vestibular artery provides blood supply to the LSCC, ASCC, utricle, and saccule. The common cochlear artery branches to the main cochlear artery and the vestibulo-cochlear artery. The vestibulo-cochlear artery branches to the posterior vestibular artery and the cochlear branch. The cochlear branch provides

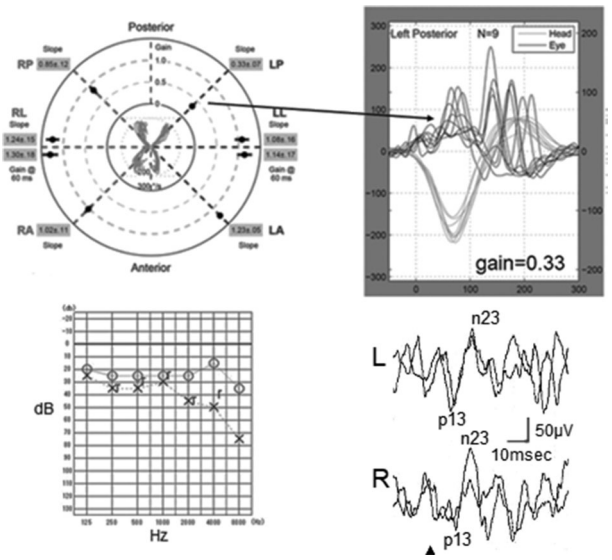

Fig. 1 Findings of patient #3 (a 65–69-year-old woman). Upper left: summary of vHIT results. Only left PSCC showed a decreased VOR gain (0.33). Upper right: vHIT results in left PSCC. Catch-up saccades were observed. Lower left: pure tone audiometry. O = right air-conducted hearing levels, X = left air-conducted hearing levels. Lower right: cVEMP findings. Both ears showed normal cVEMP responses

blood supply to the basal turn of the cochlea. The posterior vestibular artery provides blood supply to PSCC and saccule. Ischemia in the vestibulo-cochlear artery could result in dysfunction of the basal turn of the cochlea and PSCC. As the saccule also has blood supply from the anterior vestibular artery, the saccule function could be spared.

If Idiopathic AHSHL with vertigo was caused by viral labyrinthitis, the saccule must be damaged, because expansion of viral labyrinthitis to PSCC from the cochlea must involve the saccule. In the series of Pogson et al., four patients had AHSHL. One patient had only mild vestibular damages. As the same in our series, in other three patients, PSCC was most severely damaged in the semicircular canals. Although one patient had normal cVEMP, two patients had absent cVEMP. Therefore, they could not conclude they had infarction instead of labyrinthitis. Park et al. showed that cVEMP was affected more frequently than PSCC vHIT in vestibular neuritis. It might indicate another etiology [8].

Herein, we propose the hypothesis that idiopathic AHSHL with vertigo could be diagnosed as "vestibulo-cochlear artery syndrome" when patients show AHSHL, isolated PSCC dysfunction but preserved saccule function. At the current stage imaging cannot prove ischemia in these cases. It should be shown in the future study.

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Compliance with ethical standards

Conflicts of interest There is no conflict of interest in this study.

Ethical approval This study was performed in accordance with Declaration of Helsinki (1964) and its later amendments. This study was approved by the ethics committee of Teikyo University.

References

1. Pogson JM, Taylor RL, Young AS, McGarvie LA, Flanagan S, Halmagyi GM, Welgampola MS (2016) Vertigo with sudden hearing loss: audio-vestibular characteristics. J Neurol 263:2086–2096
2. Iwasaki S, Takai Y, Ozeki H, Ito K, Karino S, Murofushi T (2005) Extent of lesions in idiopathic sudden hearing loss with vertigo. Study using click and galvanic vestibular evoked myogenic potentials. Arch Otolaryngol Head Neck Surg 131:857–862
3. Murofushi T, Tsubota M, Kitao K, Yoshimura E (2018) Simultaneous presentation of definite vestibular migraine and definite Ménière's disease: overlapping syndrome of two diseases. Front Neurol 9:749. https://doi.org/10.3389/fneur.2018.00749
4. Murofushi T, Nakahara H, Yoshimura E, Tsuda Y (2011) Association of air-conducted sound oVEMP findings with cVEMP and caloric test findings in patients with unilateral peripheral vestibular disorders. Acta Otolaryngol 131:945–950
5. Stachler RJ, Chandrasekhar S, Archer SM, Rosenfeld RM, Schwartz SR, Barrs DM, Brown SR, Fife TD, Ford P, Ganiats TG, Hollingsworth DB, Lewandowski CA, Montano JJ, Saunders JE, Tucci DL, Valente M, Warren BE, Yaremchuk KL, Robertson PJ (2012) Clinical practice guideline: sudden hearing loss. Otolaryngol Head Neck Surg 146:S1–S35
6. Murofushi T (2016) Clinical application of vestibular evoked myogenic potential (VEMP). Auris Nasus Larynx 43:367–376
7. Schuknecht HF (1993) Pathology of the ear, 2nd edn. Lea & Febiger, Philadelphia
8. Park JS, Kim CH, Kim MB (2018) Comparison of video head impulse test in the posterior canal plane and cervical vestibular evoked myogenic potential in patients with vestibular neuritis. Otol Neurotol 39:e263–268