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Inferior Vestibular Neuritis: Absence of Vestibular Evoked Myogenic Potentials in the Presence of Normal Caloric Responses

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Background and Objectives: To report the clinical features of six patients diagnosed with cases of inferior vestibular neuritis based on abnormal vestibular evoked myogenic potential (VEMP) responses with normal caloric test results. **Materials and Methods:** We retrospectively reviewed 62 patients presenting with dizziness. All patients underwent a battery of audiovestibular testing, including hearing tests, caloric test and VEMP test. **Results:** Six patients were diagnosed as inferior vestibular neuritis. All patients presented with acute onset of prolonged vertigo. The pure tone audiograms and caloric test results were normal. VEMP response was absent unilaterally, and normal in the contralateral ear. **Conclusion:** Inferior vestibular neuritis should be considered in patients presenting with acute vertigo, but normal caloric responses. Comprehensive vestibular testing including VEMP is necessary.

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Key Words: Vertigo; Vestibular neuritis; Vestibular function test

Introduction

Vestibular neuritis is a common cause in acute vertiginous patients. Typical manifestation of sudden onset of vertigo and spontaneous nystagmus, gradually recovering over several days, and confirmation of unilateral vestibular hypofunction by decreased or absent caloric response usually suffice its diagnosis. Although an invaluable tool in vestibular laboratory, the caloric test reflects the function of the lateral semicircular canal, hence the superior division of the vestibular nerve. Vestibular evoked

myogenic potential (VEMP) testing evaluates the saccular function, innervated by the inferior vestibular nerve.¹ Vestibular neuritis is generally attributed to viral inflammation of the vestibular nerve, usually limited to the superior division of the nerve.² However, recently a few patients with clinical characteristics of vestibular neuritis, who show normal caloric responses but absent or abnormal VEMP test results, have been identified as inferior vestibular neuritis.³⁻⁵ In this study, we analyze the clinical presentation of six patients suspected of inferior vestibular neuritis, based on abnormal VEMP responses with normal caloric test results.

Materials and methods

1. Patients

We reviewed the clinical records of 62 consecutive patients (24 males and 38 females; age range 12-85 years; mean age 51.8 years) who presented with acute onset of dizziness and underwent audio-vestibular test, from January 2008 to December 2009 at the ENT department of Gangnam Severance Hospital. The inclusion criteria were as the following: 1) sudden onset of prolonged vertigo, 2) normal caloric responses bilaterally and 3) unilateral absence of VEMP response by click stimulation. The exclusion criteria were: 1) history of cerebrovascular disorder, 2) abnormality on neurologic examination or brain imaging study, 3) past history of excessive noise exposure, head injury or exposure to ototoxic drugs, 4) sensorineural hearing loss except for presbycusis, 5) the existence of air-bone gap (more than 10 dB) or abnormal tympanic membrane finding,⁶ 6) confirmed diagnosis of other audio-vestibular disorders, and 7) difficulty in maintaining muscle activity at a sufficient level (electromyographic [EMG] activity $>150 \mu\text{V}$).⁷

2. Methods

1) Caloric test

The bithermal caloric test was used, and eye movements were recorded by means of a video-based system (VisualEyes™ video nystagmography system, Micromedical Technologies, Inc., Chatham, IL, USA). Each ear was irrigated alternately with a constant flow of water at temperatures of 30°C and 44°C and for a constant period (30 sec). The maximum slow-phase eye velocity of nystagmus was calculated after each irrigation, and Jongkees' formula was used to determine canal paresis (CP). Canal paresis greater than 20% was defined as abnormal.

2) VEMP test

Vestibular evoked myogenic potentials were recorded using GSI Audera (Grason-Stadler, Eden Prairie, MN, USA). Briefly, surface electromyographic activity was recorded in a sitting position from symmetric sites over the upper half of each sternocleidomastoid muscle. Reference electrode was placed on the upper end of the sternum. During recording, the patients were instructed to turn

their heads and continuously contract the sternocleidomastoid muscle. The electromyographic signal from the stimulated side was amplified and bandpass-filtered (20-2000 Hz). Click stimuli (0.1 ms, 95 decibel normal hearing level [95 dB nHL]) were presented through a headphone and the initial positive-negative biphasic waveform comprised peaks P1 and N1 and the latencies were within 15-30 msec.

Results

The study included 62 patients who presented with dizziness and underwent both caloric test and VEMP test. Twelve patients were identified to fulfill the inclusion criteria, but 6 patients were excluded according to the exclusion criteria. One patient had a history of pontine hemorrhage, two patients presented with ipsilateral Bell's palsy, one patient was presented with concomitant benign paroxysmal positional vertigo, and two patients were diagnosed with possible Meniere's disease. The patients who had absent or abnormal VEMP results on both sides were not included. A diagnosis of inferior vestibular neuritis was established in the six patients who fulfilled both inclusion and exclusion criteria based on the analysis of history review and test results (Table 1). All patients had normal hearing and normal caloric responses. Spontaneous nystagmus was not present in any of the patients. The minimum follow-up period was 3 months. All patients reported recovery of the symptoms within 2 months since onset. Three patients underwent follow up VEMP test after 3 months since initial visit, and VEMP response remained absent on the lesion side in 2 patients and recovered partially in only one patient.

1. Typical case

A 38-year-old male (patient No. 1) presented with a history of sudden onset, whirling type vertigo with nausea which began 5 days ago. At the time of presentation, he reported slightly ameliorated symptoms, and complained of dizziness more similar floating sensation. Physical examinations did not reveal spontaneous or evoked nystagmus or any other neurologic findings. Caloric testing demonstrated no asymmetry. Pure tone audiometry was normal. Vestibular evoked myogenic potentials were normal on the right side, but absent on the left side (Figure 1). The symptoms dis-

Table 1. Clinical data for 6 patients with inferior vestibular neuritis

Patient no.	Sex/ Age (yr)	Presenting symptom	Delay of testing since onset of symptom	SN/HSN on initial visit	Resolution of symptoms after 2 months since onset/ recurrent vertigo episode	Initial VEMP (Right/Left)	Follow-up VEMP after 3 months (Right/Left)
1	M/38	Dysequilibrium, self-rotatory sensation	5 days	SN-/HSN-	Resolved/None	Absent/Normal	Decreased amplitude /Normal
2	M/62	Dysequilibrium,	1 month	SN-/HSN-	Resolved/None	Normal/Absent	-
3	F/47	Spinning sensation, floating sensation,	1 month	SN-/HSN, not recorded	Resolved/None	Normal/Absent	-
4	M/43	Dysequilibrium, aggravation on head turning	2 days	SN-/HSN, not recorded	Resolved/None	Normal/Absent	Normal/Absent
5	F/36	Dysequilibrium self-rotatory sensation	1 month	SN-/HSN-	Resolved/None	Absent/Normal	-
6	F/68	Dysequilibrium	1 month	SN-/HSN-	Resolved/None	Absent/Normal	Absent/Normal

SN, spontaneous nystagmus; HSN, head shaking nystagmus; VEMP, vestibular evoked myogenic potential.

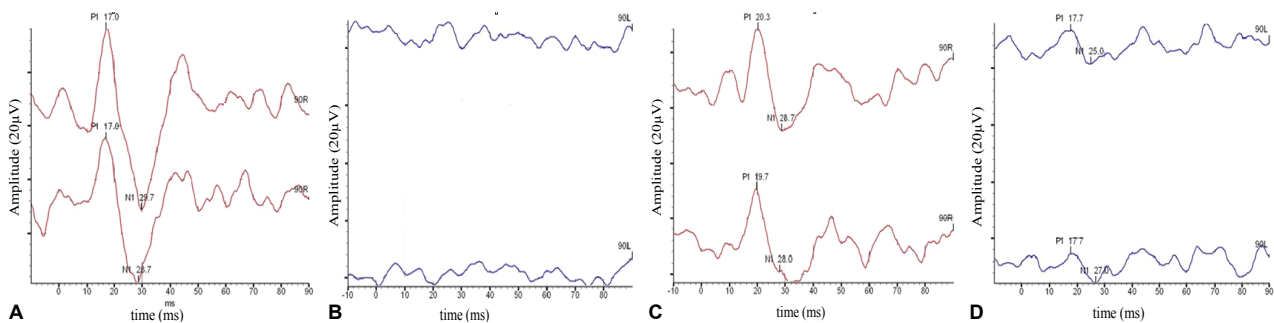


Figure 1. Vestibular evoked myogenic potential (VEMP) test results indicate normal response in the right ear (A) and absent response in the left ear (B) of the patient number 1 at initial presentation. After 3 months, VEMP response returned on the left ear after 3 months (D). VEMP was normal in the right ear (C).

appeared over the next two weeks. When the VEMP tests were repeated 3 months later, the patient was free of symptoms, and VEMP was recorded in the left ear, but the amplitude was smaller than in the left ear. Interaural amplitude difference (IAD) between P1 and N1 peaks was -70.7%.

Discussion

Inferior vestibular neuritis is a recently introduced concept as the superior vestibular nerve is generally involved in viral inflammation of the vestibular nerve.² Halmagyi et al.⁸ reported two possible cases presenting with acute onset vertigo and showing intact caloric responses and abnormal VEMP responses. In their re-

view of a large series of patients with balance problems, Iwasaki et al.³ identified eight patients with normal caloric responses and abnormal VEMP responses that were not attributed to other identified cause. Monstad et al.⁴ reported three cases with clinical picture of vestibular neuritis, loss of VEMP, and normal caloric responses. Recently, Zhang et al.⁵ described eight patients of inferior vestibular neuritis among 216 patients with vestibular neuritis. In the current study, vestibular function test results were reviewed in 62 patients who underwent both caloric and VEMP tests among 261 patients whose clinical diagnosis was acute vestibular neuritis based on clinical manifestation. Six patients were identified to have normal hearing and caloric responses but absent vestibular evoked myogenic potentials. Some suspected cases of inferior ves-

tibular neuritis were accompanied by ipsilateral hearing loss,⁸ but the patients in our study all had normal hearing. During the follow up, the patients recovered from severe dizziness, but mild transient dizziness upon rapid head movement lingered for several weeks. Clinical characteristics of inferior vestibular neuritis closely resemble classical neuritis of the superior vestibular nerve, but tend to be less severe in the acute phase. The VEMP tests were repeated at least 2 months from initial tests in 3 of our patients. The response returned to normal in only one (patient 1). Considering the recovery of normal caloric responses after classical vestibular neuritis is limited,⁹ the reappearance of normal VEMP response may signify that less severe damage to the inferior division of the vestibular nerve may be reversible as well.

Clinical manifestation of vestibular neuritis may be deceiving, as cerebrovascular lesions can present similar symptoms.^{10,11} Usually a diffusion weighted magnetic resonance imaging is recommended to rule out a possibility of central origin in patients with risk factors for vascular accidents. Also, loss of VEMP in the presence of normal caloric responses can occur in various conditions causing dizziness, both peripheral¹²⁻¹⁴ and central in origin (such as vestibular schwannoma, cerebellopontine angle tumors, and multiple sclerosis).³ Loss of saccular function with preserved lateral canal function can be expected in many peripheral vestibular lesions such as sudden hearing loss with vertigo, benign paroxysmal positional vertigo, and meniere's disease.^{3,12,14,15} The patients in our study were carefully reviewed to rule out other diseases where dissociation of VEMP and caloric test results has been reported. Two patients were excluded from our study because we suspected fluctuation of hearing loss around the onset of vertigo based on their reports although hearing test results were normal at the time of presentation. The possibility of an early stage Meniere's disease could not be ruled out. Also, one patient presenting with clinical features of vestibular neuritis was excluded because he was also diagnosed with posterior semicircular canal canalolithiasis ipsilateral to the side with abnormal VEMP response. Interestingly, few patients presenting with acute onset vertigo to our clinic did show bilaterally absent or abnormal VEMP responses and yet normal caloric responses. Fujimoto et al.⁷ have introduced cases of idiopathic bilateral vestibulopathy limited to the inferior vestibular nerve system. However, the symptoms of our patients were not typical of bilateral vestibulopathy and other causes of abnormal VEMP re-

sults could not be excluded.

Limitation of our study includes the observation that there lies a possibility that initially present caloric loss has recovered over time, along with amelioration of the symptoms. Previous studies reported slower recovery of caloric responses compared to otolith-related test results.^{5,13} However, about 25% of vestibular neuritis patients confirmed of pathologic canal paresis recovered to show normal caloric responses during follow up of two months.⁹ Since the vestibular tests were performed about a month after the onset of vertigo in our patients, recovery of caloric response may be possible, but unlikely, to have preceded the recovery of VEMP responses. The diagnosis of selective involvement of inferior vestibular nerve yet remains elusive: exclusion of other possible causes of vestibulopathy is mandatory. Meticulous review of patient reported symptoms and timely evaluation using comprehensive vestibular function tests would yield more accurate diagnosis in vertiginous patients.

Conclusion

The patients in our study presented with acute onset vertigo and laboratory findings support the diagnosis of inferior vestibular neuritis. Clinical suspicion and comprehensive vestibular function test battery including vestibular evoked potential tests are indispensable in the effort to localize the lesion site in the peripheral vestibular system.

중심 단어: 어지럼, 전정신경염, 전정기능검사

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