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Periodic Tinnitus and Direction-Changing Nystagmus in Vestibular Paroxysmia

Chang-Hee Kim[a]
Kwang-Dong Choi[b]

[a]Department of Otorhinolaryngology-Head and Neck Surgery, Konkuk University Medical Center, Research Institute of Medical Science, Konkuk University School of Medicine, Seoul, Korea
[b]Department of Neurology, Pusan National University Hospital, Pusan National University School of Medicine and Biomedical Research, Busan, Korea

Dear Editor,

Vestibular paroxysmia (VP) is characterized by short vertiginous spells with or without hearing symptoms such as tinnitus.[1,2] Neurovascular cross-compression of the eighth cranial nerve has been assumed to be the underlying pathophysiology of the VP, and VP is diagnosed mainly based on clinical manifestations and treatment responses.[1,2] The characteristics of paroxysmal nystagmus during the spells of VP have rarely been described.[3-5] We report a patient with VP who presented with periodic tinnitus and direction-changing nystagmus during the attacks.

A 78-year-old female developed recurrent left-sided "clicking tinnitus" and experienced a brief attack of vertigo 1 month prior to presentation, which occurred two or three times every day. A video-Frenzel examination revealed weak right-beating horizontal nystagmus. Bithermal caloric tests showed 39% canal paresis on the left side (Fig. 1A). Video head impulse tests revealed decreased vestibulo-ocular reflex gain with covert saccades in the left horizontal semicircular canal (Fig. 1B). Pure-tone audiometry showed symmetric slight high-frequency hearing loss on both sides (Fig. 1C), and auditory brainstem response testing revealed no interpeak delay on the left side (Fig. 1D). Fast imaging employing steady-state acquisition (FIESTA) MRI demonstrated neurovascular cross-compression between the anterior inferior cerebellar artery and the cisternal segment of the left vestibular nerve (Fig. 1E and F).

Treating the patient with 300 mg oxcarbazepine daily for 2 years resulted in considerable improvement of the symptoms. However, about 2 years after treatment initiation the patient reported experiencing 30-second-long vertigo attacks every 2 minutes throughout the day, which were always preceded by left-sided "clicking tinnitus." There was no trigger factor that provoked these symptoms. Pure-tone audiometry revealed symmetric slight age-related deterioration on both sides since the last examination 2 years previously (Supplementary Fig. 1 in the online-only Data Supplement). A video-Frenzel examination showed persistent right-beating nystagmus that was interposed by left horizontal and torsional nystagmus with downbeating components every 90−105 seconds lasting for 30−35 seconds (Supplementary Video 1 in the online-only Data Supplement). The paroxysmal vertigo was relieved by increasing the dosage of carbamazepine to 400 mg daily, which had no side effects.

A review of the literature revealed three additional cases presenting with paroxysmal nystagmus during VP attacks: this occurred spontaneously in two of the cases and was triggered by hyperventilation in the third (Supplementary Table 1 in the online-only Data Supplement).[3-5] Paroxysmal nystagmus lasted for 5−30 seconds, and was accompanied by tinnitus in one patient. The paroxysmal nystagmus was pure ipsilesional horizontal (n=1), ipsilesional horizontal and torsional (n=1), or horizontal and torsional with a downbeating component (n=1). The paroxysmal nystagmus was periodic, with a frequency ranging from 47 seconds to 10 minutes in two patients, and one of them showed periodic direction-changing

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Correspondence
Kwang-Dong Choi, MD, PhD
Department of Neurology, Pusan National University Hospital, Pusan National University, School of Medicine, 179 Gudeok-ro, Seo-gu, Busan 49241, Korea
Tel +82-51-240-7317
Fax +82-51-245-2783
E-mail kdchoi@pusan.ac.kr

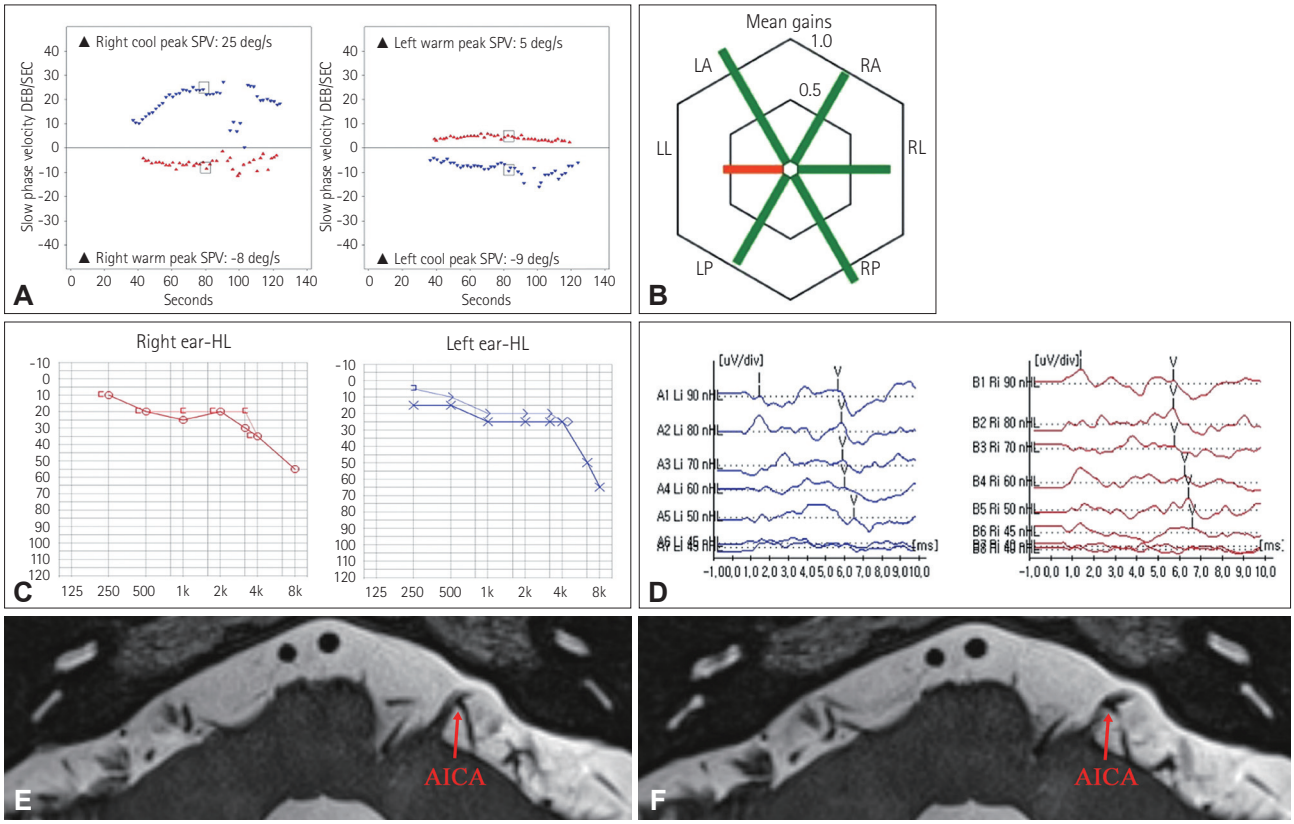

Fig. 1. Results of laboratory and radiological evaluations of the patient. A: Bithermal caloric tests reveal left unilateral canal paresis. B: Video head impulse tests reveal decreased vestibulo-ocular reflex gain in the left horizontal semicircular canal. C: Pure-tone audiometry shows slight high-frequency loss on both sides. D: Auditory brainstem response testing reveals an interaural latency difference of 0.09 milliseconds, which is within the normal range. E, F: 3D balanced FIESTA (fast imaging employing steady-state acquisition) MRI demonstrates neurovascular compression between the anterior inferior cerebellar artery and the cisternal segment of the left vestibular nerve (red arrows). FIESTA: fast imaging employing steady-state acquisition.

nystagmus at 5 years after vertigo with an irregular frequency had first appeared. The present case also developed periodic direction-changing nystagmus 2 years after irregular-frequency vertigo had first appeared. Persistent right horizontal nystagmus was periodically interposed by 30- to 35-second-long paroxysms of left horizontal torsional nystagmus every 90–105 seconds. Chronic left peripheral vestibulopathy was evident from background right-beating nystagmus and the results of a caloric test and video head impulse test. Remarkably, periodic direction-changing nystagmus in the present case was always preceded by left-sided "clicking tinnitus," which is a distinguishing finding of the present study.

Patients with VP can develop persistent contralesional nystagmus resulting from chronic peripheral vestibulopathy, and variable patterns of paroxysmal ipsilesional nystagmus with or without tinnitus. Paroxysmal nystagmus and tinnitus would be periodic in some patients with long-standing VP, which can be explained by secondary central hyperactivity in the vestibular and cochlear nuclei caused by long-standing compression or direct pulsatile compression with ephaptic discharges in the peripheral vestibular nerve.[2,3] This mechanism is supported by the transition from irregular-frequency vertigo for several years into periodic attacks and a case with unilateral vestibular nuclear hemorrhage developing similar periodic direction-changing nystagmus.[6] Central sensitization and decreased central inhibitory activity have also been reported when symptoms developed in the fifth cranial nerve.[7]

Supplementary Video Legend

Video 1. Video-Frenzel examination shows a persistent right-beating nystagmus which is interposed by a left torsional and horizontal nystagmus every 90–105 s. The left torsional and horizontal nystagmus lasts for 30–35 seconds and is accompanied by vertigo.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.3988/jcn.2021.17.3.493.

Author Contributions

Conceptualization: Chang-Hee Kim, Kwang-Dong Choi. Data curation: Chang-Hee Kim, Kwang-Dong Choi. Formal analysis: Chang-Hee Kim. Funding acquisition: Kwang-Dong Choi. Investigation: Chang-Hee Kim.

Methodology: Chang-Hee Kim. Project administration: Chang-Hee Kim. Resources: Chang-Hee Kim. Supervision: Kwang-Dong Choi. Validation: Kwang-Dong Choi. Visualization: Chang-Hee Kim. Writing—original draft: Chang-Hee Kim. Writing—review & editing: Kwang-Dong Choi.

ORCID iDs

Chang-Hee Kim https://orcid.org/0000-0001-5667-861X
Kwang-Dong Choi https://orcid.org/0000-0002-9373-4710

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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