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LETTER TO THE EDITORS

The nystagmus of vestibular paroxysmia

Seo-Young Choi[1] · Jae-Hwan Choi[2] · Kwang-Dong Choi[1]

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Dear Sirs,

Vestibular paroxysmia is a rare vestibular disorder characterized by brief attacks of spinning or non-spinning vertigo which lasts from a second up to a few minutes, and occurs with or without ear symptoms [1, 4, 6]. A neurovascular cross-compression of the eighth cranial nerve is assumed to be the cause of short episodes of vertigo in vestibular paroxysmia that was recently documented in a patient who underwent a successful operation [1, 2, 4, 6, 7]. The frequency of attacks of vertigo in vestibular paroxysmia greatly varies among patients from 30 attacks per day to a few attacks per year. We report a patient with vestibular paroxysmia which presented periodic vertigo with paroxysmal nystagmus.

A 59-year-old man, a professional conductor of classical music first developed vertigo attacks about 5 years ago. Each attack would last only about 10 s but he would have 2–3 attacks every day. He sought our medical attention but was too busy to undergo investigation. He now presented because the attacks had increased in frequency over the last 3 months. He was now having the same 10 s attacks every minute. Video-oculography disclosed a persistent left-beating nystagmus which would reverse every 47 s, to right-beating nystagmus for 10 s and would be accompanied by vertigo (figure A and B, video). The mean and maximum slow phase velocities of resting spontaneous left-beating nystagmus were 2 and 4°/s, respectively, while those of the paroxysmal right-beating nystagmus were 3 and 7°/s (Fig. 1b). Hyperventilation did not affect the pattern of spontaneous and paroxysmal nystagmus. Video head impulse tests (HITs) showed decreased vestibulo-ocular reflex gain with covert saccades in right horizontal semicircular canal (Fig. 1c), and bithermal caloric tests revealed a right canal paresis of 59% (Jongkees equation) (Fig. 1d). Cervical and ocular vestibular-evoked myogenic potentials (VEMPs) were decreased during right ear stimulation with an interaural difference at 29% (normal range 22%) and 49% (normal range < 18%), respectively (Fig. 1e, f). Pure tone audiometry and brainstem auditory evoked potentials were normal. Constructive interference in steady state (CISS) MRIs demonstrated a neurovascular cross-compression between the anterior inferior cerebellar artery and the cisternal segment of the right vestibular nerve (Fig. 1g). The periodic vertigo with nystagmus resolved by administration of 300 mg of oxcarbazepine daily.

In our patient, recurrent stereotyped vertigo lasting for seconds, resolution of vertigo by a treatment with oxcarbazepine, and neurovascular cross-compression on brain MRI meet the diagnostic criteria for vestibular paroxysmia [6]. Our patient with vascular compression of the right vestibular nerve developed a chronic right peripheral vestibulopathy (evident from background left-beating nystagmus, and vestibular impairment on caloric, head impulse and VEMP testing), and also developed vestibular paroxysmia during which he would have 10 s paroxysms of right-beating nystagmus every 47 s. Previously, a patient showed spontaneous upward nystagmus during the attack of transient vertigo in vestibular paroxysmia [1].

The periodicity of vertigo with paroxysmal nystagmus in our patient can be explained by direct pulsatile compression with ephaptic discharges in the peripheral vestibular nerve or secondary central hyperactivity in the vestibular nuclei which is induced and maintained by long-standing compression. The transition from irregular frequency of vertigo for several years into periodic attacks may support the latter mechanism. Central sensitisation and decreased central

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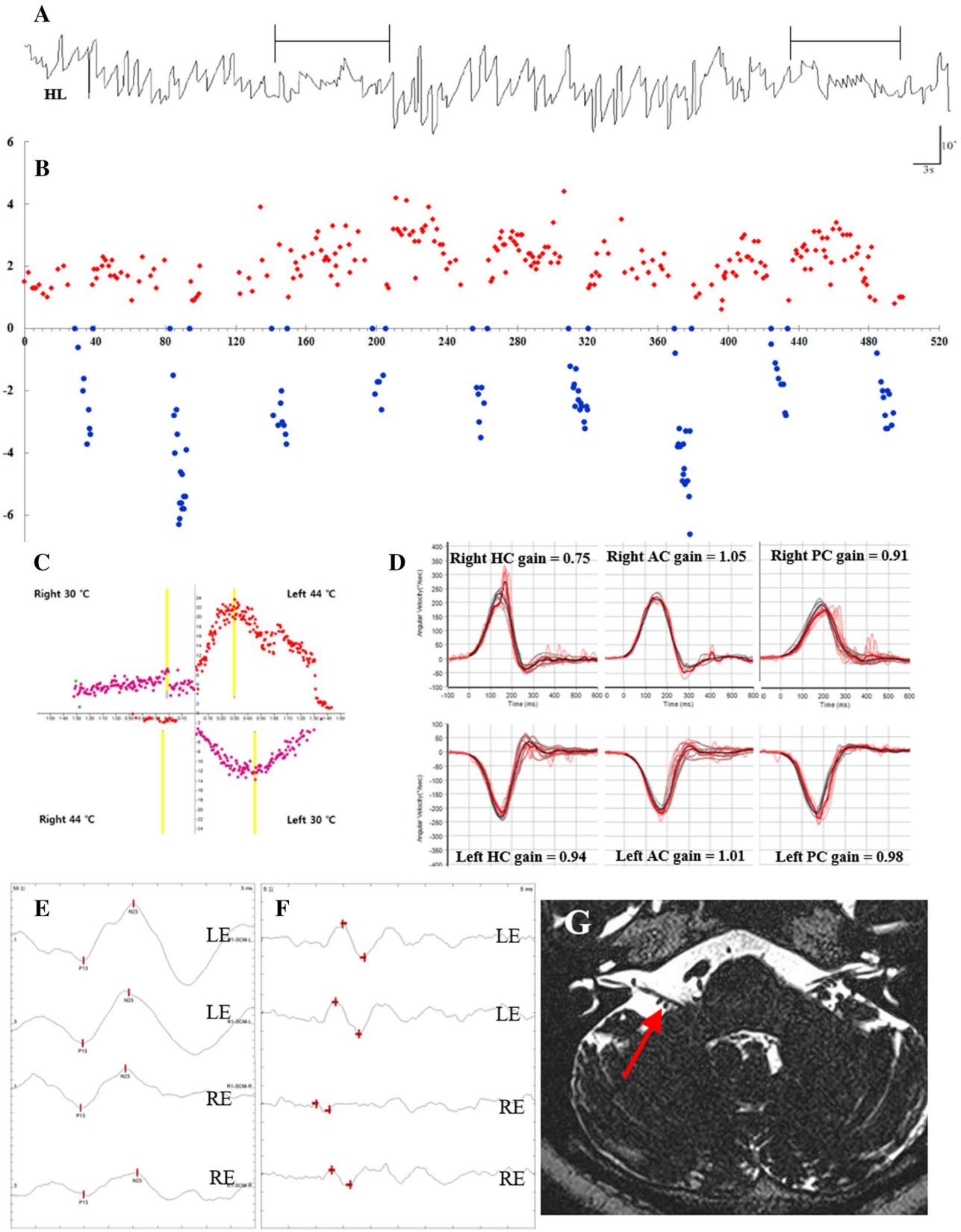
✉ Kwang-Dong Choi
kdchoi@pusan.ac.kr

[1] Department of Neurology, College of Medicine, Pusan National University Hospital, Pusan National University School of Medicine and Biomedical Research Institute, 179, Gudeok-ro, Seo-gu, Busan 602-739, South Korea

[2] Department of Neurology, Research Institute for Convergence of Biomedical Science and Technology, Pusan National University Yangsan Hospital, Pusan National University School of Medicine, Busan, South Korea


A
HL
10°
3s
B
C
Right 30 °C
Left 44 °C
Right 44 °C
Left 30 °C
D
Right HC gain = 0.75
Right AC gain = 1.05
Right PC gain = 0.91
Left HC gain = 0.94
Left AC gain = 1.01
Left PC gain = 0.98
E
LE
LE
RE
RE
F
LE
LE
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G

◂**Fig. 1** **a** Three-dimensional video-oculography shows spontaneous left-beating nystagmus without fixation which is periodically intervened by paroxysmal right-beating nystagmus. **b** The plotting of the nystagmus reveals a persistent left-beating nystagmus (red dots) which reverses every 47 s to right-beating nystagmus (blue dots) for 10 s. Negative value means rightward direction of the slow components of nystagmus. **c** Bithermal caloric tests show a right canal paresis of 59%. **d** Video head impulse tests show decreased vestibulo-ocular reflex gain with covert saccades in right horizontal semicircular canal (normal gain = 0.88–1.03), but normal gains for the left horizontal and both anterior (normal gain = 0.91–1.07) and posterior semicircular canals (normal gain = 0.88–1.06). Cervical (**e**) and ocular (**f**) vestibular-evoked myogenic potentials are decreased during right ear stimulation with an interaural difference at 29% (normal range < 22%) and 49% (normal range < 18%), respectively. **g** Brain constructive interference in steady state magnetic resonance images demonstrates a neurovascular compression between the anterior inferior cerebellar artery and the cisternal segment of the right vestibular nerve (red arrow). *HL* horizontal recording of the left eye movement, *HC* horizontal semicircular canal, *AC* anterior semicircular canal, *PC* posterior semicircular canal, *LE* left ear stimulation, *RE* right ear stimulation. Upward deflection indicates rightward eye motion

inhibitory activity both resulting in hyperexcitability were described in the context of symptom development for the fifth cranial nerve [3]. Furthermore, similar periodic patterns of nystagmus were reported in a patient with unilateral vestibular nuclear hemorrhage [5]. At 2-min intervals, a paroxysmal burst of nystagmus reversed an asymptomatic resting nystagmus during the inter-paroxysmal period. Intermittent hyperactivity in the subpopulation of vestibular nucleus can induce regularly pathologic brief bursts, and cause the episodic reversal of resting nystagmus.

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Compliance with ethical standards

Conflicts of interest We have no disclosure of any competing interest.

Ethical approval for research involving human participants and/or animals All experiments followed the tenets of the Declaration of Helsinki, and this study was approved by the Institutional Review Board.

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