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Acute Vertical Pendular Nystagmus and Delayed Oculopalatal Tremor in a Patient with Bilateral Horizontal Gaze Palsy

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Dear Editor,

Oculopalatal tremor (OPT) is a delayed complication of damage to the dentatorubro-olivary pathway and subsequent hypertrophic olivary degeneration.[1] Various patterns of eye movement abnormalities due to the acute damage precede vertical pendular nystagmus (PN) and palatal tremor.[2] We observed acute vertical PN and delayed OPT in a patient with bilateral horizontal gaze palsy.

A 46-year-old man experienced the sudden onset of vertigo and diplopia. The patient exhibited a complete limitation of horizontal conjugate eye movements including saccades, pursuit, and the vestibulo-ocular reflex. Vertical eye movements were intact but the patient had difficulty holding a vertical gaze. Symmetric vertical PN was observed in both eyes during visual fixation, was augmented during vertical gaze (Supplementary Video 1 in the online-only Data Supplement), and was accompanied by intermittent clockwise torsional beating during downward gaze. Three-dimensional video-oculography (VOG) performed at 10 days after the onset of acute symptoms showed pure vertical, binocular, conjugate PN at a frequency of 3–4 Hz in the primary position during visual fixation and vertical gaze. The amplitude was increased during vertical gaze [i.e., gaze-evoked nystagmus (GEN)]. No nystagmus occurred in the primary position without fixation. Slow upward deviation of both eyes with corrected downward eye movement was observed (Fig. 1A). Axial brain diffusion-weighted MRI performed 1 hour after the onset of his symptoms revealed an acute infarction involving the medial medullae bilaterally and extending to the tegmentum and both paramedian caudal pontine tegmenta (Fig. 1B). Acute PN was not observed at the 3-month follow-up (Supplementary Video 2 in the online-only Data Supplement). Eighteen months later the patient developed oscillopsia and a change in voice color. He had partial limitation of bilateral horizontal eye movements but his horizontal vestibulo-ocular reflex was normal. The torsional component with small-amplitude PN and asymmetric palatal tremor were also observed (Supplementary Video 3 in the online-only Data Supplement). Follow-up VOG showed asymmetric vertical PN at a frequency of 3–4 Hz with a higher amplitude in the right eye (2.5–3°) that was attenuated during visual fixation. Slow upward deviation was observed in both eyes in the absence of fixation (Fig. 1C). Brain MRI showed a subtle high-intensity signal in the inferior olivary nucleus (ION) on the left side (Fig. 1B).

We observed different patterns of vertical PN during the acute and delayed stages in a patient with bilateral horizontal gaze palsy. His acute PN was pure vertical PN augmented by visual fixation and vertical gaze, while this delayed PN showed asymmetric vertical movements with a torsional component that was suppressed during visual fixation. We speculated that the pathogeneses of acute vertical PN and OPT may differ since they had a different onsets and patterns.

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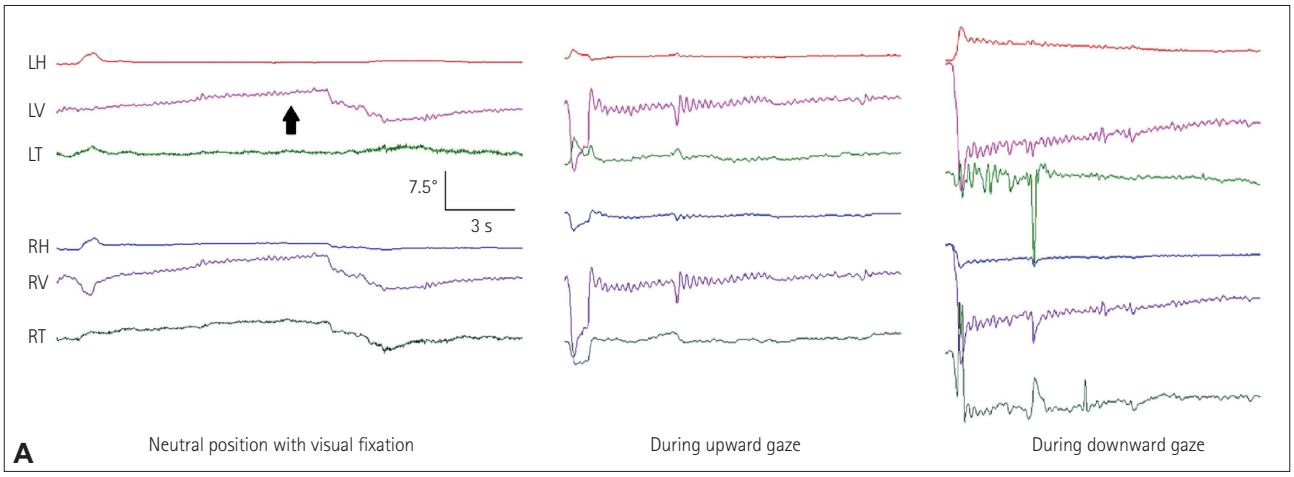

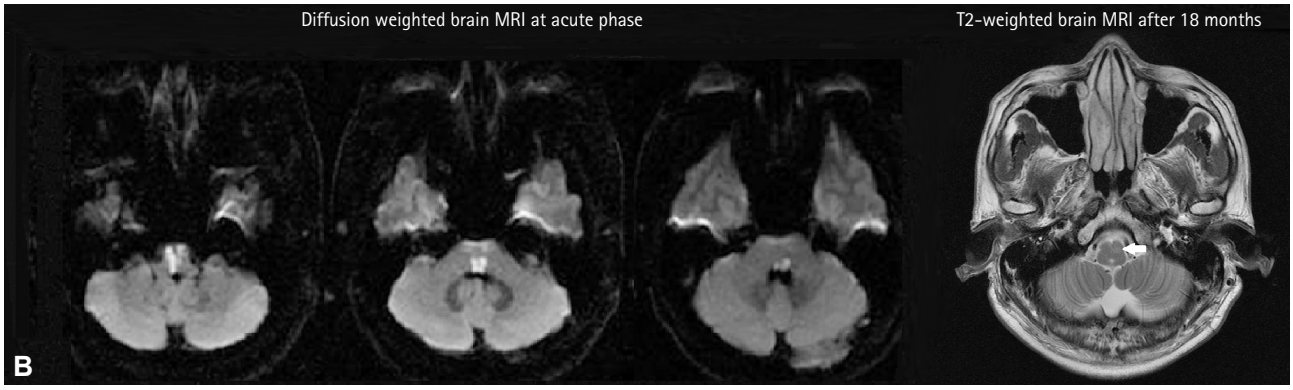

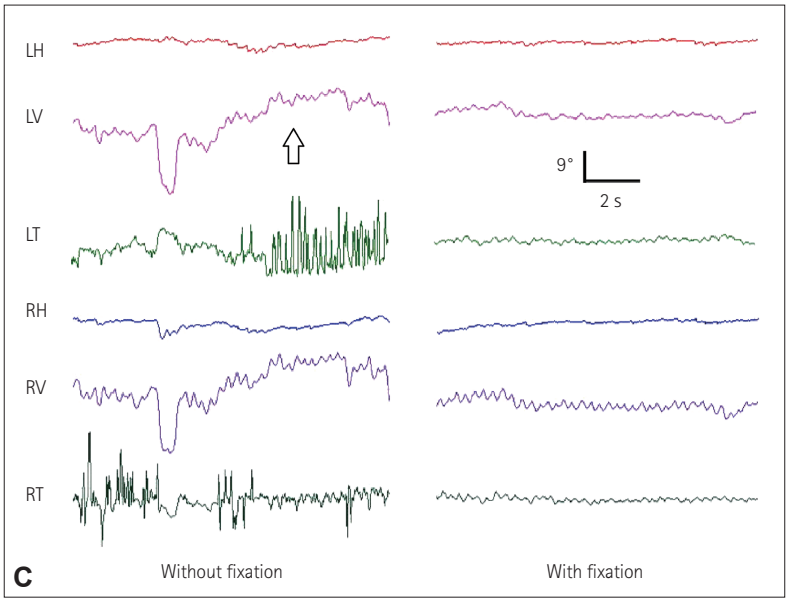

Fig. 1. Acute and chronic vertical PN in bilateral pontine lesion. A: Three-dimensional VOG during the acute period showed small-amplitude (0.5–1°) oscillations in both eyes at a frequency of 3–4 Hz during visual fixation. The amplitude increased during vertical gaze (up to 3–4°) (i.e., gaze-evoked nystagmus); note the slow upward deviation of both eyes (arrow) with corrected downward eye movement. B: Axial brain diffusion-weighted MRI performed during the acute phase showed hyperintense lesions involving both medial medullae extending to the tegmentum, and both paramedian caudal pontine tegmenta. Brain T2-weighted MRI performed 18 months later showed a subtle high-intensity signal in the inferior olivary nucleus (arrow). C: Three-dimensional VOG performed 18 months after the acute event showed asymmetric vertical PN at a frequency of 3–4 Hz and a higher amplitude in the right eye (2.5–3°). The amplitude was decreased during visual fixation (2–2.5°). The torsional component was more prominent in the delayed PN than in the acute PN. Note the slow upward deviation of both eyes in the absence of fixation (arrow). LH: horizontal position of left eye, LT: torsional position of left eye, LV: vertical position of left eye, PN: pendular nystagmus, RH: horizontal position of left eye, RT: torsional position of right eye, RV: vertical position of right eye, VOG: video-oculography.

A dual mechanism model has recently been suggested as a pathophysiologic mechanism of OPT.[3] Lesions in the Guillain-Mollaret triangle result in hypertrophy in the ION that causes the development of abnormal soma-somatic gap junctions.[4] The ION acts as a generator of PN, but its low-amplitude signal needs to be amplified/modulated by the deep cerebellar nuclei.[3] However, PN in the acute stage in our patient could not be explained by delayed soma-somatic gap enlargement. With the ION, paramedian tract (PMT) cell group act to optimize the control of gaze in response to the changing interface between the organism and its environment, and helps the role of the cerebellum in both the long-term adaptive and immediate control of eye movement.[4,5] Therefore, we can speculate that damage to PMT cell groups distorts the efforts to control gaze in a condition of horizontal gaze palsy, which may have produced acute PN during visual fixation in our patient. Moreover, PMT neurons in the pons are thought to participate primarily in vertical gaze holding, and the involvement of the PMT neurons might be related to the vertical GEN exhibited by the present patient.[6] A previous study observed acute vertical PN after pontine hemorrhage, which was attenuated during fixation,[7] and the pathomechanism underlying that case might have differed from that in our patient. Another study found acute vertical isolated PN after severe pontine strokes with ocular bobbing in patients with severe pontine strokes, and the authors speculated that this could have been caused by damage to the central tegmental tracts bilaterally.[8] Our patient showed upward deviation of both eyes in both the acute stage and during follow-up. Damage to the central tegmental tracts bilaterally could cause upward eye deviation but it cannot explain the vertical eye oscillation observed in our patient.

Supplementary Video Legend

Video 1. Symmetric and sinusoidal vertical eye movements are observed in both eyes during the acute phase. Vertical PN appears during visual fixation and is augmented during vertical gaze.

Video 2. Vertical PN does not appear at the 3-month follow-up.

Video 3. Asymmetric vertical PN with a torsional component and asymmetric palatal tremor are observed at the 18-month follow-up.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.3988/jcn.2020.16.1.163.

Author Contributions

Conceptualization: Hyun Ah Kim. Data curation: Hyun Ah Kim. Supervision: Hyun Ah Kim. Writing—original draft: Hyung Lee, Hyun Ah Kim. Writing—review & editing: Hyung Lee, Hyun Ah Kim.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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