



Epileptic Vertigo and Nystagmus in Cerebral Hemiatrophy Syndrome

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Dear Editor,

A 47-year-old right-handed woman presented with recurrent vertigo and visual hallucinations (rainbow colored lines) in the left hemifield, which had been present for 10 years. The patient experienced vertigo around a dozen times daily, but never lost consciousness or fell. She reported that the left half of her body had been weak since her childhood, but she had not received any medical evaluation or treatment. She did not show any sign of intellectual disability. The findings of a neurologic examination were unremarkable with the exception of mild left-sided weakness (Medical Research Council grade 4+) and brisk deep tendon reflexes. Dystonia, limb ataxia, and truncal ataxia were not observed. We performed video-oculography and video head impulse tests during the interictal period, which produced unremarkable findings for spontaneous nystagmus, gaze-evoked nystagmus, saccades, and video head impulse tests, while smooth pursuit was impaired bilaterally (Supplementary Fig. 1 in the online-only Data Supplement). During the ictus, left-beating nystagmus without altered consciousness immediately after the perception of visual hallucination was observed. The slow phase of nystagmus appeared to cross the midline. The ictal electroencephalogram (EEG) revealed fast rhythmic activity in the right occipital region (Fig. 1A and B, Supplementary Video 1 in the online-only Data Supplement). Brain MRI disclosed prominent atrophy of the right hemisphere in T1-weighted images and seizure-related cortical hyperintensities in T2- and diffusion-weighted images (Fig. 1C, D, and E). The asymmetry index of the left and right cerebral hemispheres obtained by a quantitative automated volumetric analysis was 13.1% (normal range: -1.1% to 2.2%).¹ The asymmetry index of the cerebellar hemisphere was within the normal range. Seizures occurred up to eight times per hour during the video-EEG monitoring, and this frequency was reduced to four times per day after the intravenous administration of levetiracetam (3,000 mg) and fosphenytoin (20 mg PE/kg). After adding clobazam (5 mg b.i.d.), the seizure frequency decreased to three times per month.

Cerebral hemiatrophy (also called Dyke-Davidoff-Masson syndrome) is a rare condition that presents with hemiparesis, hemidystonia, seizure, or mental retardation. The characteristic neuroimaging findings are diffuse unilateral cerebral atrophy/hypoplasia and unique bony changes such as thickening of the calvarium, enlarged paranasal sinuses, and elevation of the greater sphenoid wings.² Patients with cerebral hemiatrophy syndrome can present with epileptic seizures and usually respond poorly to anticonvulsants.^{3,4} Epileptic nystagmus has been ascribed to a disruption of cortical control over saccades or smooth-pursuit eye movements, which are attributed to epileptiform discharges originating from the cortical area. The direction of epileptic nystagmus was generally directed away from the epileptic focus that predominantly arose from the posterior part of the cerebral hemisphere.^{5,6} Epileptic vertigo may be considered when short and recurrent vertigo is accompanied by cortical symptoms such as visual hallucinations.

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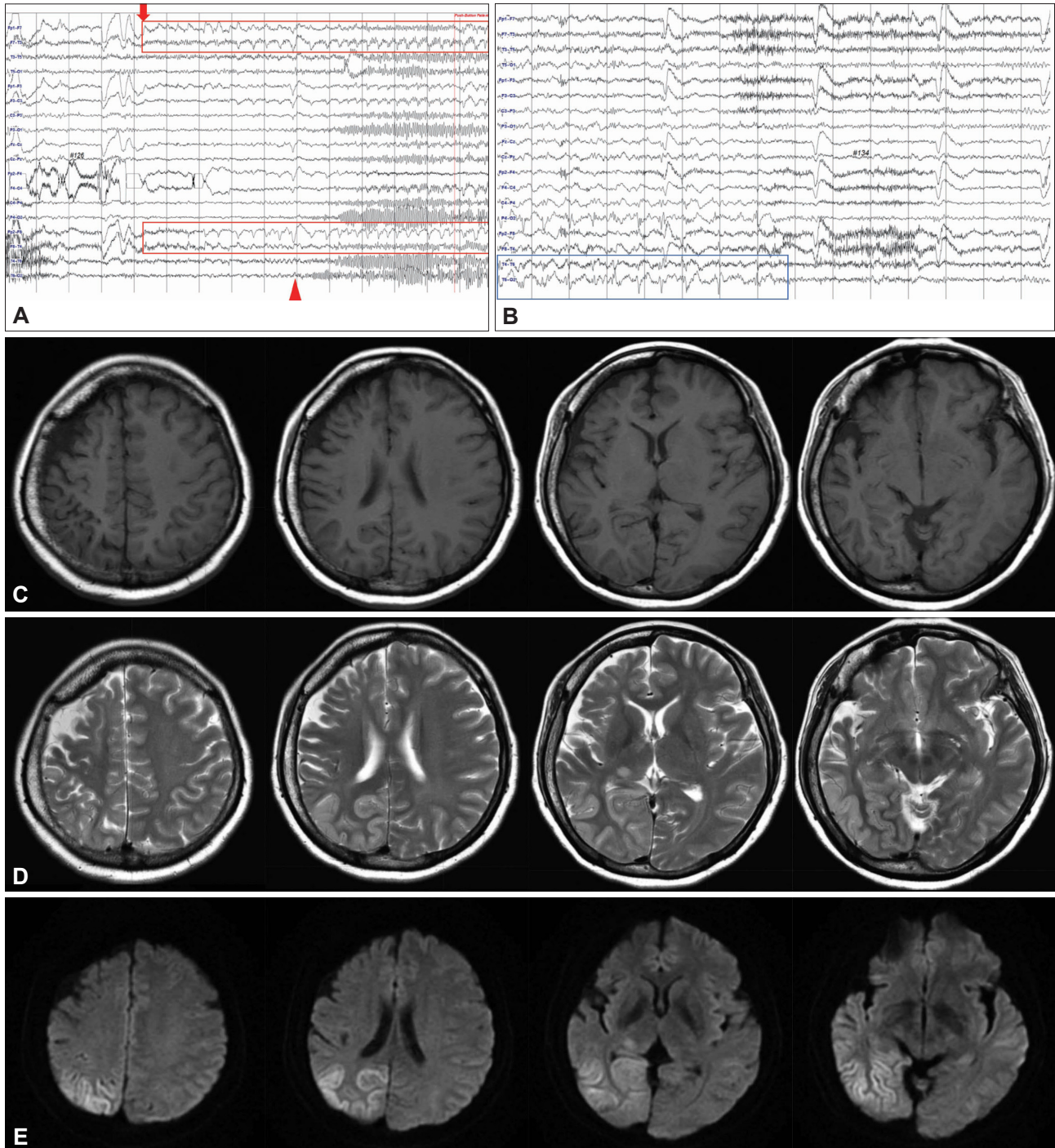


Fig. 1. Ictal electroencephalogram (EEG) and brain MRI. (A) Ictal EEG demonstrating the evolution of rhythmic fast activity (red arrowhead) at 18–20 Hz over the right occipital region within 6 seconds after the onset of nystagmus (red arrow). (B) At approximately 50 seconds after seizure onset, the EEG shows seizure offset as repetitive sharp-and-wave activities involving the right occipital region (blue box). (C) T1-weighted MRI demonstrating prominent atrophy with calvarial thickening of the right cerebral hemisphere. Seizure-related cortical hyperintensities were observed in the right parietal, occipital, temporal, and thalamic regions in (D) T2-weighted images and (E) diffusion-weighted images.

Supplementary Video Legend

Video 1. The video shows ictal left beating nystagmus with intact consciousness.

Supplementary Material

The online-only Data Supplement is available with this article at <https://doi.org/10.3988/jcn.2020.16.2.341>.

Author Contributions

Conceptualization: Hye-Jin Moon. Funding acquisition: Eek-Sung Lee, Hye-Jin Moon. Investigation: Keun Tae Kim. Writing—original draft: Eek-Sung Lee. Writing—review & editing: Tae-Kyeong Lee, Hye-Jin Moon.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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