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LETTER TO THE EDITOR

Revisiting the diagnostic value of Evans' index: lessons from an unusual case of normal pressure hydrocephalus with Evans' index less than 0.3

Jung E. Park[1] · Hyunjin Ju[2] · Kayeong Im[2] · Kyum-Yil Kwon[2]

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Idiopathic normal pressure hydrocephalus (iNPH) is a potentially treatable movement disorder and is diagnosed when Evans' index (EI) is greater than 0.3 [1, 2]. However, the most reliable biomarker of iNPH remains unknown and the diagnosis of iNPH has been regarded to be difficult for this reason. We describe a case of iNPH with EI of less than 0.3 presenting clinically as ventriculoperitoneal (VP) shunt–responsive gait abnormality.

A 77-year-old man was admitted to the hospital for evaluation of ambulatory difficulty with occasional falls while standing up or turning. The patient's daily medications included those for diabetes mellitus and hypertension, and there were no offending drugs for neurological disorders. On bedside examination, subtle signs of parkinsonism including postural instability were noted (see Video 1). We did not observe any falls during his hospitalization. Brain magnetic resonance imaging (MRI) revealed mild periventricular hyperintensities (Fig. 1a) and DESH (disproportionately enlarged subarachnoid space hydrocephalus) sign (Fig. 1b), while hydrocephalus was not documented (EI = 0.28). In addition, the patient's dopamine transporter (DAT) imaging was normal (Fig. 1c), thereby ruling out Parkinson's disease and atypical parkinsonism. While there was a clinical suspicion for iNPH, the patient was not intervened upon and was followed as an outpatient.

The patient's gait gradually worsened over the course of a year, and he sustained more frequent falls. On repeat examination, severe postural instability and freezing of gait were noted (see Video 1), although rigidity and bradykinesia were mildly pronounced in bilateral upper and lower extremities. The patient also reported subjective memory loss, for which a detailed neuropsychological assessment was conducted, revealing findings consistent with multi-domain amnestic mild cognitive impairment. Meanwhile, the patient did not complain of urinary difficulty. Follow-up brain MRI revealed a slight progression of initial MRI findings including narrow callosal angle over the high convexity and enlargement of temporal horns (Fig. 1D and E). Given the patient's clinical course, we had a high suspicion for iNPH, despite the EI value of 0.29. Multiple attempts to perform lumbar puncture, including several trials of fluoroscopy-guided lumbar puncture, were unsuccessful due to the patient's unfavorable anatomy. The patient underwent a VP shunt operation, resulting in dramatic improvement (see Video 2). The patient remained asymptomatic during his 8-month follow-up period.

The accurate diagnosis of iNPH has been regarded as challenging, and two international guidelines are currently used as diagnostic criteria [1, 2]. Both guidelines define ventriculomegaly as EI greater than 0.3 as the core feature of hydrocephalus. Accordingly, the EI of 0.29 in the patient's MRI was insufficient for hydrocephalus according to the current cut-off value for diagnosing iNPH. However, we concluded that the clinical diagnosis of the patient was iNPH for the following reasons: (1) he presented with progressive symptoms of gait/balance disturbance and cognitive decline; (2) other MRI findings including narrow callosal angle over the high convexity, enlargement of temporal horns, periventricular signal changes, and the DESH sign were found to be worsened over the following year with no therapeutic intervention; and (3) other parkinsonian disorders including progressive supranuclear palsy were excluded not only by brain MRI but also by DAT imaging [3, 4]. This case suggests

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✉ Kyum-Yil Kwon
denovo78@naver.com

[1] Department of Neurology, Dongguk University Ilsan Hospital, Dongguk University College of Medicine, Goyang, Republic of Korea

[2] Department of Neurology, Soonchunhyang University Seoul Hospital, Soonchunhyang University School of Medicine, Seoul 04401, Republic of Korea

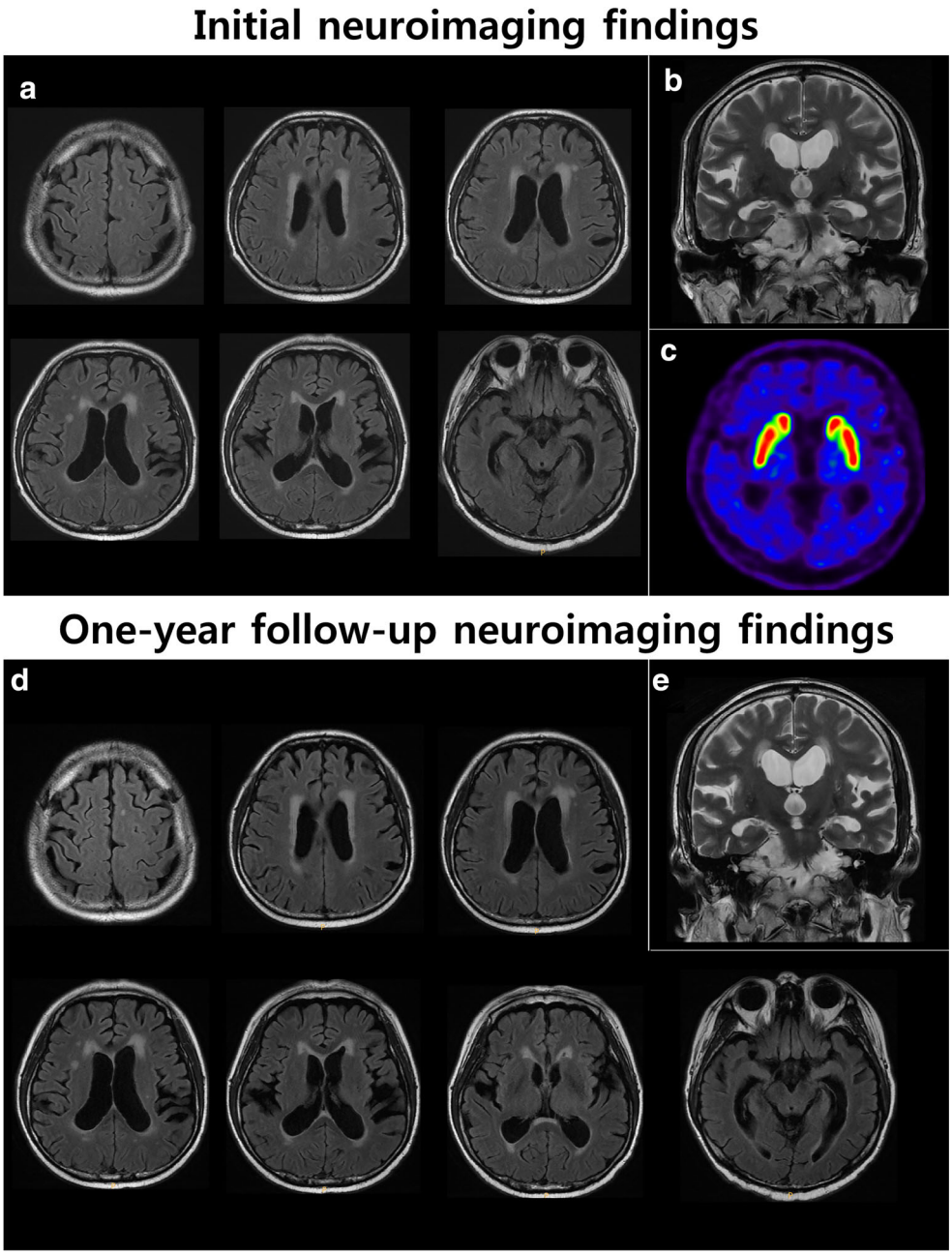

Fig. 1 Neuroimaging findings of the patient. (**a**) Axial FLAIR images show mild ventricular enlargement (the Evans' index = 0.28) and mild periventricular hyperintensity. (**b**) A coronal T2-weighted image reveals narrow callosal angle and DESH (disproportionately enlarged subarachnoid space hydrocephalus) sign. (**c**) A dopamine transporter PET scan with 18F-FP-CIT results in no abnormal finding of striatal dopamine uptake. (**d** and **e**) After 1 year, the previous MRI abnormalities of narrow callosal angle with high convexity tightness, the DESH sign, enlargement of temporal horns and lateral ventricle, and periventricular hyperintensity were slightly aggravated, although the change of hydrocephalus was not considerably prominent (the Evans' ratio = 0.29), compared to the alteration of clinical symptoms

that an EI of 0.3 for ventriculomegaly should not be uniformly applied, and it is also reasonable to infer that other MRI findings including DESH sign may even precede EI of 0.3 in certain cases with iNPH. One might also consider performing DAT imaging to exclude the possibility of Parkinson's disease or other Parkinsonian disorders to further support the clinical diagnosis of iNPH.

In conclusion, this unusual case of iNPH with EI of less than 0.3 suggests that the causal relationship between clinical symptoms and brain MRI findings should be interpreted with more caution in diagnosing iNPH. While the diagnostic value of EI has been questioned by previous studies, the current diagnostic criteria for iNPH include EI as an absolute requisite [5]. This case once again emphasizes the need to consider this diagnosis even in the setting of an insufficiently elevated EI, when the clinical picture is suggestive.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

Informed consent Written informed consent was obtained from the patient.

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