Teaching NeuroImage: Paravermal Lesions in Neuronal Intranuclear Inclusion Disease

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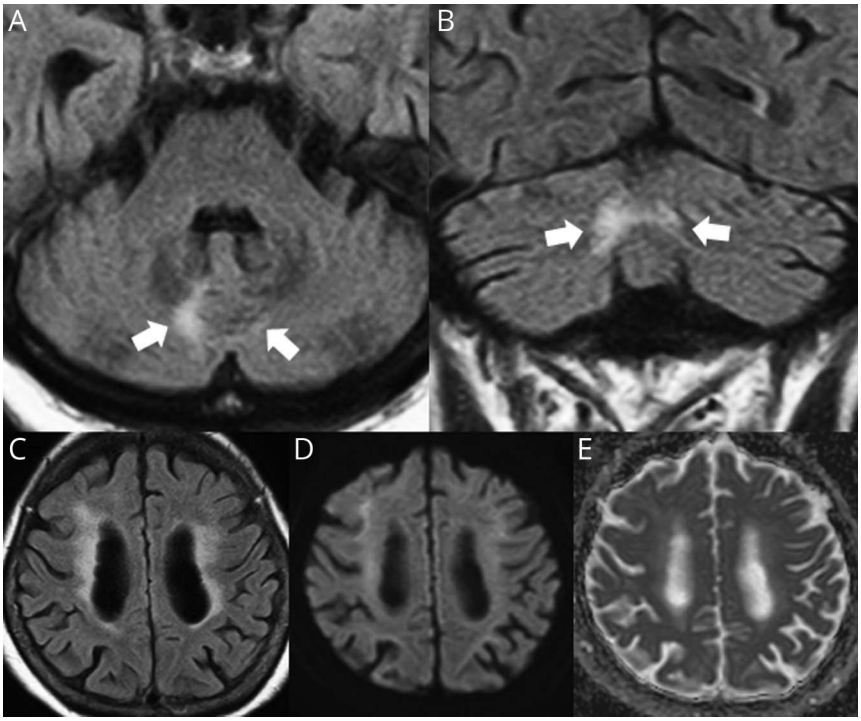
Figure 1 Brain MRI Without Contrast



Brain MRI showing a bilateral high-intensity signal in the medial part of the cerebellar hemisphere by the vermis (paravermal lesions, white arrows; A, B), cerebral white matter (C) on fluid-attenuated inversion recovery, a high-intensity signal along the corticomedullary junction on diffusion-weighted imaging (D) without restrictions to the apparent diffusion coefficient map (E).

A 77-year-old woman presented with a several-year history of progressive cerebellar ataxia and cognitive impairment. MRI revealed paravermal lesions on fluid-attenuated inversion recovery and a high-intensity signal along the corticomedullary junction on diffusion-weighted imaging (Figure 1). Abnormal expansion of GGC repeats in the *NOTCH2NLC* gene confirmed the neuronal intranuclear inclusion disease (NIID) diagnosis. NIID is a clinically heterogeneous neurodegenerative disorder usually occurring at age 50 years or older in sporadic cases. Paravermal lesions are a characteristic MRI finding in NIID.[1] Paravermal lesions are not specific to NIID alone (Supplement, links.lww.com/WNL/C182) but precede other imaging findings and can be the sole radiologic indication for NIID diagnosis.[2]

MORE ONLINE

Teaching slides
links.lww.com/WNL/C181

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Name	Location	Contribution
Atsuhiko Sugiyama, MD, PhD	Chiba University, Japan	Concept and design; drafted the manuscript
Jun Sone, MD, PhD	Aichi Medical University, Japan	Genetic analysis; revised the manuscript for intellectual content
Satoshi Kuwabara, MD, PhD	Chiba University, Japan	Revised the manuscript for intellectual content; supervised the study and gave the final approval

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