Teaching NeuroImage: Paravermal Lesions in Neuronal Intranuclear Inclusion Disease

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Figure 1 Brain MRI Without Contrast

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. 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.

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Appendix Authors

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