Teaching NeuroImages: Paravermal Lesions in Neuronal Intranuclear Inclusion Disease

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Jun Sone: Drafting/revision of the manuscript for content, including medical writing for content
Satoshi Kuwabara: Drafting/revision of the manuscript for content, including medical writing for content

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Case Summary
A 77-year-old female presented with a several year history of progressive cerebellar ataxia and cognitive impairment. MRI revealed paravermal lesions on fluid-attenuated inversion recovery and high signal intensity 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 ≥50 years in sporadic cases. Paravermal lesions are a characteristic MRI finding in NIID.\(^1\) Paravermal lesions are not specific to NIID alone (Supplement) but precede other imaging findings and can be the sole radiological indication for NIID.
diagnosis.²

**Appendix 1. Co-investigators**

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<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
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<tr>
<td>Atsuhiko Sugiyama, MD, PhD Chiba</td>
<td>University</td>
<td>Concept and design; drafted the manuscript.</td>
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<td>University</td>
<td>Genetic analysis; revised the manuscript for intellectual content.</td>
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<td>Satoshi Kuwabara, MD, PhD Chiba</td>
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<td>Revisited the manuscript for intellectual content; supervised the study and</td>
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<td>gave the final approval.</td>
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**Supplement** - [http://links.lww.com/WNL/C182](http://links.lww.com/WNL/C182)

**References**


Figure Legends
Figure 1. Brain MRI without contrast

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

![Brain MRI images](image-url)
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