Teaching NeuroImages: Paravermal Lesions in Neuronal Intranuclear Inclusion Disease

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Atsuhiko Sugiyama: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design
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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. Coinvestigators**

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atsuhiko Sugiyama, MD, PhD</td>
<td>Chiba University</td>
<td>Concept and design; drafted the manuscript.</td>
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<td>Jun Sone, MD, PhD</td>
<td>Aichi Medical University</td>
<td>Genetic analysis; revised the manuscript for intellectual content.</td>
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<tr>
<td>Satoshi Kuwabara, MD, PhD</td>
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<td>Revised the manuscript for intellectual content; supervised the study and gave the final approval.</td>
</tr>
</tbody>
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Teaching Slides - [http://links.lww.com/WNL/C181](http://links.lww.com/WNL/C181)
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).
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