Teaching NeuroImages: Paravermal Lesions in Neuronal Intranuclear Inclusion Disease

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Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication.

This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.

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Jun Sone: Drafting/revision of the manuscript for content, including medical writing for content
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Figure Count:
1

Table Count:
0

Search Terms:
[ 120 ] MRI, [ 161 ] All Movement Disorders

Acknowledgment:
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
2

Appendix 1. Coinvestigators

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<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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<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>
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Teaching Slides - http://links.lww.com/WNL/C181
Supplement - 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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Neurology published online July 8, 2022
DOI 10.1212/WNL.0000000000200984

This information is current as of July 8, 2022