RESIDENT & FELLOW SECTION

# Teaching NeuroImage: Dorsal Medullary Lesions in Juvenile-Onset Alexander Disease

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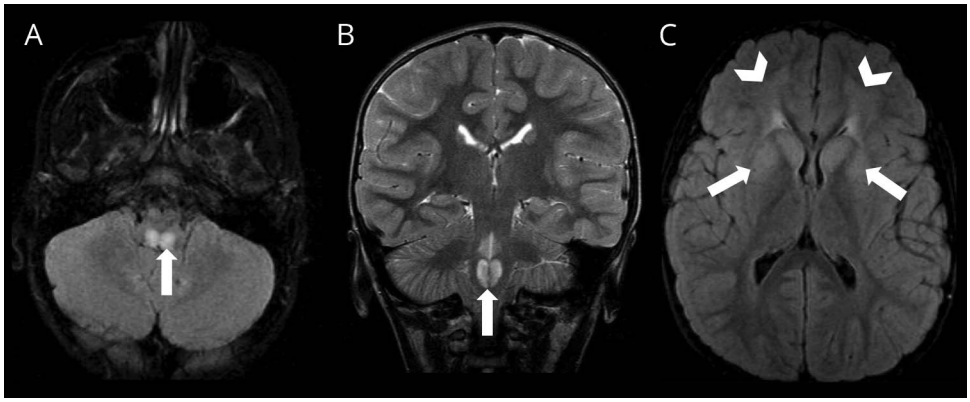
**Figure** Dorsal Medullary Lesions in Alexander Disease




Brain MRI in Alexander disease reveals distinctive hyperintense bilateral dorsal medullary lesions on (A) axial FLAIR and (B) coronal T2-weighted images in a heart-shaped appearance. (C) Additional diagnostic criteria (T2 hyperintense signal abnormality in the frontal white matter [arrow heads] and basal ganglia [arrows]) are present on axial FLAIR images.[1]

A 6-year-old boy presented with dysphagia, vomiting, and weight loss. Early developmental milestones were notable for mild gross motor and speech delay. Hypotonia was present on examination. Brain MRI revealed bilateral enhancing dorsal medullary lesions (figure, contrast not shown). The differential diagnosis included a leukodystrophy or mitochondrial disease. Alexander disease was confirmed genetically (de novo variant in *GFAP*-targeted testing: p.Arg-376-Gly). Typical features also include hypernasal speech with subsequent motor difficulties and autonomic dysfunction over time.[2] *GFAP* sequencing should be considered in patients with unilateral or bilateral dorsal medullary lesions with localizing symptoms (e.g., vomiting and dysphagia).

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**Teaching slides**
links.lww.com/WNL/B454

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## Disclosures
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## Appendix Authors

| Name | Location | Contribution |
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| **John Sollee, BS** | Warren Alpert Medical School of Brown University, Providence, RI | Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data |
| **Amy Waldman, MD, MSCE** | Division of Neurology, Children's Hospital of Philadelphia, PA | Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data |

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