Teaching NeuroImages: Infantile-onset Krabbe disease with tigroid appearance of the white matter

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Figure Imaging

An 8-month-old girl presented with a 1-month history of developmental regression, irritability, and opisthotonic posturing. A brain MRI revealed abnormal T2 hyperintensities in the periventricular and deep cerebral white matter, and peridentate cerebellar white matter. Reduced galactocerebrosidase activity in leukocytes confirmed the diagnosis of Krabbe disease.

Though typically characteristic of metachromatic leukodystrophy (MLD), the tigroid MRI pattern can also be seen in Pelizaeus-Merzbacher and Krabbe disease. Clinically, irritability, opisthotonos, and developmental regression, and radiographically, early cerebellar involvement, decreased thalamic T2 signal, and, if present, optic nerve enlargement, can help differentiate MLD from Krabbe disease (figure).
Appendix

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<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
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<tr>
<td>E. Corina Andriescu, BS</td>
<td>The University of Texas Health Science Center at Houston</td>
<td>Designed, conceptualized, and drafted the manuscript</td>
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Disclosure
E. Corina Andriescu, Dr. Russo, and Dr. Pérez report no disclosures. Go to Neurology.org/N for full disclosures.

References
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