

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

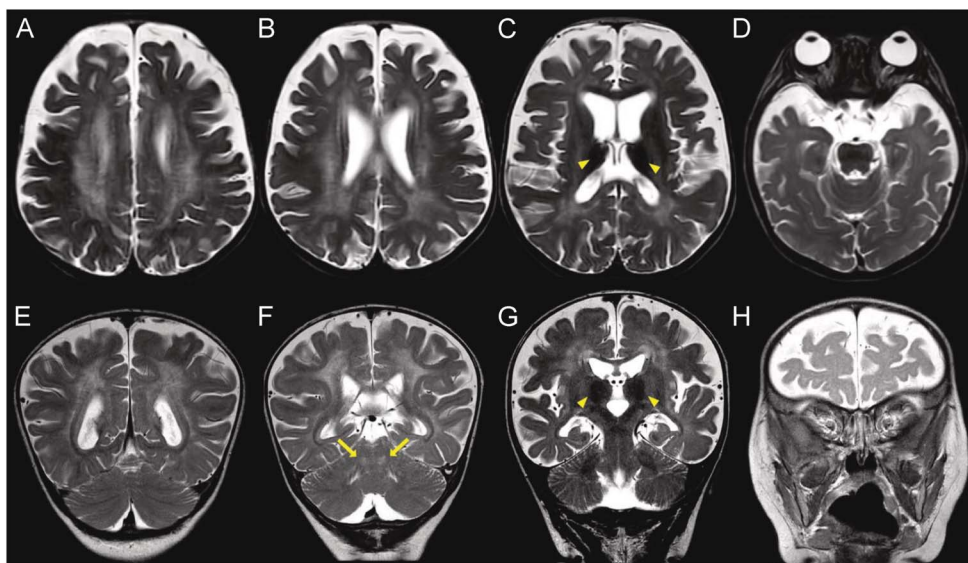
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Figure Imaging



(A–D) Axial T2-weighted views. (A, B) Bilateral symmetric hyperintensity of the periventricular and deep cerebral white matter with a tigroid appearance. (C) Bilateral T2 thalamic hypointensities (arrowheads) and predominant parieto-occipital white matter involvement. (D) Normal, nonenlarged optic nerves. (E–H) Coronal T2-weighted views. (E) Bilateral symmetric hyperintensity of the periventricular and deep cerebral white matter with a tigroid appearance. (F) Peridentate cerebellar white matter hyperintensities (arrows). (G) Bilateral T2 thalamic hypointensities (arrowheads). (H) Normal, nonenlarged optic nerves.

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.^{1–3} 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).

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Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

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Carlos A. Pérez, MD	The University of Texas Health Science Center at Houston	Critically reviewed the manuscript for intellectual content and contributed equally to the drafting of the manuscript

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Disclosure

E. Corina Andriescu, Dr. Russo, and Dr. Pérez report no disclosures. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

References

1. Duffner PK, Barczykowski A, Jalal K, et al. Early infantile Krabbe disease: results of the World-Wide Krabbe Registry. *Pediatr Neurol* 2011;45:141.
2. Wenger DA. Krabbe disease. *GeneReviews*. Available at: [ncbi.nlm.nih.gov/books/NBK1238/](https://www.ncbi.nlm.nih.gov/books/NBK1238/). Accessed December 14, 2019.
3. Muthusamy K, Sudhakar SV, Thomas M, et al. Revisiting magnetic resonance imaging pattern of Krabbe disease: lessons from an Indian cohort. *J Clin Imaging Sci* 2019;9:25.

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