Teaching NeuroImages: Selective corticospinal tract involvement in late-onset Krabbe disease

R. Sehgal, MD*
S. Sharma, DM*
N. Sankhyan, DM
A. Kumar, MD
S. Gulati, MD

Address correspondence and
reprint requests to Dr. Sheffali
Gulati, Division of Pediatric
Neurology, Department of
Pediatrics, All India Institute of
Medical Sciences, New Delhi
110029, India
sheffaligulati@gmail.com

Figure Brain MRI

Axial (A) and coronal (B) fluid-attenuated inversion recovery MRI scans show bilateral symmetric hyperintense signal changes involving the corticospinal tracts. (C) Sagittal T2-weighted MRI shows the hyperintense signal along the corticospinal tract from the cortical area extending into the internal capsule.

A 4-year-old girl presented with increasing falls and progressive limb stiffness noticed for 2 months. There was no history of cognitive decline, seizures, or vision impairment. Examination revealed spastic quadriparesis, with no sensory or cerebellar abnormalities. A suggestive MRI scan of the brain (figure) and reduced galactocerebrosidase activity in leukocytes confirmed the diagnosis of late-onset Krabbe disease. MRI in late-onset Krabbe disease classically shows parieto-occipital periventricular white matter and posterior corpus callosal signal changes with sparing of subcortical U fibers and cerebellar white matter.¹ Isolated corticospinal tract involvement is an unusual pattern described in adult- and late-onset forms of Krabbe disease.¹ In the setting of childhood neurodegeneration, similar changes have also been described in X-linked adrenoleukodystrophy, acyl-coenzyme A (CoA) oxidase

deficiency, and 3-hydroxy-3-methylglutaryl-CoA lyase deficiency.^{2,3}

AUTHOR CONTRIBUTIONS

R.S., S.S., and N.S. did the clinical/diagnostic workup of the child and reviewed the literature. A.K. provided neuroradiologic inputs. R.S., S.S., N.S., A.K., and S.G. contributed to the content of the manuscript. S.G. was in charge of the case overall and provided final approval of the manuscript.

REFERENCES

1. Loes DJ, Peters C, Krivit W. Globoid cell leukodystrophy: early-onset from late-onset disease using a brain MR imaging scoring method. *AJNR Am J Neuroradiol* 1999;20:316–323.
2. Suzuki Y, Iai M, Kamei A, et al. Peroxisomal acyl CoA oxidase deficiency. *J Pediatr* 2002;140:128–130.
3. Yılmaz Y, Ozdemir N, Ekin G, Baykal T, Kocaman C. Corticospinal tract involvement in a patient with 3-HMG coenzyme A lyase deficiency. *Pediatr Neurol* 2006;35:139–141.

*These authors contributed equally to this work.

From the Division of Pediatric Neurology, Department of Pediatrics (R.S., S.S., N.S., S.G.), and Department of Radiodiagnosis (A.K.), All India Institute of Medical Sciences, New Delhi, India.

Disclosure: Dr. Sehgal, Dr. Sharma, Dr. Sankhyan, and Dr. Kumar report no disclosures. Dr. Gulati serves on the editorial board of the *Indian Journal of Pediatrics* and has received research support from the NIH, Autism Speaks, and National Trust.

Neurology®

Teaching *NeuroImages*: Selective corticospinal tract involvement in late-onset Krabbe disease

R. Sehgal, S. Sharma, N. Sankhyan, et al.

Neurology 2011;77:e20

DOI 10.1212/WNL.0b013e318225aaf5

This information is current as of July 18, 2011

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/77/3/e20.full
References	This article cites 3 articles, 1 of which you can access for free at: http://n.neurology.org/content/77/3/e20.full#ref-list-1
Citations	This article has been cited by 1 HighWire-hosted articles: http://n.neurology.org/content/77/3/e20.full##otherarticles
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Cognitive Disorders/Dementia http://n.neurology.org/cgi/collection/all_cognitive_disorders_dementia All Demyelinating disease (CNS) http://n.neurology.org/cgi/collection/all_demyelinating_disease_cns All Pediatric http://n.neurology.org/cgi/collection/all_pediatric MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright Copyright © 2011 by AAN Enterprises, Inc.. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.