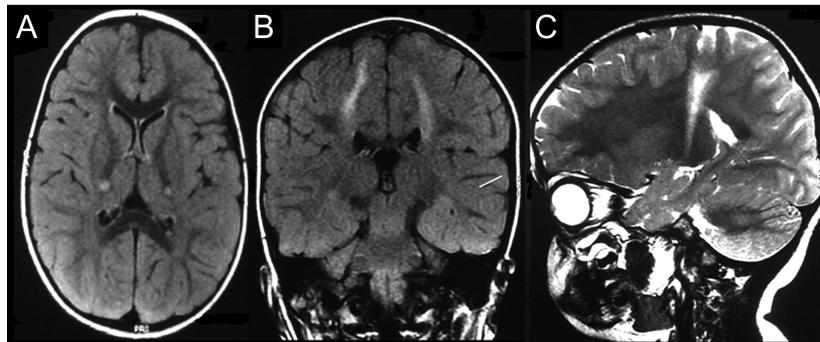


Teaching NeuroImages: Selective corticospinal tract involvement in late-onset Krabbe disease

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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.

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