Bright tongue sign in Pompe disease

Figure Brain MRI

Sagittal T1 MRI of the brain shows abnormal diffuse T1 hyperintensity of the tongue musculature.

A 38-year-old man was referred for progressive gait difficulty. A brain MRI was obtained as part of his workup prior to referral. Upon review of the brain MRI—reported normal by the radiologist—the tongue looked bright on the T1 sequence (figure). Investigation revealed that he had Pompe disease. Bright tongue sign is well-known in patients with bulbar amyotrophic lateral sclerosis but also can be seen in other neuropathies or myopathies that can affect the tongue muscle. Attention to abnormalities of the tongue, even on nondedicated brain MRI studies, may help neurologists with the diagnosis.¹

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