“Bright tongue sign” in ALS

Figure Brain MRI

Sagittal T1 MRI of the brain shows abnormal diffuse T1 hyperintensity of the tongue musculature (left). A normal tongue is shown on the right.

A 62-year-old woman presented with 1 year of slowly progressive dysarthria, difficulty chewing food, sialorrhea, dysphagia, and unintentional 20-pound weight loss. Neurologic examination demonstrated moderate dysarthria, marked tongue weakness with atrophy and fasciculations, multifocal muscular atrophy, and diffuse hyperreflexia, including a jaw jerk. EMG showed diffuse fibrillation potentials and positive sharp waves. The history, examination, and EMG results fulfilled diagnostic criteria for amyotrophic lateral sclerosis. Brain MRI showed pronounced T1 hyperintensity of the tongue, consistent with chronic denervation of the tongue musculature with fatty replacement (figure).^{1,2}

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