Teaching NeuroImages: Mills syndrome

Metabolic and structural neuroimaging aids to the diagnostic

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Figure Left motor cortex iron deposition and hypometabolism and asymmetry between pyramidal tracts

Axial susceptibility-weighted image shows hypointensity in the left precentral cortex (A, arrow), signaling neurodegeneration-related abnormal iron deposition, accompanied by hypometabolism on [^{18}F]FDG-PET (B, arrow), also seen on quantitative 3D stereotactic surface projection analysis (C). Tractography (D) demonstrates asymmetry between corticospinal tracts (reduced fiber density on the left, dark blue).

Examination of a 57-year-old woman with a 3-year history of progressive right-sided weakness (first arm, then leg) and dysarthria disclosed right-sided weakness, spasticity, hyperreflexia, and Babinski sign. There were no signs of lower motor neuron disease or sensory involvement on clinical and electroneuromyographic examinations. MRI and [^{18}F]FDG-PET results are shown in the figure. CSF and serum screening were normal. Mills syndrome (progressive hemiparesis) due to primary lateral sclerosis is a rare phenotype of motor neuron disease.¹ Neuroimaging is essential to not only rule out alternative etiologies but also demonstrate functional and anatomic involvement of pyramidal tract.²

AUTHOR CONTRIBUTIONS

Fábio H.G. Porto: study concept and design, acquisition of data, and analysis and interpretation. Artur M. Coutinho: study concept and design, acquisition of data, and analysis and interpretation. Leandro T. Lucato: study

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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