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Primary Lateral Sclerosis Presenting With Focal Onset Spreading Through Contiguous Neuroanatomic Regions

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A 66-year-old woman complained a 3-year history of left ankle focal spasticity. Brain MRI revealed subcortical white matter hyperintensity in the medial region of the right precentral gyrus. Subsequently, motor impairment spread proximally, to the contralateral lower limb and then to ipsilateral arm, ultimately resulting in a progressive asymmetric tetraparesis and a mild dysarthria. After a 4-year follow-up, a diagnosis of definite Primary Lateral Sclerosis was established.¹ Clinical progression was mirrored by corresponding spreading of the MRI hyperintensity through motor areas (Figure 1A) and extensive microstructural damage of the corticospinal tract shown by diffusion tensor (DT) MRI tractography (Figure 1B), consistently with a model of TDP-43 protein prion-like transmission through the complex 3D anatomy of the motor system.²

Figure legend

Figure [A] Conventional MRI. T2-weighted/FLAIR sequences revealing subcortical white matter hyperintensity in the medial right motor cortex, spreading over time to contiguous right frontal and left motor regions, mirroring clinical progression. **[B] DT MRI analysis.** DT MRI tractography analysis revealing fractional anisotropy (FA) decrease and mean diffusivity (MD) increase along the corticospinal tract, indicating the absence of obstacles to free diffusion of water molecules due to diffuse loss of white matter fiber integrity compared to 13 age-matched healthy female subjects (see eTable 1, eMethods and eResults in the Supplement for further information,<http://links.lww.com/WNL/B802>).

A. Conventional MRI

B. DT MRI

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Ethics approval

The study was performed in accordance with the ethical standards statement.

Informed consent: the patient provided written informed consent.

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