

Neuroimaging in amyotrophic lateral sclerosis

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DESCRIPTION

A 54-year-old man presented with a 4-month history of gait difficulty and progressive leg weakness, predominantly on the left side. A neurological examination revealed slurred speech, tongue fasciculations, spastic weakness and hyper-

reflexia of both lower extremities. A left Babinski sign and ipsilateral foot drop were also noted. Positive sharp waves, fasciculations and fibrillation potentials indicative of active and chronic denervation were seen on electromyography. An MRI of the brain revealed bilateral signal hyperintensities

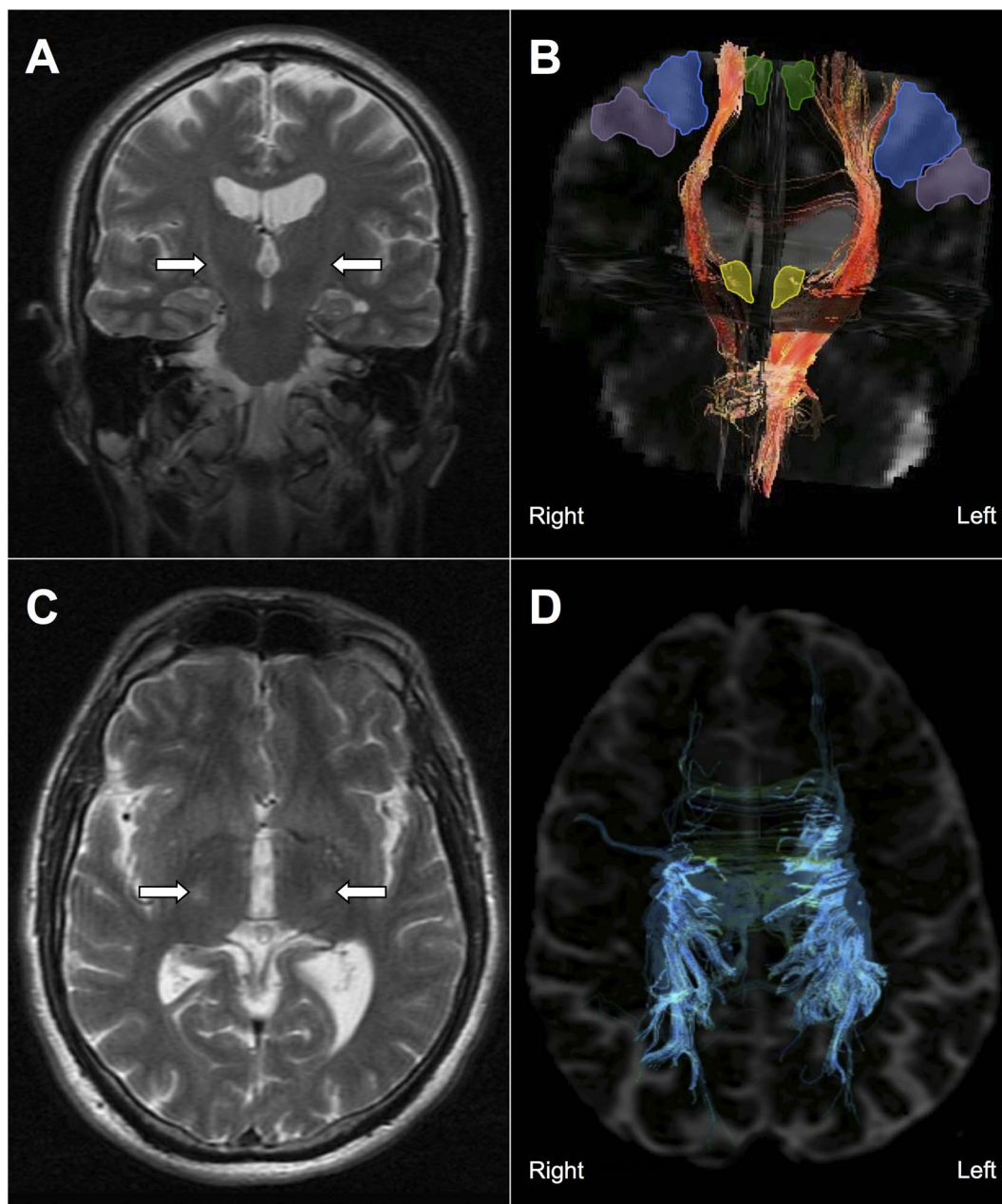


Figure 1 MRI and diffusion tensor tractography of a 54-year-old man with amyotrophic lateral sclerosis. (A and C) T2-weighted coronal and axial MRI of the brain revealed bilateral signal hyperintensities along the corticospinal tracts (arrows). (B and D) Diffusion tensor tractography showed asymmetry of the corticospinal tracts with fewer fibres on the right compared with the left side. In (B), the coloured regions illustrate the anatomical relationship of the corticospinal tract (shown in orange) with the thalamus (yellow) and the various motor areas of the cortex: leg (green), hand (blue) and face (purple).



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along the corticospinal tracts (CST; figure 1A,C). Tractography showed volume loss of the CST with a lower number of fibres on the right side (figure 1B,D). A diagnosis of the amyotrophic lateral sclerosis (ALS) was performed based on the clinical and neurophysiological evidence of upper and lower motor neuron involvement. His symptoms gradually progressed over several months and caused increasing disability.

ALS is an idiopathic neurodegenerative syndrome reflecting motor neuron loss.¹ Its diagnosis still largely depends on clinical criteria; nevertheless neuroimaging can provide important insights into the structural and functional changes associated with this disease.²

MRI is usually unremarkable in ALS, although hyperintensities along CST on T2-weighted or fluid-attenuated inversion recovery images have been described. This finding is believed to reflect degeneration of CST.³

Tractography is a promising MRI technique, which provides a volumetric analysis of CST and allows early identification of pyramidal tract abnormalities.² Neurological signs in this patient were more noticeable on the left side of his body, which correlates with the lower number of corticospinal fibres seen on the right side.

ALS is a relentlessly progressive disorder. Symptomatic treatment is the cornerstone of management.¹

Learning points

- ▶ Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterised by the combination of upper and lower motor neuron signs.
- ▶ Appropriate use of neuroimaging techniques may add valuable information to the current clinical and neurophysiological assessment of ALS.
- ▶ Symptomatic treatment with the aim of improving quality of life is the mainstay of therapy for patients with ALS.

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Competing interests None.

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