A distinct imaging phenotype in amyotrophic lateral sclerosis confidently detected on T1 MTC

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Accepted 12 November 2014

DESCRIPTION

A 57-year-old woman presented with a progressing asymmetric (left to right) weakness and wasting that spread in the next 3 months from her lower to upper limbs; she exhibited mild hyper-reflexia in all four limbs. No cognitive or sensory impairment was detected. Cerebrospinal fluid and blood analyses were unremarkable. Electromyography showed pathological signs of denervation and chronic reinnervation with spontaneous activity at rest, consistent with a motor neuron disease. The presence of upper motor neuron (UMN) and lower motor neuron (LMN) signs together in multiple regions, on clinical and paraclinical basis, supported amyotrophic lateral sclerosis (ALS) diagnosis, according to current criteria.[1] An MRI was performed and abnormal corticospinal tract signal intensity was remarkably detected on T1 magnetisation transfer contrast (MTC) images (figure 1A–C), which supported the presumptive diagnosis of UMN degeneration. The current diagnostic criteria have recently been introduced to better define LMN degeneration; however, paraclinical proof of UMN compromise remains a challenge, particularly in the early stages of the disease. In searching for UMN biomarkers, some conventional and non-conventional MRI techniques have emerged in routine clinical practice to demonstrate UMN degeneration in ALS.[1]

While proton density-weighted imaging (figure 2A–C) has only moderate sensitivity for ALS diagnosis, a newer MTC technique based on the exchange between stationary (macromolecules) and mobile protons (free water) has been elucidated to highlight structural abnormalities not detected on conventional techniques.[2] T1 MTC is better able to demonstrate the hyperintensity of the corticospinal tracts in a particular phenotype of imaging in early disease with UMN involvement.[2 3]

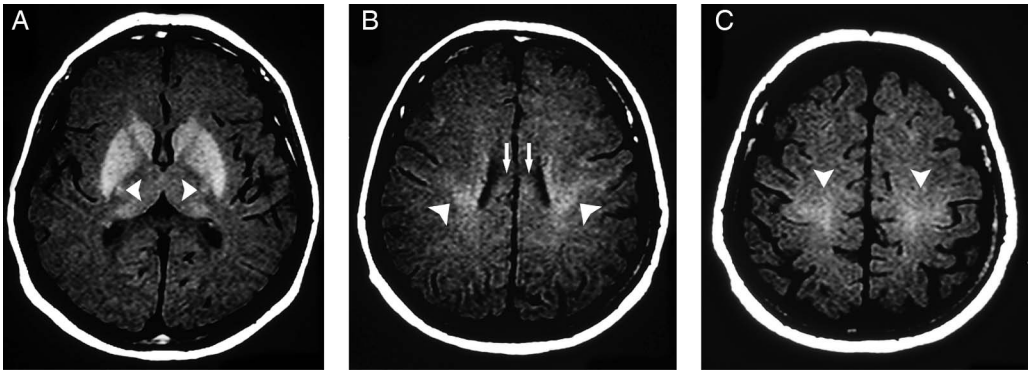

Figure 1 Axial T1 magnetisation transfer contrast (MTC; A–C). Selective hyperintensity (arrowheads) throughout the corticospinal tracts is depicted on T1 MTC axial images, assuming a typical 'W-like appearance' crossing the corpus callosum (arrows).

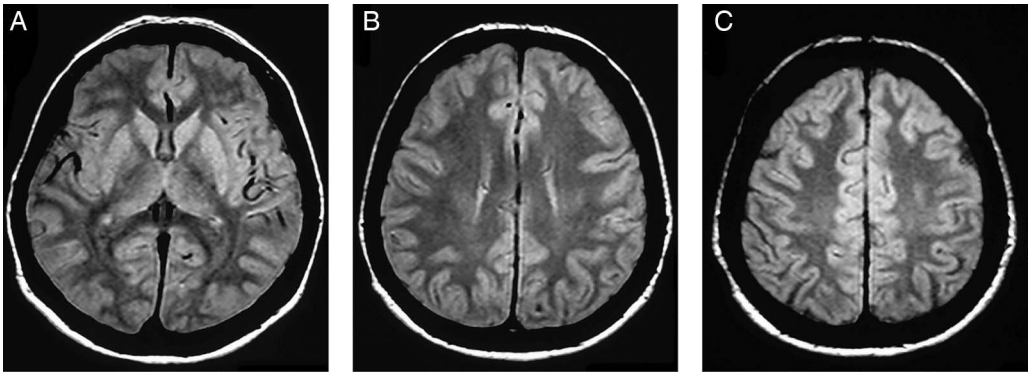

Figure 2 Axial proton density-weighted imaging (PDWI; A–C). PDWI only revealed a faint hyperintensity in the same regions, making the diagnosis more difficult.

To cite: da Rocha AJ, Nunes RH. *BMJ Case Rep* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2014-206511

Learning points

- The early diagnosis of amyotrophic lateral sclerosis might be a challenge; MRI has a potential role as a paraclinical tool for prompt detection of UMN involvement.
- In this case, the MRI signal intensity was useful for detecting upper motor neuron degeneration based on corticospinal tract signal intensity, reinforcing the relevance of the T1 magnetisation transfer contrast (MTC) to demonstrate corticospinal tract degeneration and corroborate an amyotrophic lateral sclerosis diagnosis in a single patient.
- Selective hyperintensity throughout the corticospinal tracts is depicted on T1 MTC axial images, assuming a typical 'W-like appearance' crossing the corpus callosum.

Contributors AJR and RHN were responsible for the study concept, drafting the manuscript, and for acquisition and interpretation of data.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 de Carvalho M, Dengler R, Eisen A, *et al*. Electrodiagnostic criteria for diagnosis of ALS. *Clin Neurophysiol* 2008;119:497–503.
2 da Rocha A J, Oliveira A S, Fonseca R B, *et al*. Detection of corticospinal tract compromise in amyotrophic lateral sclerosis with brain MR imaging: relevance of theT1-weighted spin-echo magnetization transfer contrast sequence. *AJNR Am J Neuroradiol* 2004;25:1509–15.
3 Carrara G, Carapelli C, Venturi F, *et al*. A distinct MR imaging phenotype in amyotrophic lateral sclerosis: correlation between T1 magnetization transfer contrast hyperintensity along the corticospinal tract and diffusion tensor imaging analysis. *AJNR Am J Neuroradiol* 2012;33:733–9.