

‘Wine-glass’ sign

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A 56-year-old woman presented with a nine-month history of rapidly progressive limb weakness. Initial examination revealed spastic tetraparesis, dysarthria and dysphagia. Clinical and electrophysiological signs of lower motor neuron (LMN) affection were absent. Brain magnetic resonance imaging (MRI) demonstrated ‘wine-glass’ sign with symmetrical hyperintensities on T2 and

FLAIR sequences, extending along the corticospinal tracts reaching the pontine level (Fig. 1). Immunological panel, onconeural antibodies (Hu, Yo, Ri), very long chain fatty acids, and ganglioside antibodies were all normal. Four months after discharge, the patient underwent control electromyography showing denervation and LMN affection. Six months later, the patient died. According to

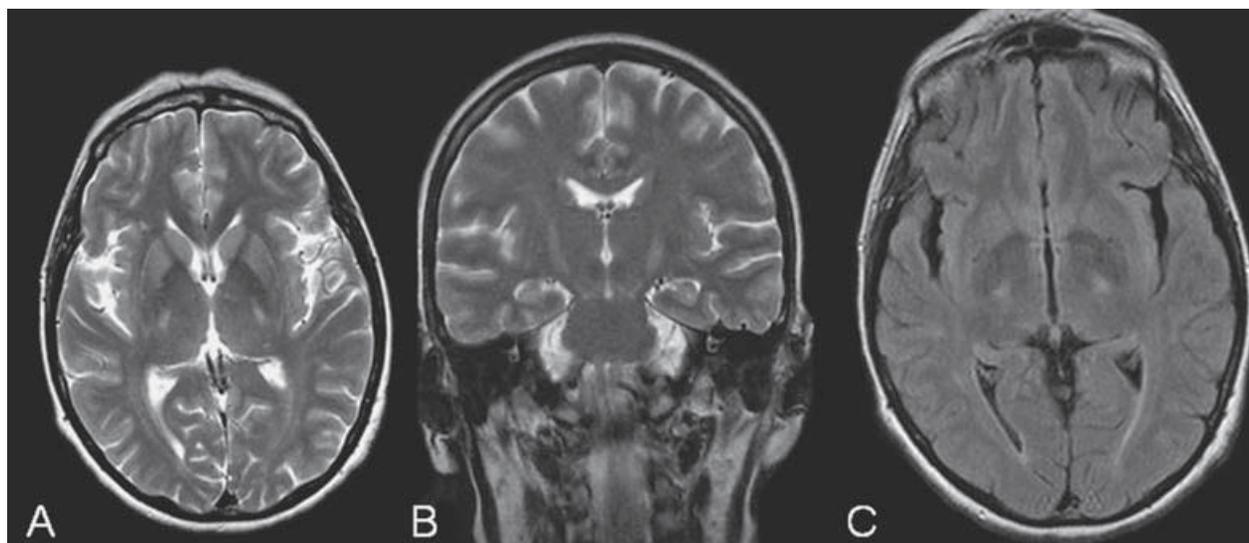


Fig. 1 (A, C). Axial T2 and FLAIR weighted MR images showing bilateral hyperintensities in posterior limb of the internal capsule; (B) coronal T2-weighted MR image showing ‘wine-glass’ sign with symmetrical corticospinal tract hyperintensities reaching the pontine level.

clinical picture and disease progression, the diagnosis of amyotrophic lateral sclerosis (ALS) was made.

According to current guidelines, use of conventional MRI in patients suspected of having a motor neuron disease is restricted to exclude other possible causes (1). Even more advanced neuroimaging techniques, such as diffusion tensor imaging and proton magnetic resonance spectroscopic imaging, have only modest discriminatory capability in making the diagnosis of ALS (2). Corticospinal tract hyperintensities are not disease-specific, however, lacking a specific image-biomarker for ALS, a unique neuroradiological finding such as 'wine-glass' sign should not be ignored and should complement clinical finding in supporting appropriate diagnosis.

Key words: amyotrophic lateral sclerosis, corticospinal tract hyperintensities, wine-glass sign

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