Case report

MRI findings in a patient with upper motor neuron dominant amyotrophic lateral sclerosis

A S Pallewatte[1], **I K Gooneratne**[2], **P Weeratunga**[2], **R Gamage**[2]

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Introduction

Amyotrophic lateral sclerosis (ALS) is part of the clinical spectrum of motor neuron disease and is a progressive neurodegenerative disorder with muscle wasting and paralysis which ultimately results in death. Upper motor neuron dominant ALS (UMN-ALS) is part of the clinical spectrum of ALS.

Case report

A 55-year-old previously healthy female presented with progressive difficulty in speech and walking for 8 months. There were no risk factors for vascular disease, or features of autoimmune disease. There was no family history of neurological disease.

On examination she had a spastic speech, pseudobulbar palsy and quadrispasticity with hyperreflexia. There was no demonstrable wasting or fasciculations in any muscle group. Cerebellar signs were negative. She did not have any evidence of Parkinsonism. Cognitive function was normal and bladder and bowel was continent. There was no evidence of sensory dysfunction. Ophthalmoscopy was normal. Other systems examination was normal.

Electromyography (EMG) showed mild denervation changes. Magnetic resonance imaging (MRI) with T2 weighted axial image demonstrated bilateral symmetrical hyperintensity indicating involvement of both corticospinal tracts in the periventricular white matter, posterior internal capsule and cerebral peduncles (Figure). FLAIR coronal MRI images also showed similar bilateral symmetrical involvement of corticospinal tracts. Investigations including full blood count, fasting

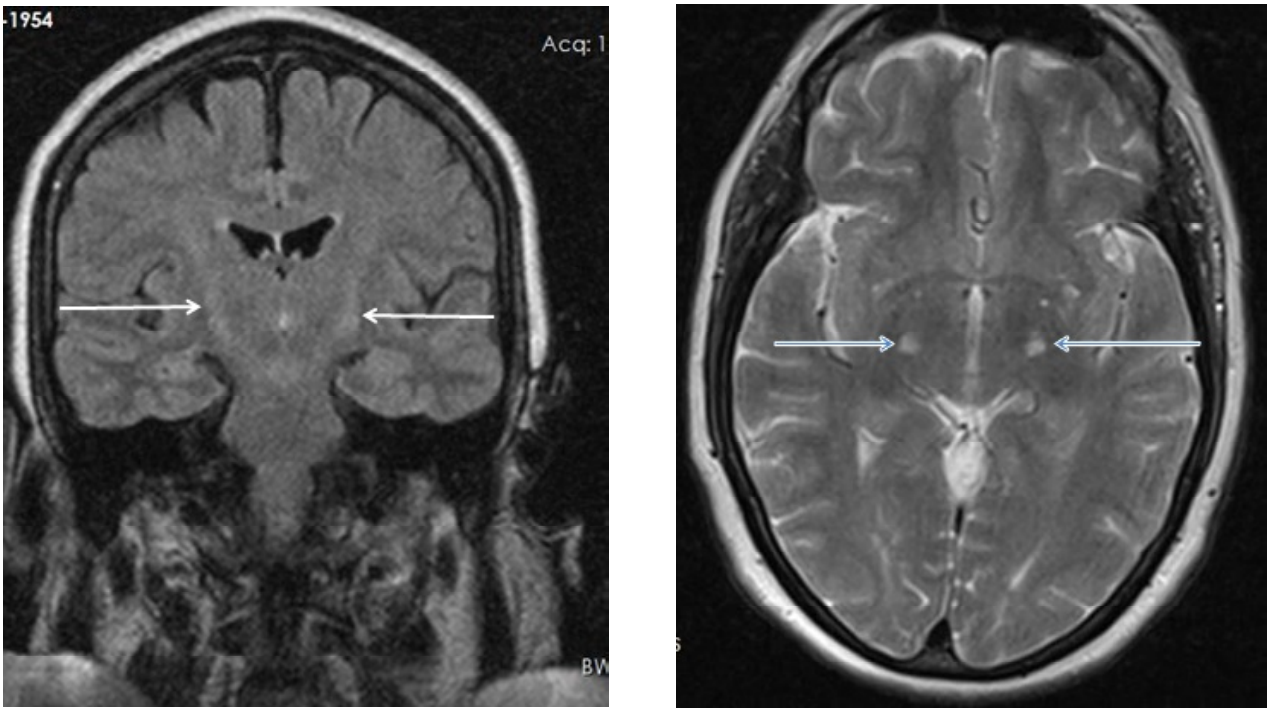

Figure. T2 axial image showing symmetrical hyperintensity indicating involvement of both corticospinal tracts in posterior internal capsule.

[1]*Department of Radiology,* [2]*Institute of Neurology, National Hospital of Sri Lanka.*
Correspondence: IK Gooneratne E-mail: kishig@gmail.com

blood glucose, renal and liver function tests, and autoimmune profile were unremarkable. HIV screening was negative and VDRL was normal. Cerebrospinal fluid examination was unremarkable with normal IgG index.

Discussion

As per the diagnostic criteria which define UMN – ALS[1] – evidence of UMN involvement with minor EMG/clinical evidence of LMN involvement within 4 years of onset, this patient fits in to a profile of UMN-ALS. Primary lateral sclerosis (PLS) was the other considered diagnosis. However this requires absence of clinical and EMG evidence of denervation within 4 years of symptom onset. The MRI images in this patient demonstrate bilateral and symmetrical T2/Flair hyperintensities in the regions corresponding to the corticospinal tracts extending from the periventricular white matter, cerebral peduncles and posterior limb of the internal capsule. T1 weighted imaging and DWI did not show any abnormalities. ALS results in motor neuron death and subsequent gliosis extending from the pyramidal cell layer in the cortex to the corticospinal tracts in a retrograde manner. This accounts for the above MRI appearance and is keeping with previous case reports[2-4]. It is postulated that these changes may reflect progression of the disease.

The differential diagnosis for a similar MRI appearance includes ischemia, demyelinating diseases such as multiple sclerosis and vasculitis. However these are rarely symmetrical. Bilateral symmetrical change occurring in normal individuals has also been reported[5]. The clinical and laboratory assessment in the above patient excluded these differential diagnoses.

Even though pathologically evident, patients with ALS rarely demonstrate the above MRI abnormalities and the prevalence ranges from 14-50% of ALS cases[6]. In addition to the above abnormalities patients with ALS also have shrinkage of white matter in the pre central gyrus demonstrable by MRI[7]. This is more prominent in PLS.

The role of MRI as a diagnostic tool in ALS is not clearly defined. EMG is of little value in the setting of predominant UMN involvement as in this case. Therefore MRI may be important in diagnosis of probable UMN dominant ALS with minimal EMG findings to demonstrate localization of the pathology to the corticospinal tracts.

Furthermore the potential of visualization of the MRI changes in the corticospinal tracts as an early marker[8] for the disease and for monitoring disease progression has been explored with the advent of more sensitive advanced MRI techniques such as diffusion tensor imaging (DTI) with fractional anisotropy and functional MRI.

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