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CASE REPORT

Magnetic Resonance Imaging Findings in a case of Amyotrophic Lateral Sclerosis

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Abstract

Amyotrophic lateral sclerosis is a slow, progressive fatal motor neuron disease of both upper motor neuron (UMN) and lower motor neuron (LMN) type. Degeneration of these neurons causes progressive weakness starting from hands and legs spreading towards centrally with a short life span of 3 to 5 years. We report a case of ALS with classical imaging findings.

Keywords: ALS-Amyotrophic Lateral Sclerosis, UMN-upper motor neuron, LMN-lower motor neuron, Garland sign.

Introduction

Amyotrophic lateral sclerosis is a slow, progressive fatal motor neuron disease of both upper motor neuron (UMN) and lower motor neuron (LMN) type. Degeneration of these neurons causes progressive weakness starting from hands and legs spreading towards centrally with a short life span of 3 to 5 years. We report a case of ALS with classical imaging findings.

Case Report

A 50-year-old man was referred to our department from outside for MRI brain examination with history of progressive weakness of both upper and lower limbs. Patient refused admission. Hence, further details could not be made. MRI brain showed symmetrical involvement of bilateral corticospinal tracts as shown in figures 1 and 2.

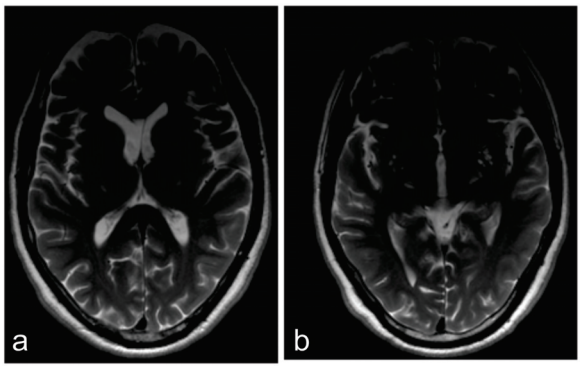

Fig. 1: T2 weighted MRI showing symmetrical bilateral hyperintensities in posterior limb of internal capsule (a) extending to midbrain (b).

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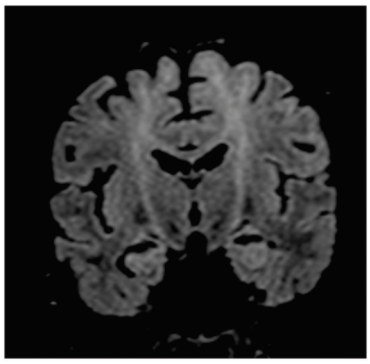

Fig. 2: Oblique coronal T2 FLAIR MRI showing symmetrical bilateral hyperintensities involving both corticospinal tracts from subcortical white matter to brain stem- classic Garland sign.

Discussion

ALS is also known as Charcot disease or Lou Gehrig's disease. Dr. Jean Martin Charcot was the first one to describe the case in 1969 and Lou Gehrig a famous basketball player was the first known person who died with this disease, hence his name for this disease. ALS stands for A- negative, myo-muscle, trophic –nourishment, lateral - spinal cord location and sclerosis-scarring. Two forms of the ALS are seen- sporadic and familial. Motor neurone disease may be primary lateral sclerosis with UMN type and progressive bulbar sclerosis with LMN type. The mean age of onset is 40 to 60 years [1] with slight male preponderance and no race or ethnic predisposition. The clinical diagnosis is followed as per El Escorial criteria – 2015 [4]. The MRI features are [3]; high signal intensity involving symmetrical corticospinal tracts at both T2 - weighted and proton density weighted imaging extending from the corona radiata, through the most caudal aspect of the posterior limb of the internal capsule, into the ventral aspect of the brain stem, and finally into the anterolateral column of the spinal cord thus showing **"Garland Sign"(fig 2), "Wine Glass Sign"** and **"Snake Eyes Sign"** in cervical spinal cord. T2-

weighted MR imaging typically demonstrates low signal intensity in the motor cortex in amyotrophic lateral sclerosis and motor cortex atrophy. MR spectroscopy, diffusion weighted imaging, diffusion tensor imaging help in identifying disease changes earlier. Long TE proton spectroscopy may reveal significantly decreased NA/Cr values consistent with neuronal dysfunction and/or loss.

Differential diagnosis of these MRI findings in ALS are based on anatomy, clinical features of certain pathologies involving brain [Adult polyglucosan body disease], brain stem [Adrenomyeloneuropathy], spinal cord [multiple sclerosis], anterior horn cells pathologies [Kennedy's disease], Peripheral neuropathies and Neuromuscular transmission disorders [myasthenia gravis] and systemic disease [4] [Hyperthyroidism]. Extension of T2 hyperintensity from motor cortex to spinal cord rules out healthy individuals and periventricular pattern of multiple sclerosis. High signal intensity in the corticospinal tract has also been described in Friedreich ataxia and vitamin B_{12} deficiency, where in hyperintensities are only limited to internal capsule.

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