Case 14898

Eurorad

Amyotrophic lateral sclerosis

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ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 35 years, male

Clinical History:

A 35-year-old male patient was referred to our Medicine Department with a history of altered behaviour and intention tremors for six months which progressed to weakness of bilateral upper and lower limbs. An MRI brain was advised.
Imaging Findings:

MRI brain revealed symmetrical T2/FLAIR hyperintensities in the bilateral pre-central gyrus, centrum semiovale, posterior limb of internal capsule and bilateral crus cerebri along the course of corticospinal tracts associated with focal thinning of the body of corpus callosum. Mild hyperintensity was seen in the periventricular region along the occipital horns of the bilateral lateral ventricles. No restriction was seen along the altered signal intensity areas or elsewhere. No blooming was seen on susceptibility-weighted images. The rest of the brain parenchyma was normal in signal intensity. The ventricular system was within normal limits.
Discussion:

ALS is a motor neuron disease causing degeneration of motor neurons in primary motor cortex, brainstem and spinal cord. The annual incidence of this disease is 0.4 to 1.76 per 100, 000 population [1]. It manifests with features of upper and lower motor neuron degeneration characterised by weakness of hands and forearm, spasticity of legs and generalised hyperreflexia. The disease progresses to death of almost 50% patients within 3 years and 90% of them within 6 years of onset of symptoms. Diagnosis is made in patients with signs/symptoms of UMN and LMN degeneration with fast progression. Imaging and laboratory tests are done to rule out other causes of motor neuron degeneration including multiple sclerosis, cerebrovascular disease, compressive cervical myelopathy and conus lesions [2, 3].

MR imaging of the brain shows areas of hyperintensity on T2 weighted/ FLAIR images along bilateral corticospinal tracts (CST) from the centrum semiovale to brainstem, best appreciated on coronal scans [4]. Such hyperintensities have also been observed in patients with X linked Charcot-Marie tooth disease, Krabbe disease and adrenomyeloneuropathy, however, the clinical findings differ from ALS. Apart from corticospinal hyperintensities, low signal intensity of precentral cortex on T2 –weighted images and T2 signal intensity changes in anterior subcortical white matter are also reported [5]. Some studies have also reported mild cerebral atrophy in patients with ALS [6]. Hyperintensities in the anterolateral columns of the cervical cord have been reported in MRI of the cervical spine [7]. Diffusion Tensor Imaging reveals decreased FA and Increased MD values along the CST [8].

In patients with ALS, survival is variable though death occurs due to respiratory failure within 3-5 years after onset of symtoms. MR imaging can help by identifying changes earlier in the course of disease and institution of

neuroprotective treatments.
Differential Diagnosis List: Amyotrophic lateral sclerosis, Primary lateral sclerosis, Metabolic disease involving the corticospinal tracts

Final Diagnosis: Amyotrophic lateral sclerosis

References:

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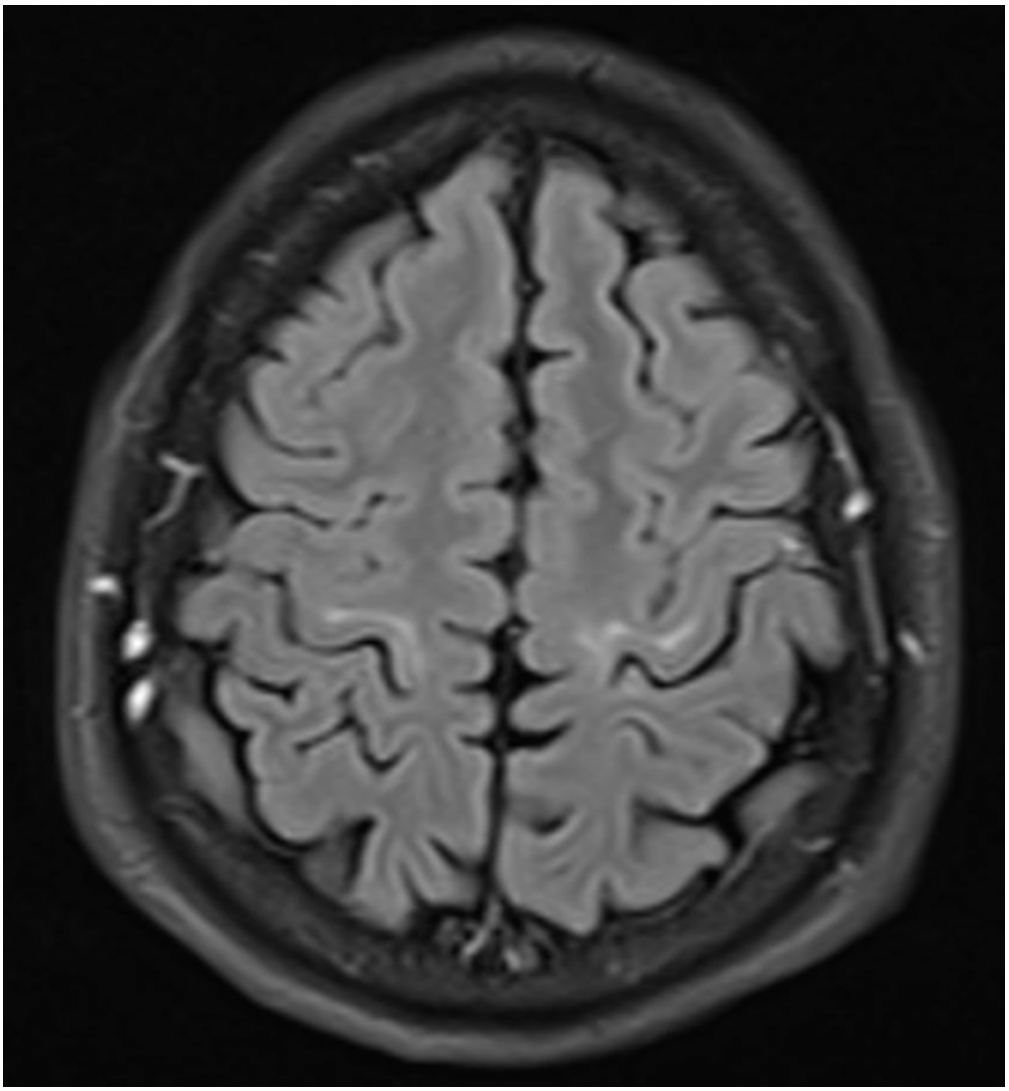
Sage CA, Peeters RR, Gorner A, et al (2007) Quantitative diffusion tensor imaging in amyotrophic lateral sclerosis. Neuroimage 34:486–99 (PMID: 17097892)

Figure 1

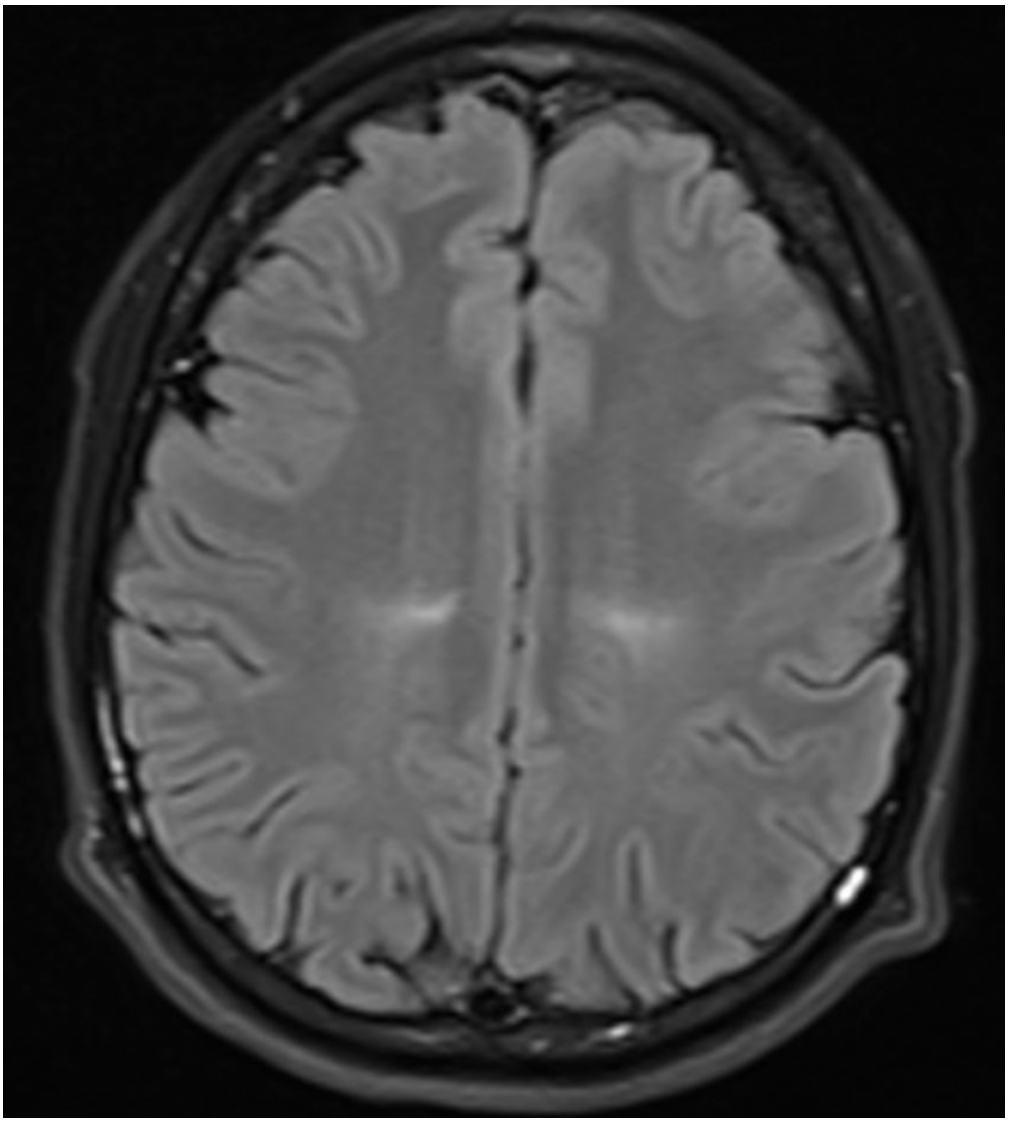
a



Description: MRI brain axial image reveals symmetrical FLAIR hyperintensities in bilateral pre-central gyrus. **Origin:** Department of Radiology, Medanta-The Medicity, Gurgaon,India

Figure 2

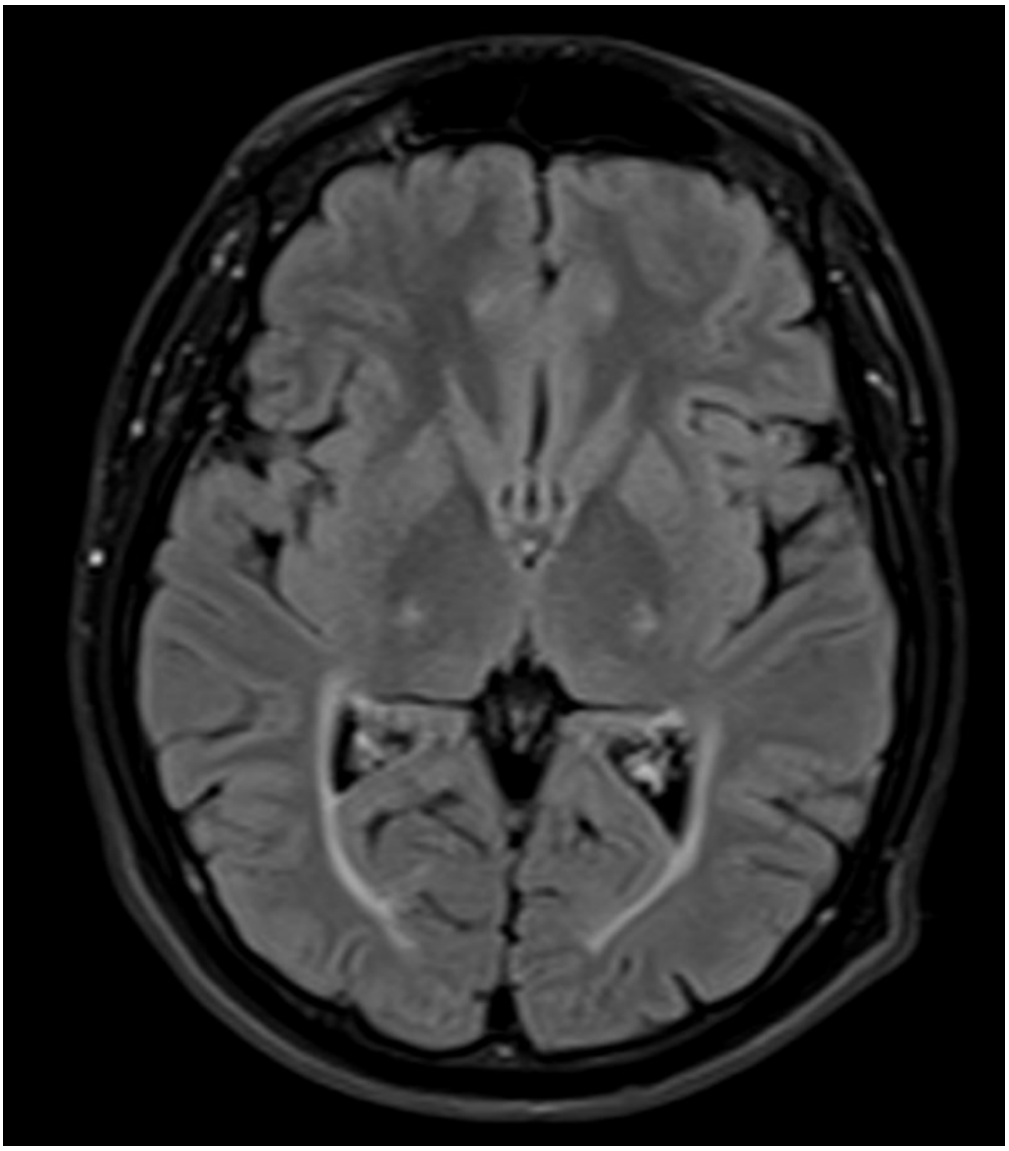
a



Description: MRI brain axial image reveals symmetrical FLAIR hyperintensities in bilateral centrum semiovale. **Origin:** Department of Radiology, Medanta-The Medicity, Gurgaon,India

Figure 3

a



Description: MRI brain axial image reveals symmetrical FLAIR hyperintensities in posterior limb of both internal capsules. **Origin:** Department of Radiology, Medanta-The Medicity, Gurgaon,India

Figure 4

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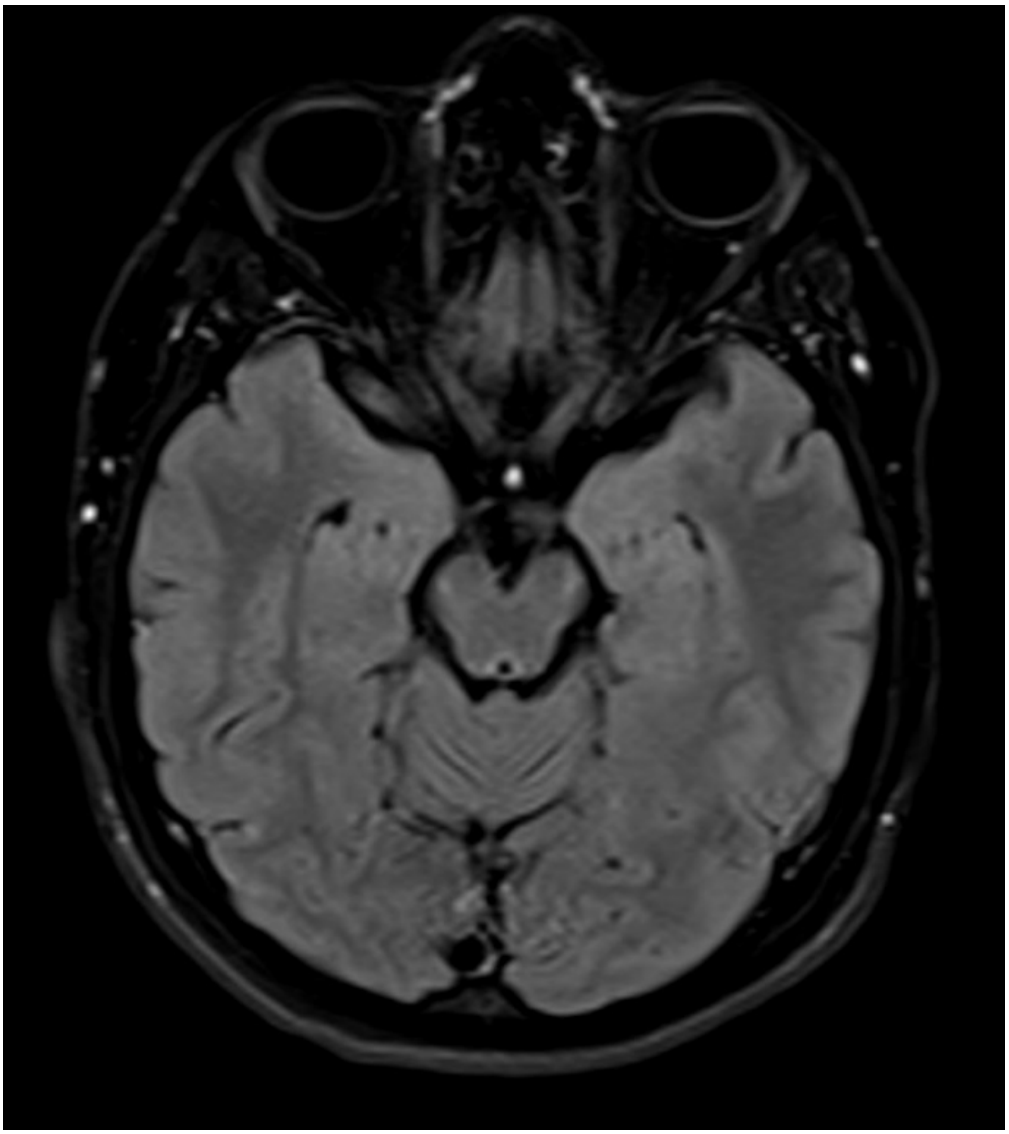

Description: MRI brain axial image reveals symmetrical FLAIR hyperintensities in bilateral crus cerebri.
Origin: Department of Radiology, Medanta-The Medicity, Gurgaon,India

Figure 5

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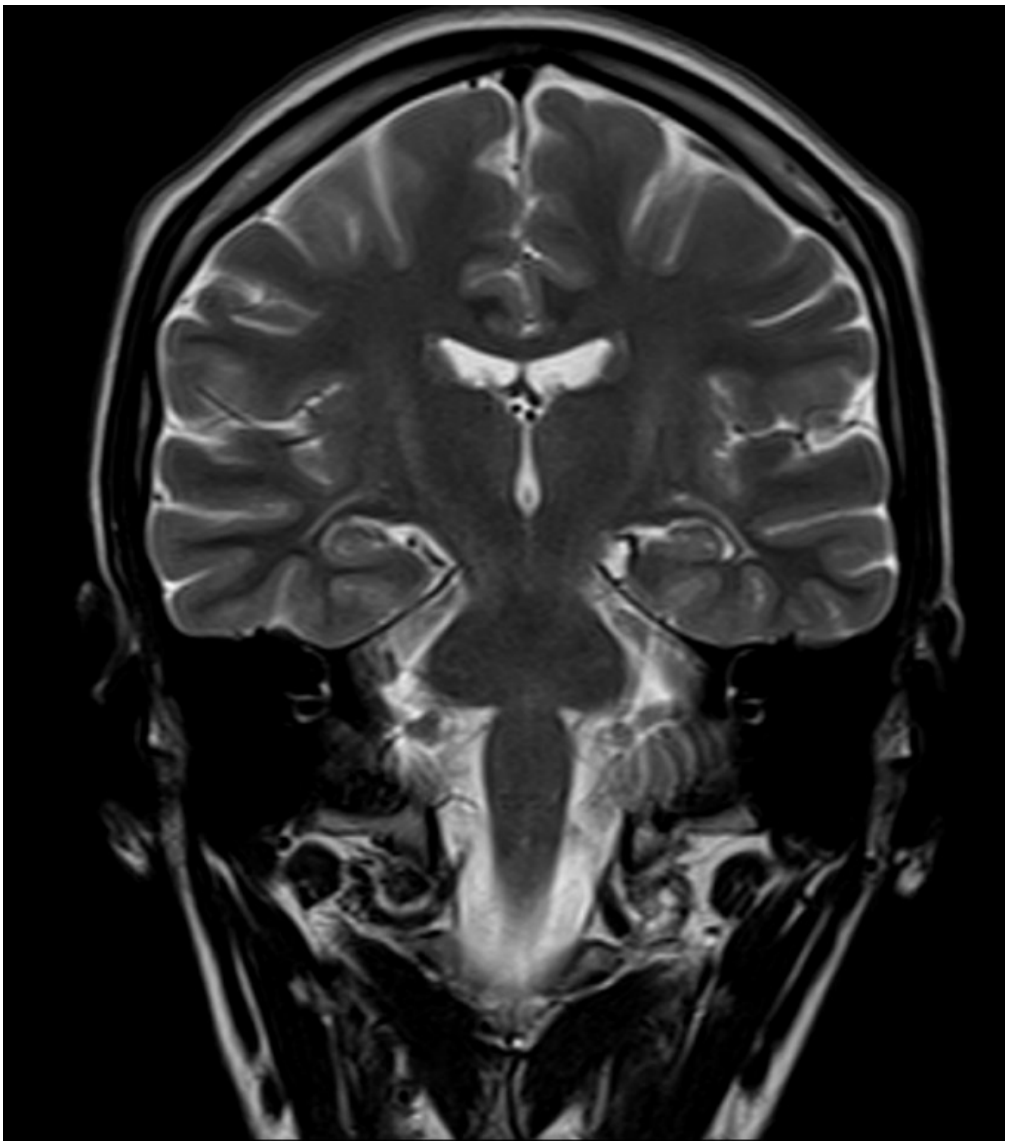

Description: MRI brain coronal T2 image shows hyperintensity in subcortical white matter along the corticospinal tract. **Origin:** Department of Radiology, Medanta-The Medicity, Gurgaon,India