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Diagnostic clue from brain MRI in a 91 year old with Amyotrophic Lateral Sclerosis (ALS)

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Abstract

ALS is a neurodegenerative disorder that involves both upper and lower motor neurons, which has a median survival of three to five years. We report a 91-year-old woman who presented with an 18-month history of progressive weakness. Her speech had become increasingly slower. She denied any urinary or bowel abnormalities or any sensory loss and had grossly normal cognition. Physical examination was remarkable for nasal dysarthria, weak palatal movements and atrophic tongue. She had diffuse muscle atrophy with an increased tone in all 4 extremities. Strength was 1/5 in all extremities. Reflexes were brisk throughout with mute toes. MRI Brain showed T2 hyper intensities of the cortico-spinal tract (CST). In ALS cortical motor cells degenerate leading to retrograde axonal loss and gliosis in the CST, which may appear as T2 hyper-intensities in the MRI. Our patient presented at the terminal stage of her disease, with a rapidly progressing ALS and had symmetric T2 hyper-intensities of CST. This supports the prior studies of hyper-intensities and symmetry being related to the rate of disease progression and severity. The age of presentation in our patient was very unusual for ALS but the MRI findings guided us to the correct work up and helped to predict her disease course.

Keywords

Amyotrophic lateral sclerosis, ALS, MRI, Age, Hyperintensity of cortico-spinal tract

Introduction

ALS is a relentlessly progressive neurodegenerative disorder that involves both upper and lower motor neurons. It causes muscle weakness, disability, and eventually death, with a median survival of three to five years after onset. The incidence of ALS in Europe and North America is between 1.5 and 2.7 per 100,000/year and increases with each decade, especially after age 40 years, peaking at age 74, and decreasing thereafter with only 2% of cases being reported over 85 years.[1,2]

MRI is usually done to exclude other possible diagnoses. Conventional MRI of the brain and spinal cord is usually normal in ALS, although hyper-intensity in the corticospinal tracts and hypo-intensity of the motor cortex on T2-weighted images has been reported. [3,4]We report a rare case of ALS with striking MRI findings in a 91-year-old woman.

Case Presentation

A 91-year-old woman presented with an 18-month history of progressive weakness, which began in the legs and gradually progressed to involve the arms. She had started to walk with a cane 12 months prior to presentation, requiring a walker within 6 months, a wheelchair for 3-4 months and had been bedridden for 3-4 weeks prior to presentation. Her speech had become progressively slower over the last 8 months. She denied any urinary or bowel abnormalities or any sensory loss and had a normal cognition.

Physical examination was remarkable for a harsh spastic and nasal dysarthria; there was mild bifacial weakness. Palatal movements were weak bilaterally. Tongue muscles were noted to be atrophic with slowed lateral movements. Jaw jerk was present. She had diffuse muscle atrophy with an increased tone in all 4 extremities. No fasciculation's were noted. Strength was 1/5 in bilateral upper extremities and 0/5 in bilateral lower extremities. Reflexes were brisk throughout and Babinski's reflex was absent bilaterally. She had a normal sensory exam and cognition was grossly intact. Patient's BUN was 10 mg/dl (normal: 5-31 mg/dl), which was at the lower end of normal consistent with muscle atrophy. Other CBC, CMP results were unremarkable.

MRI Brain with and without contrast (1.5 T) demonstrated increased T2 signal intensity involving the corticospinal tracts bilaterally, extending into the anterior aspects of the midbrain consistent with Wallerian degeneration or ALS as shown in the images (Figure 1-3).

EMG revealed diffuse ongoing denervation and chronic reinnervation with frequent fasciculation potentials in several muscles at the bulbar, cervical, thoracic and lumbosacral level, consistent with the diagnoses of ALS. Patient wanted to go back home to her family with home nursing services. She was discharged home after 3 days of inpatient admission and died 2 months later.

Discussion

ALS is characterized by motor neuron degeneration and death with gliosis replacing lost neurons. Cortical motor cells (Betz cells of the pyramidal tract) degenerate leading to retrograde axonal loss and gliosis in the corticospinal tract. This gliosis results in the bilateral white matter changes sometimes seen in the brain MRI of patients with ALS as T2 hyper-intensity. Findings of pathological examination have demonstrated that these areas of hyper-intensity correspond to areas of degeneration of the pyramidal

tracts. [5] Hyper-intensities of corticospinal tract are seen in about 35% cases of ALS, ranging from mild to rarely being iso-intense to CSF. [6] Studies have shown a positive correlation between rapid progression of disease and positive MRI findings. [6,7] The symmetry of ALS lesions has been thought to reflect the severity of the disease. Clinical correlation remains important as, subtle T2 hyper intensities of cortico-spinal tract have been seen in healthy control patients as well. [8,9]In addition decreased signal intensity in motor cortex on T2 MRI images is supportive of ALS and both findings combined can improve the sensitivity and diagnostic accuracy of MRI for ALS. [4]Our patient presented at the terminal stage of her disease, with a rapidly progressing ALS and had symmetric T2 hyper-intensities of corticospinal tract. This supports the prior studies of hyper-intensity and symmetry being related to rate of disease progression and severity. [6,7]The age of presentation in our patient was exceptionally rare for ALS but the MRI findings guided us to the correct work up and helped predict the progression. Advanced MRI techniques like diffusion tensor imaging; voxel-based morphometry and resting-state functional MRI have a higher sensitivity for assessing disease condition in ALS but their use is debatable considering cost effectiveness.[3,10]

Figures

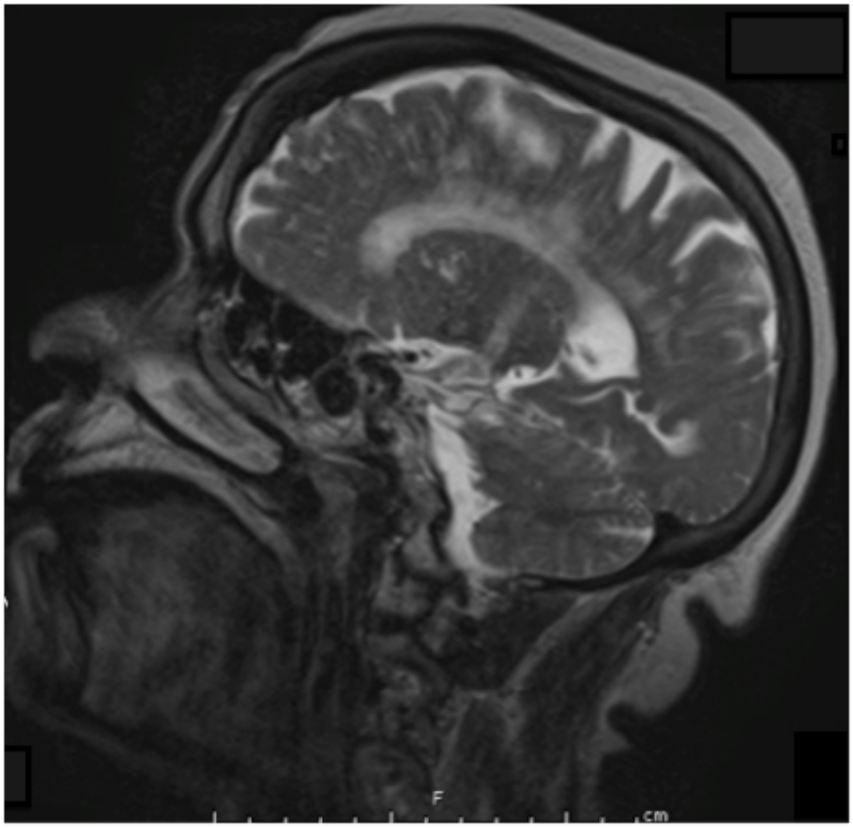

Figure 1: Sagittal T2 showing hyper intensity in the internal capsule along the corticospinal tract

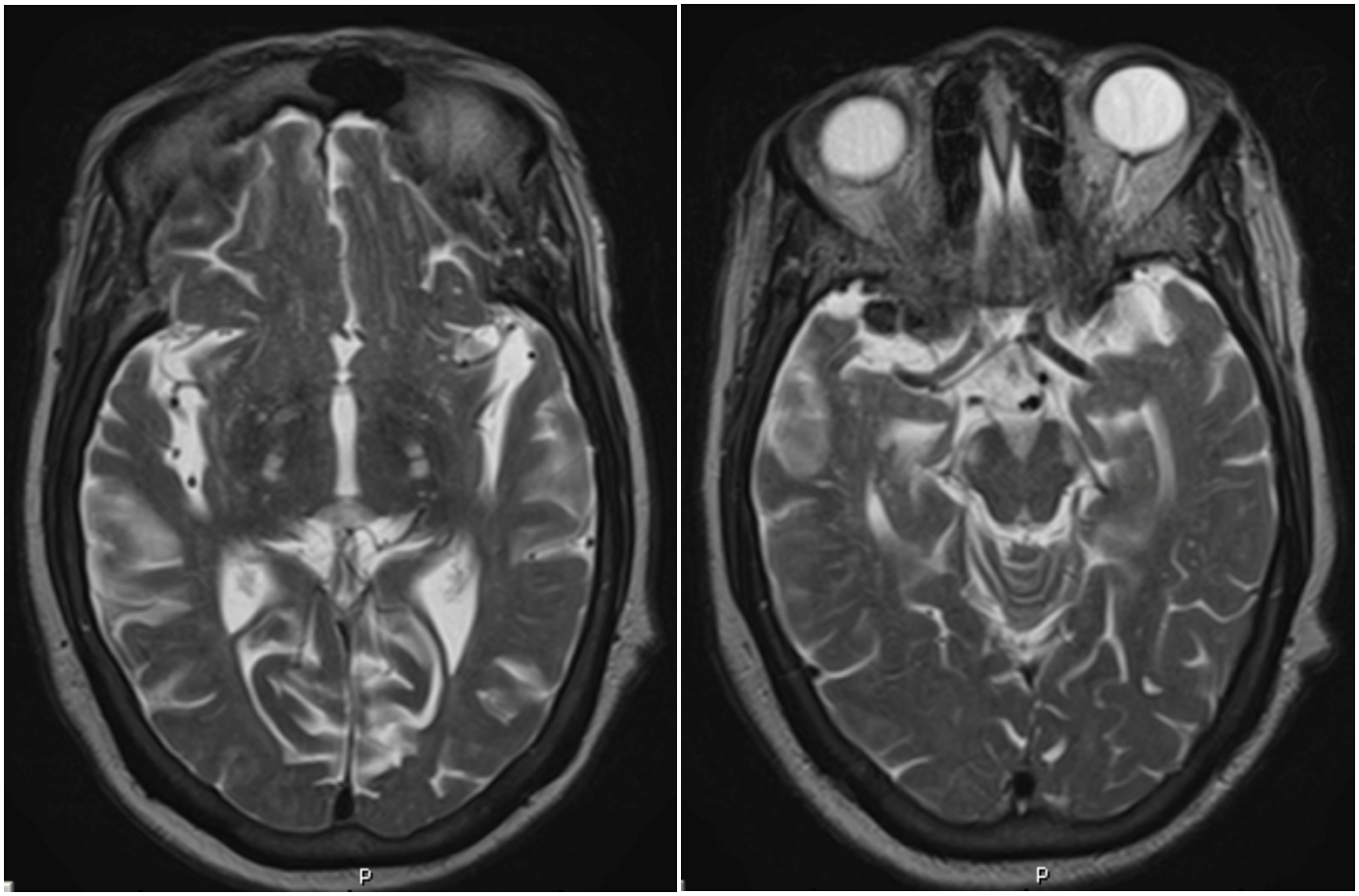

Figure 2,3: Axial T2 images showing hyper intensity in internal capsule and cerebral peduncles along the corticospinal tract

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