'Wine Glass' Sign in a Case of Juvenile Amyotrophic Lateral Sclerosis

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ABSTRACT

Amyotrophic lateral sclerosis is a neurodegenerative disorder affecting upper and lower motor neurons in primary motor cortex, brainstem and spinal cord. We present a case of a 24-year-old young male who presented with progressive quadriparesis and bulbar palsy with MRI brain revealing characteristic hyperintensities of the corticospinal tracts bilaterally, extending from the internal capsule to the brainstem, producing a 'wine glass' appearance on coronal sections.

Keywords: Amyotrophic lateral sclerosis, corticospinal tracts, wine glass

Amyotrophic lateral sclerosis (ALS) is a form of motor neuron disease (MND) characterized by the degeneration of upper and lower motor neurons. The mean age of onset is 57 years. Juvenile ALS is reserved for patients 25 years of age or less and is characterized by a prolonged survival. The degeneration of corticospinal tracts in the brain leads to the development of hyperintensities along the tracts extending from internal capsule to brainstem, producing a 'wine glass' appearance on magnetic resonance imaging (MRI). Thus, MRI brain can be a modality to provide an early evidence of corticospinal tract degeneration in MNDs.

CASE REPORT

A 24-year-old male presented with history of subacute onset weakness of all four limbs for last 1 year starting with left lower limb, manifesting as difficulty in clearing off foot from ground. This was followed by a similar involvement of right foot after 4-5 months. There was difficulty in gripping objects along with thinning of muscles and guttering noted between the thumb and the first dorsal interosseous. The patient started experiencing difficulty in swallowing, nasal regurgitation of fluids along with nasal twang to his voice. There was no associated sensory complaint, bowel or bladder involvement.

On examination, the vitals were normal. On neurological examination, jaw jerk was brisk. There was wasting of posterior fibers of deltoid, both anterior compartments of forearms bilaterally, interossei, chiefly the first dorsal interosseous and calf muscles. Generalized spasticity was present. The muscle power was MRC Grade 3/5 and 4/5 in the right and left upper limbs, respectively, and 3/5 in lower limbs bilaterally. Deep tendon reflexes were exaggerated and bilateral Babinski's sign was present. Sensory and cerebellar examination was unremarkable. The patient was subjected to neurophysiological studies, with electromyography revealing neurogenic affection with multiple fasciculations. MRI of the brain revealed linear, bilaterally symmetrical hyperintensities (Figs. 1-3) involving the corticospinal tracts in internal capsule, crus cerebri and pons on T2 weighted image (T2WI), giving a 'wine glass' appearance, seen in the coronal plane (Fig. 4).

DISCUSSION

Amyotrophic lateral sclerosis is a neurodegenerative disease characterized by involvement of both upper and lower motor neurons and is diagnosed by using revised El Escorial criteria. Thus, ALS has been conventionally diagnosed on the basis of clinical and electromyographic data. Motor neurons undergo degeneration and result in axonal edema apparent on electron microscopy. The degeneration of motor neurons may result in cellular loss and axonal edema.

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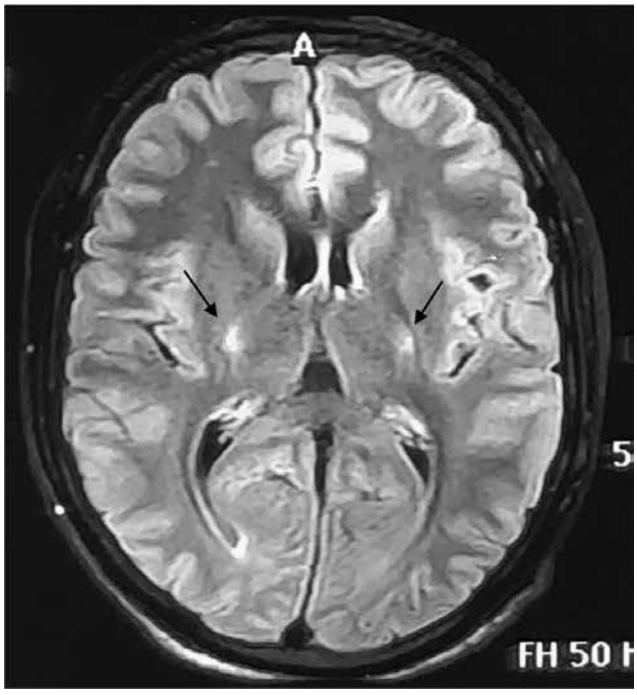

Figure 1. MRI brain axial section showing bilaterally symmetrical hyperintensities involving the corticospinal tracts in internal capsule.

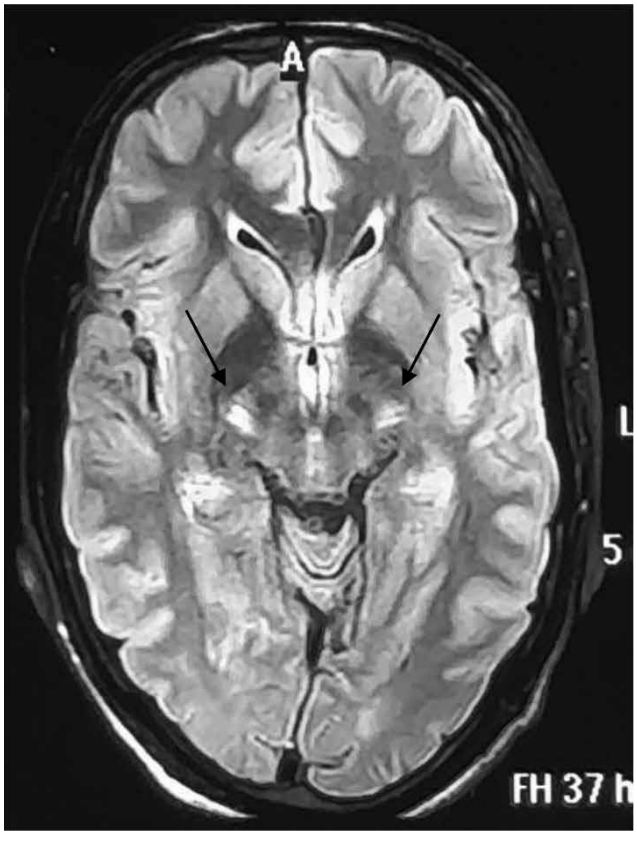

Figure 2. MRI brain axial section showing bilaterally symmetrical hyperintensities involving the corticospinal tracts in crus cerebri.

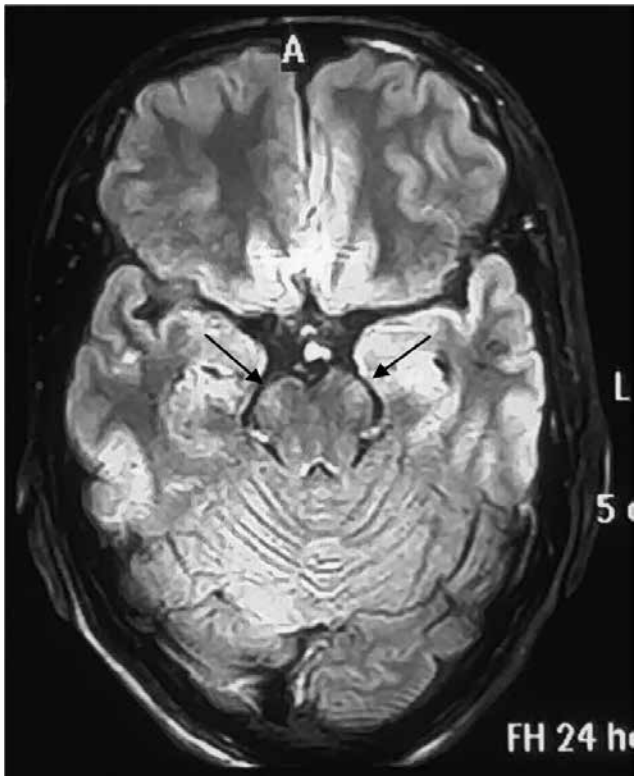

Figure 3. MRI brain axial section showing bilaterally symmetrical hyperintensities involving the corticospinal tracts in pons.

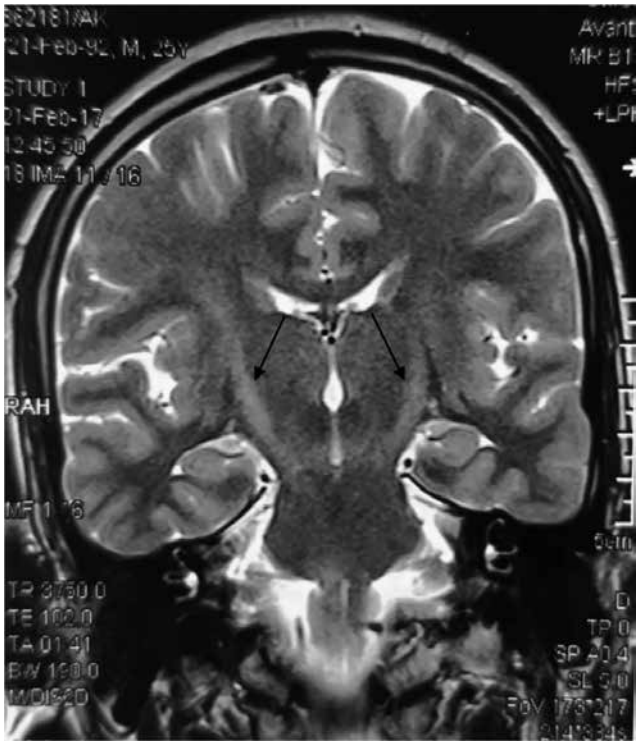

Figure 4. MRI brain coronal section showing bilaterally symmetrical hyperintensities involving the corticospinal tracts giving a 'wine glass' appearance.

About 90% of ALS cases are sporadic and 10% familial, with majority of the cases of juvenile ALS being familial. Juvenile ALS is inherited as autosomal recessive in majority, mapped to chromosome regions 2q33 and 15q12-21. Some are autosomal dominant, mapped to chromosome 9q34. Neuroimaging, till date, has been of limited use in diagnosing juvenile ALS. Kumar et al, in their case report, found the typical 'wine glass' appearance in a 9-year-old male presenting with both upper motor neuron (UMN) and lower motor neuron (LMN) features. Midani et al showed bilateral hyperintensities along the corticospinal tracts on T2WI MRI in a patient with juvenile ALS. There have been several studies on adult patients of MND showing MRI changes. But juvenile ALS, being a rare entity, doesn't have much literature on its MRI changes. In this case report, we suggest that the involvement of corticospinal tracts results in a typical 'wine glass' pattern in coronal section of brain MRI and thus, has a diagnostic utility in these cases.

CONCLUSION

Juvenile ALS can be suspected in children with both UMN and LMN features. MRI brain can provide an early clue to the diagnosis and prognostication of this disease.

SUGGESTED READING

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