Pictorial CME

Foot Drop as the Initial Presentation of Amyotrophic Lateral Sclerosis

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Pseudo polyneuritic ALS mimics motor neuropathy as it is characterized by unilateral foot drop which progresses as the disease advances. In this report we are discussing a case that presented with foot drop. Detailed neurological examination,supported by electrophysiological findings helped us to arrive at the diagnosis of this unique variant of ALS. Pseudopolyneuritic form has a slow progression and better prognosis when compared to other ALS subtypes. Recognition of this form of ALS is important for clinicians because the combination of distal weakness of the lower limb and absence of Ankle jerk usually suggests peripheral neuropathy.

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Amyotrophic lateral sclerosis ALS is defined as adult onset idiopathic progressive degeneration of anterior horn cells and pyramidal tract resulting in progressive muscle weakness, wasting and fasciculations. Patients with pseudo polyneuritic variant of ALS presents with distal weakness and foot drop as the initial manifestation. Hereby we report this case of ALS with foot drop as initial manifestation.

Case Report

58 year old male presented with subacute onset, progressive flaccid weakness of left lower limb, distal more than proximal, associated with thinning of left foot since 1 year. It was followed by weakness of right lower limb after 1month. Patient also reported muscle twitching over both thighs. He developed weakness of both upper limbs, left followed by right over past 2months, distal more than proximal, flaccid type associated with thinning and twitching. Higher mental functions were normal. Cranial nerve examination revealed exaggerated gag reflex .Tongue showed atrophy, weakness and fasciculations. Motor examination revealed flaccid weakness of both hands and feet with MRC grade of 4/5 proximally and 1/5 distally associated with wasting and fasciculation. Patient had high steppage gait with bilateral foot drop, right more than left. All deep tendon reflexes were exaggerated except bilateral ankle jerk which were absent. Jaw jerk was brisk with prominent cortical release reflexes. Sensory examination was normal. MRI brain and spine was normal. Nerve conduction study showed severe motor axonal neuropathy with normal sensory action potentials. Laboratory investigations including Complete blood count, Liver function test , Renal function test, and Thyroid profile were within normal limits. Serum electrophoresis, Urine Bence Jones Proteins and Paraneoplastic work up were negative. Electromyography showed fasciculation and fibrillation potentials with giant polyphasic motor unit action potentials with increased duration and incomplete interference suggestive of neurogenic pattern. Hence diagnosis of Amyotrophic Lateral Sclerosis was made as per modified El escorial criteria. Patient was initiated on Riluzole and counselled regarding prognosis.

Fig 1 — Patient in sitting position demonstrating bilateral foot dorsiflexion weakness with right foot drop

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Discussion

Amyotrophic lateral sclerosis (ALS) was first described in 1869. It is frequently referred to as "Lou Gehrig's disease" in memory of the famous baseball player who died of ALS in 1941. ALS is defined as adult-onset, idiopathic, progressive degeneration of anterior horn cells and upper motor neurons resulting in progressive muscle weakness, wasting and fasciculations. The clinical picture varies, depending on the location and progression of the pathologic changes. Diagnostic criteria of the World Federation of Neurology (The "El Escorial criteria") can help define and classify ALS.

The pseudopolyneuritic form of ALS is a subtype of ALS characterised by distal weakness of unilateral lower limb and absence of ankle jerk at disease onset[1]. Pathology of this form of ALS is associated with myelinated fiber loss in the corticospinal tract of the thoracic and lumbar spinal cord segments[2].

The pseudo polyneuritic form of ALS may begin with foot drop in some patients. This form may be confused with some clinical conditions such as lumbar plexopathy, L5 radiculopathy, peroneal neuropathy and distal myopathy because of the presence of isolated foot drop at presentation. This form of ALS has better prognosis and survival rates than the other subtypes of ALS[2].

Conclusion

This case is reported here to emphasise that Pseudo polyneuritic form of ALS can present with foot drop. In this aspect, it may be misdiagnosed as neuropathy. Therefore, a patient's history, neurologic examination and electrophysiologic evaluation should be assessed carefully for a proper diagnosis.

References

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