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Research Article

A RARE CASE REPORT ON HIRAYAMA DISEASE

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ABSTRACT

Hirayama's disease also referred to as monomelic amyotrophy or Sobue disease or juvenile non-progressive amyotrophy, predominantly impacts young males, especially in regions such as India and Japan. It presents as a specific type of focal, lower motor neuron disorder, with its precise cause often elusive.

INTRODUCTION

Hirayama disease presents with a gradual onset of muscle atrophy, usually unilateral or asymmetric and oblique, affecting the myotomes of C7, C8, and T1. Its initiation coincides with the adolescent growth spurt. This condition is characterized by progressive muscle dystrophy in the distal upper limbs, particularly associated with flexion movements of the cervical spine. Diagnosis typically involves cervical spine MRI, especially when performed with the spine in a flexed position.

CASE REPORT

A 19 year old male presented with symptoms of progressive weakness of left upper limb since 2 months followed by right upper limb weakness which is gradually progressive. On examination, there was atrophy of thenar and hypothenar muscles on the left side.

There was no significant previous history of trauma or any chronic illness.

Imaging

T2 weighted MRI of cervical spine saggital section on neutral position shows no significant abnormality within the spinal cord. **Fig.1**

T2 weighted MRI of cervical spine saggital section on flexion position shows anterior translation of posterior dura with dorsal epidural soft tissue extending from C4 to D3 vertebral body **Fig.2** level (arrow) which is causing mass effect over spinal cord in the form of severe compression with abnormal hyperintensity within the cord extending from inferior end plate of C4 to superior end plate of C7.

The epidural soft tissue is having heterogeneous intermediate signal with multiple hypointense engorged epidural venous plexus.

On post contrast T1 weighted saggital image on flexion position, there is homogeneous enhancement of the epidural soft tissue with relatively less enhancement of engorged venous plexus. (Arrow) **Fig.3**

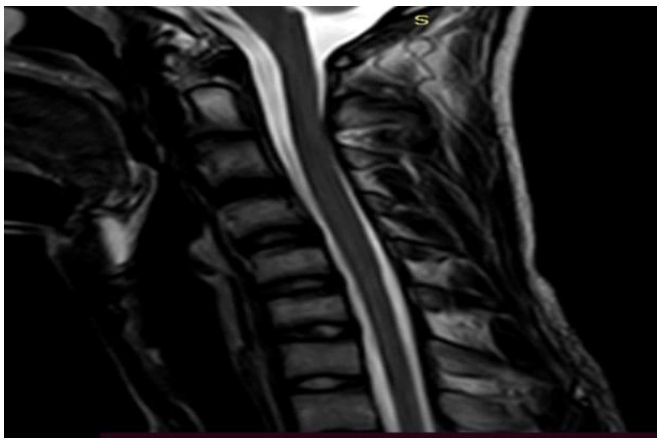

Fig.1

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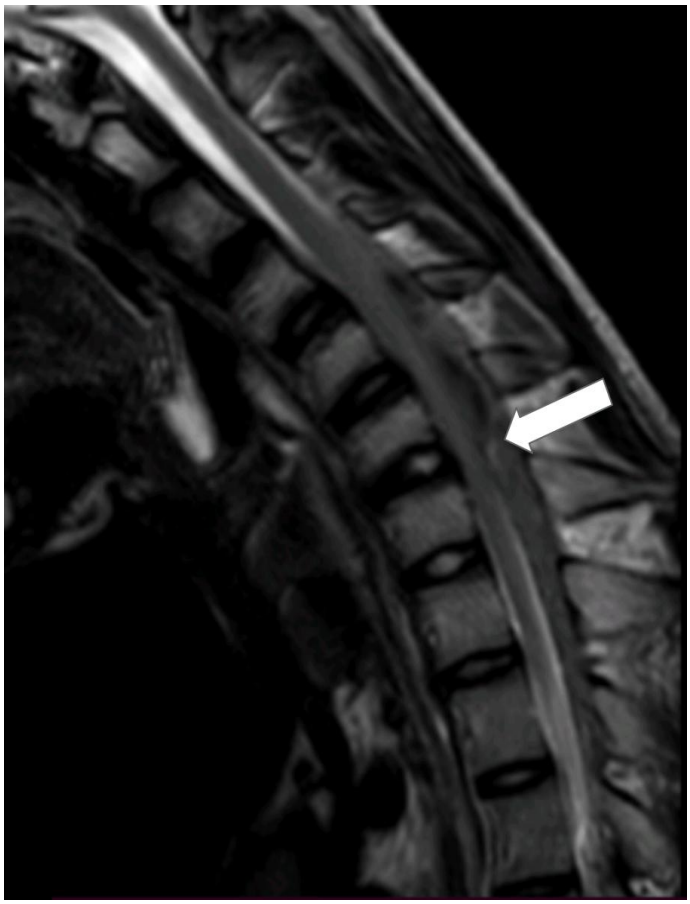


Fig.2

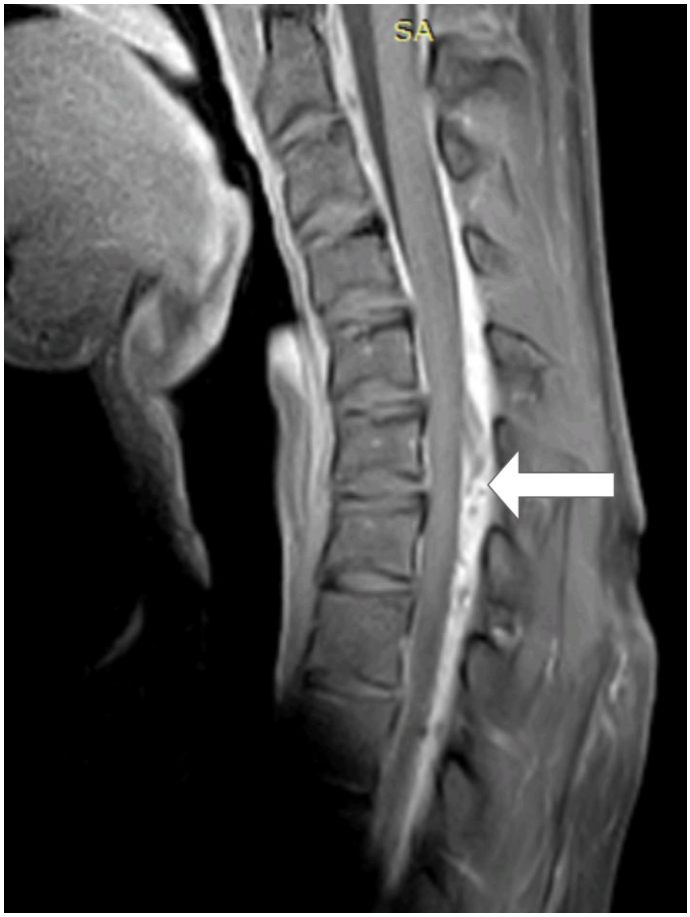
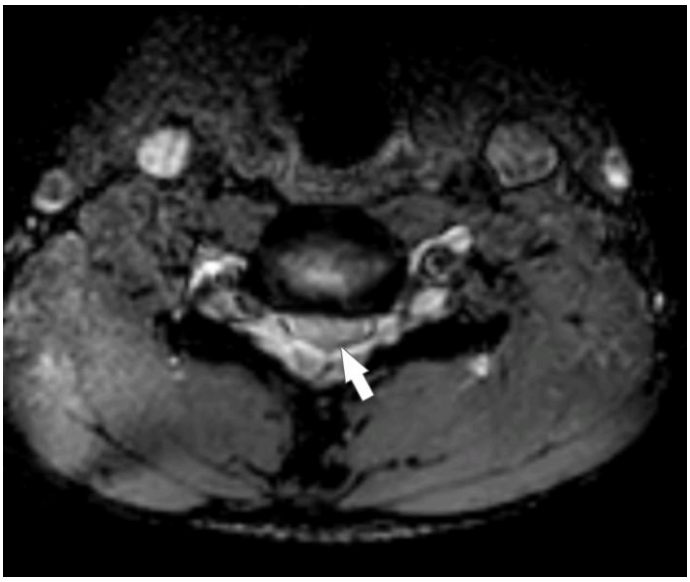


Fig.3



GRE axial section cervical spine shows crescent shaped hyperintensity (arrow) in the dorsal epidural compartment with few areas of bloomings.

All the imaging findings are suggestive of **Hirayama disease.**

DISCUSSION

Hirayama disease typically begins with a gradual onset of asymmetrical weakness and muscle wasting in the upper limbs, predominantly affecting the C7, C8, and T1 myotomes. It's more common in males aged 15 to 25. The condition usually progresses over one to three years before stabilizing, resulting in a relatively mild course. Clinical features may include irregular coarse tremors in the fingers, worsened temporarily by exposure to cold. Sensory, reflex, and cranial nerve examinations typically produce normal results, while involvement of the pyramidal tract, autonomic system, and cerebellum is rare. Electromyography often reveals chronic denervation in affected muscles, sometimes with acute denervation changes. Moreover, even muscles that appear healthy may exhibit abnormal electromyography findings.

The rarity of Hirayama disease and its numerous atypical presentations pose a diagnostic challenge. The diagnostic criteria encompass primary weakness and atrophy in the distal forearm and hand muscles, predominantly unilateral upper extremity involvement, onset typically between ages 10 and the early 20s, gradual symptom onset with progressive evolution over several years followed by stabilization, absence of lower extremity involvement, lack of sensory disturbances or abnormal tendon reflexes, and exclusion of other conditions such as motor neuron disease, multifocal motor neuropathy, brachial plexopathy, spinal cord tumors, syringomyelia, cervical vertebral abnormalities, anterior interosseous, or deep ulnar neuropathy.

The exact pathogenesis of HD remains uncertain. Pathological studies have revealed features like cell shrinkage and necrosis, varying degrees of degeneration of small and large nerve cells, mild gliosis, and some circulatory insufficiency in the anterior horns of the spinal cord from the lower cervical to upper thoracic levels, particularly at the C7 and C8 levels. Some authors have proposed atopy and elevated serum IgE levels as possible precipitating factors. The widely accepted hypothesis suggests cervical myelopathy associated with neck flexion, leading to spinal cord compression against the vertebral body.

In individuals with HD, the relatively short and tight dura mater fails to compensate for the increased length of the vertebral canal during neck flexion, resulting in spinal cord compression. Repeated neck flexion episodes cause multiple ischemic events and chronic trauma to the spinal cord, ultimately leading to myelopathy, characterized by asymmetric lower cervical cord thinning on MRI.

The differential diagnosis of HD includes other conditions such as the distal form of spinal muscular atrophy, amyotrophic lateral sclerosis (ALS), post-polio syndrome, multifocal motor neuropathy with conduction block, toxic neuropathy, and structural cervical cord lesions (e.g., syringomyelia). Diagnosis relies on typical clinical features and dynamic MRI studies during neck flexion, revealing characteristic findings such as anterior displacement of the posterior wall and a well-enhanced crescent-shaped lesion in the posterior epidural space of the lower cervical canal.

CONCLUSION

Hirayama disease is a self-limiting disorder, and there is no consensus on definitive treatment. However, early diagnosis is crucial because a cervical collar may halt the progression of the disorder by limiting neck flexion. Physiotherapy is also beneficial in preventing complications resulting from immobility, such as joint stiffness and muscle wasting.

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