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Case Report

Hirayama Disease

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Abstract
Hirayama diseaseis a relatively rare myelopathy, a self limiting juvenile spinal muscular atrophy of the distal upper limbs. The exact etiology is unknown however a limited form of ischemic cervical myelopathy has been proposed as the cause. It affects mostly males in second to third decade. Cervical MRI is the preferred investigation. We report a case of Hirayama disease in a 24 year Indian male presenting with gradually progressive weakness and wasting of both the hands.

Case Report

A 24yr old male patient presented with history of slowly progressive weakness and wasting of the right hand since 4years, which progressed over 3 years to involve the left hand as well. The weakness was limited to distal muscles which interfered with his several daily activities He had no difficulty in lifting hands above the shoulder. Patient had no history of pain, loss of sensation, dysphagia, and neck pain. No history of trauma, febrile illness, or exposure to any toxins or heavy metals with no history of similar complaints in the family.

On examination, he was conscious and oriented and his vitals were normal. The examination of motor system showed atrophy of small muscles of the hand bilaterally. Full abduction, adduction of the digits, opposition of thumbs and palmar grasps and flexion at wrists were impaired. Deep tendon reflexes of triceps was diminished and biceps and supinators normal bilaterally. Examination of lower limbs was normal. There was no abnormality of Cranial nerve, sensory system, Co odination and gait. Blood investigations of Complete blood count, renal function tests, serum electrolytes, urine routine were within normal limits.

MRI Cervical spine with contrast showed Focal cord atrophy with myelomalacic changes noted at C5 and C6 level. With flexion, there was dorsal dural detachment on T2 images with extensive posterior epidural enhancement extending from C3 to D2 levels with anterior displacement of cervical cord. Underlying vertebrae appears normal and no features suggestive of craniovertebral anomalies. Nerve Conduction studies showed Bilateral ulnar nerve axonopathy. Based on the diagnosis of clinical features and findings on MRI diagnosis of Hirayama disease was made. Patient was put on hard cervical collar and asked to follow up at regular intervals.

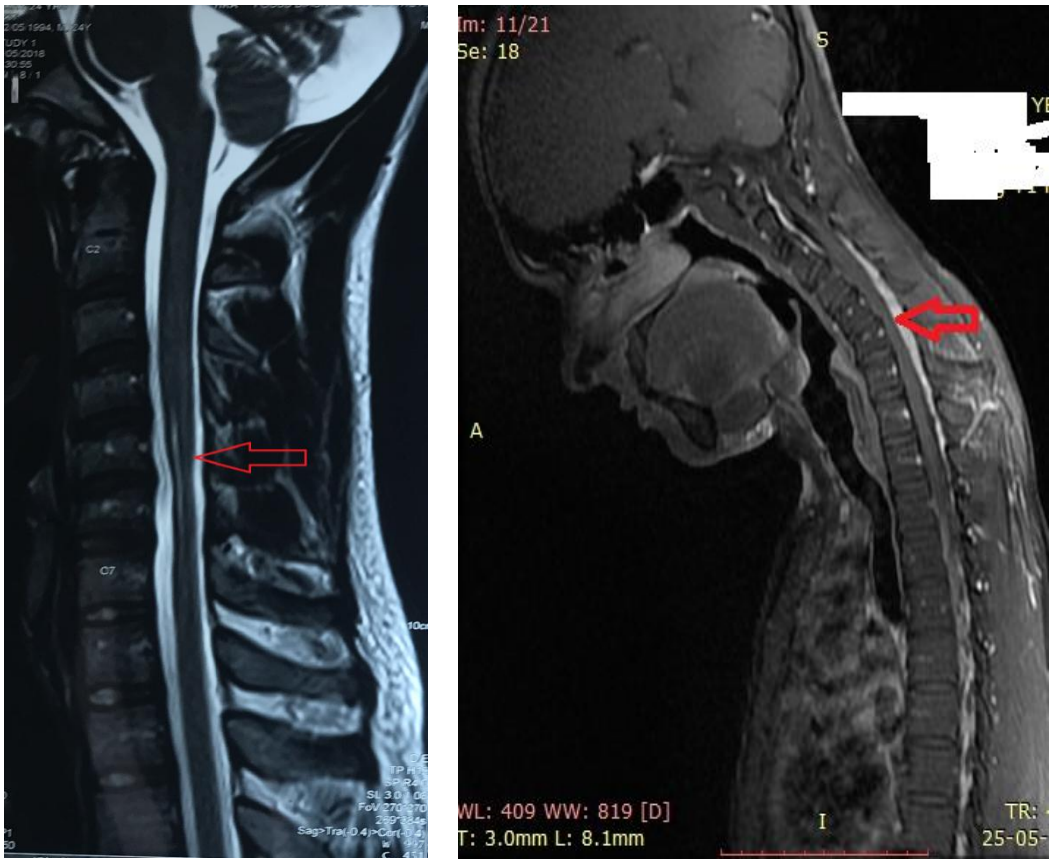

Image 1: In neutral position showing cord thinning and on flexion showing dorsal dural detachment on T2 with posterior epidural enhancement from C3 to D2.

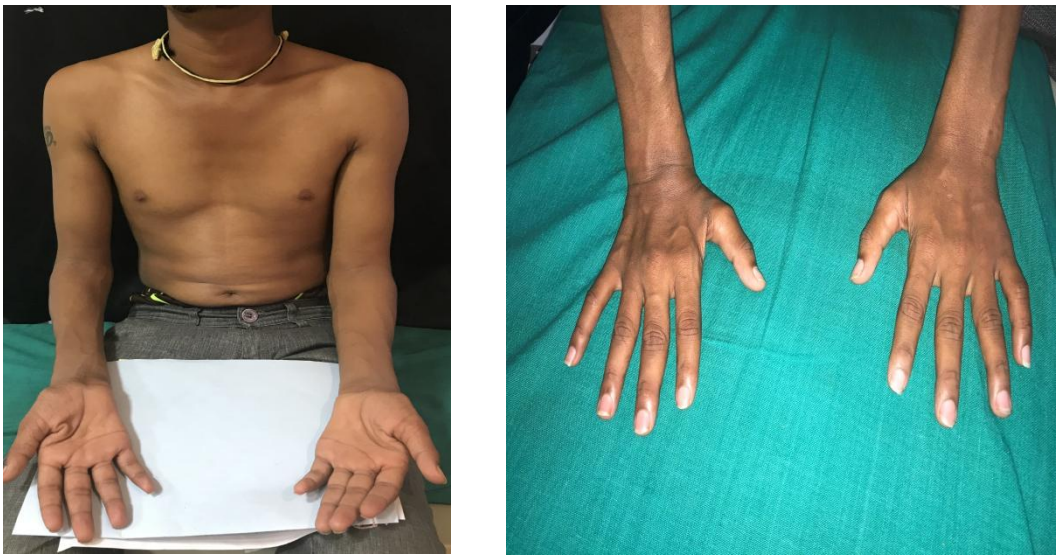

Image 2: showing atrophy of the small muscles of the hand.

Discussion

Hirayama disease is a relatively rare myelopathy, a self limiting juvenile spinal muscular atrophy of the distal upper limbs. The disease usually begins in the late teens, more than 60% of patients are men. Originally described in Indian and Japanese patients, the disorder is now recognizable around the world. The most common presentation is one of an idiopathic, slowly progressive, painless weakness and atrophy in one hand or forearm. The arm is the affected limb in approximately 75% of the patients and the leg in the remaining 25% (benign calf amyotrophy). Spread may occur to the contralateral limb in about 20% of cases and

rare patients later develop an ALS-like disease. The most common pattern is unilateral atrophy of C7–T1 innervated muscles, with sparing of the brachioradialis (the "oblique atrophy" pattern). Muscle stretch reflexes are invariably hypoactive or absent in the muscles innervated by the involved cord segment but are normal elsewhere. UMN signs are not present. The cranial nerves, pyramidal tracts, and the autonomic nervous system are normal. Weakness and atrophy may progress steadily for the initial 2 to 3 years, but most patients have stabilized within 5 years. The etiology being unknown, neuroradiological studies shows a mechanically induced limited form of ischemic cervical myelopathy, being the result of local compression of the dura and spinal cord against vertebrae during repeated neck flexion /extension, in turn due to disproportionate growth between the contents of the dural sac and the vertebral column. Motor nerve conduction studies are either normal or reveal only reduction in the maximum CMAPs; Routine laboratory test results are normal. Cervical MRI reveals segmental spinal cord atrophy or occasionally an area of increased signal on T2-weighted scans of the cervical spinal cord enlargement.Early diagnosis and therapeutic intervention may minimize the functional disability of young patients.

Therapeutic intervention using a cervical collar to minimize neck flexion halts the progression of the disease.

Acknowledgement

The patient consent was received for this case report to be published.

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