Hirayama Disease: A clinical-radiological review

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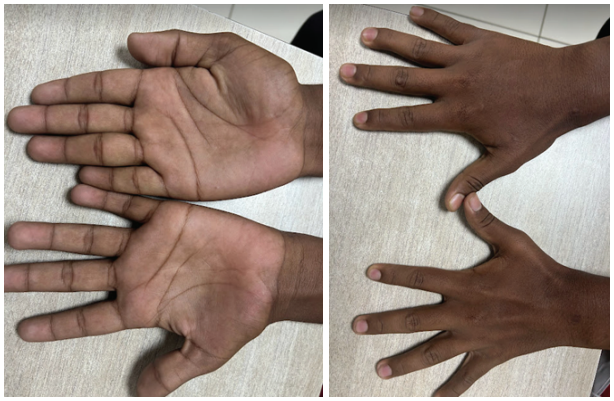
Background

Hirayama disease, also known as monomelic amyotrophic, is a rare neurological entity. The disease is characterized by progressive muscle wasting in the distribution of nerve roots C6, C7 and T1. It is believed to be a result of forward displacement of the dural sac and spinal cord induced by neck flexion. We present the case of a 16-year-old boy with progressive weakness of left upper limb.

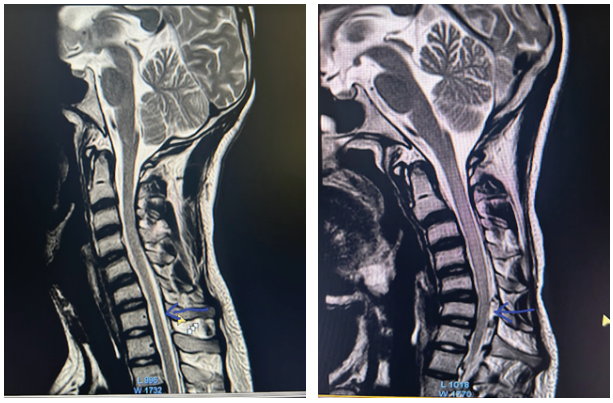
Case Presentation

A 16-year-old boy with no relevant past history presented with a history of weakness and tremors in the left upper limb. He had difficulty in gripping objects, buttoning and unbuttoning of shirt. There were no sensory complaints like paresthesia, pain, or numbness. There was no other neuro axial involvement.

Neurological examination showed an alert, conscious and oriented boy with normal higher mental functions and cranial nerves. He had jerky tremors in the fingers of the left upper limb suggestive of polyminimyoclonus. There was a wasting of hypothenar, thenar interosseous muscles and forearm muscles. Sensory examination was normal. Deep tendon reflexes were normal.



Nerve conduction studies showed reduced CMAP in left ulnar nerve when recorded from Abductor Digiti Minimi. MRI Cervical scan was done in neutral positioning. MRI C spine showed thinning of Cervical cord at C5, C6 level. Flexion neck MRI showed anterior displacement of posterior dura, and enlargement of posterior epidural space.



The patient received the diagnosis of "Hirayama disease". The patient was treated with a cervical collar to prevent neck flexion and was advised regular follow-up.

Discussion

In Hirayama disease patients, there is an imbalance in the growth of the vertebra and dura-mater leading to a "tight dural canal and overstretched cord," which cannot compensate for the increased length of the posterior wall during flexion. This causes an anterior shifting of the posterior dural wall with consequent compression of the cord. This compression causes chronic microcirculatory disturbances in the anterior portion of the spinal cord leading to necrosis of anterior horns.
Close differential diagnoses are compressive myelopathies of the cervical spine, Syringomyelia, Motor Neuron Disease, and multi-focal motor neuropathy with conduction block.

Majority of patients can be managed conservatively as the disease stabilises. In some patients, with disease progression, surgical intervention has been proposed. Surgery should be limited to severe cases that have progressed rapidly. Cervical decompression with duroplasty has shown good results.

Conclusion

Hirayama disease is a rare neurological entity. It should be suspected in male patients presenting with asymmetric distal lower motor neuron weakness of hands and forearms. It is a benign entity and can be managed conservatively in the majority of patients.

Surgery in rapidly progressive diseases is recommended.

References

[1] Gourie-Devi M. Chapter 11 Monomelic amyotrophy of upper or lower limbs. Handb Clin Neurol. 2007;82:207-27.

[2] Pal Pramod K, Atchayaram Nalini, Goel Gaurav. Central Motor Conduction in brachial monomelic amyotrophy. Neurol India. 2008;56:438–443.

[3] Hirayama K, Tokumaru Y. Cervical dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. Neurology. 2000;54:1922–1926.

[4] Hirayama K. Non-progressive juvenile spinal muscular atrophy of the distal upper limb (Hirayama's disease) In: De Jong JMBV, editor. Handbook of Clinical Neurology. Vol. 15. Amsterdam, the Netherlands: Elsevier; 1991. pp. 107–120.