Case Report

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Hirayama Disease: A Rare Cause of Posture Related Cervical Myelopathy

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Abstract

Hirayama's disease is a rare focal motor neuron disease that is reported mainly in young Asian males (15-25 years old). Hirayama disease also known as juvenile non-progressive cervical myotrophy or monomelic amyotrophy. Insidious onset and slow progression of weakness followed by a spontaneous arrest within several years are classical symptoms described with disease. MRI cervical spine in flexion is mainstay for diagnosis.

Keywords: Hirayama Disease (Hd); Cervical Myelopathy; Juvenile Non-Progressive Cervical Myotrophy.

Introduction

Hirayama's disease is a rare focal motor neuron disease that is reported mainly in young Asian males (15-25 years old). Hirayama disease also known as juvenile non-progressive cervical myotrophy or monomelic amyotrophy. The first case was reported by Hirayama in 1959.

Insidious onset and slow progression of weakness followed by a spontaneous arrest within several years are classical symptoms described with disease. Its aetiology is not clearly defined however role of dynamic chronic compression of cervical spinal cord was proposed.

Magnetic resonance imaging (MRI) of the cervical spine is main diagnostic modality which shows forward displacement of the posterior wall of the lower cervical dural canal leading to marked, often asymmetric, flattening of the lower cervical cord. Surgical decompression and physiotherapy are proposed treatment options [1-3].

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Case Report

A 20-year young male presented with two years' history of weakness and tremor in right hand. Slow progression of weakness was noted which appear to be accelerated for a period of four months. Mild tremors were also started in due course. Tremors were exaggerated on holding objects or fine movements. For last two months aggravated weakness of the right hand was noted on neck flexion. No history of sensory or bulbar symptoms, accident or other preceding illnesses was elicited. There was no medical or family history of similar complaints.

On examination, atrophic changes in the thenar, hypothenar, and interosseous muscles of the right hand were seen except the brachioradialis. Irregular, nonsynchronous fine tremors were noted in the right hand. The deep tendon reflexes, pin-prick sensation, vibration, and joint position were intact. No extra pyramidal signs, Horner sign, or abnormalities in sweating and urination were noted. Electromyography showed high amplitude polyphasic waves and fibrillation in the thenar and hypothenar muscles suggestive of an active denervation change however motor nerve conduction velocities of the median and ulnar nerves were within normal limits. These findings were compatible with an anterior horn cell disorder involving the C7–8 and T-1 levels of the cord.

On MRI of cervical spine focal thinning of spinal cord was seen without obvious intensity

changes(Figure 1). Asymmetric flattening of spinal cord with reduced posterior subarachnoid space was also seen(Figure 2). On imaging with neck flexion anteriorly shifted posterior dura was seen with congested posterior epidural venous plexus (Figure 3).On contrast application intense homogenous enhancement of venous plexus was seen(Figure 4).

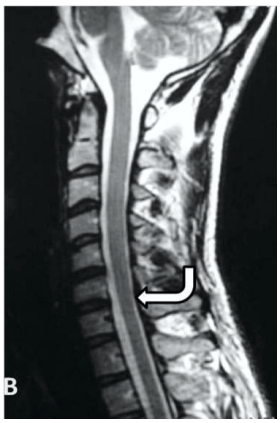

Fig. 1: Neutral
Suspicious cord thinning, obliteration of posterior epidural space

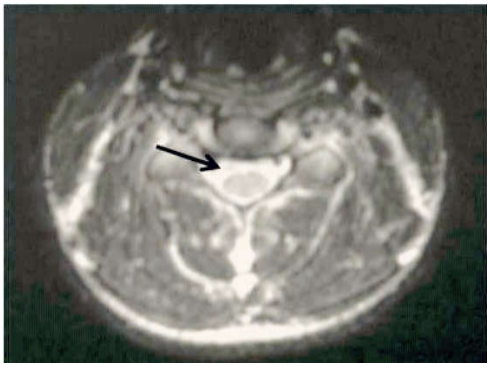

Fig. 2: Neutral
Asymmetric flattening of cord

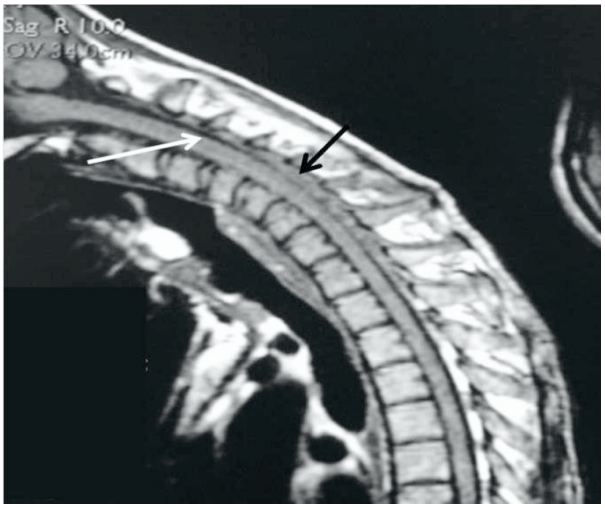

Fig. 3: Flexion
Anterior displacement of posterior dura with congested venous plexus

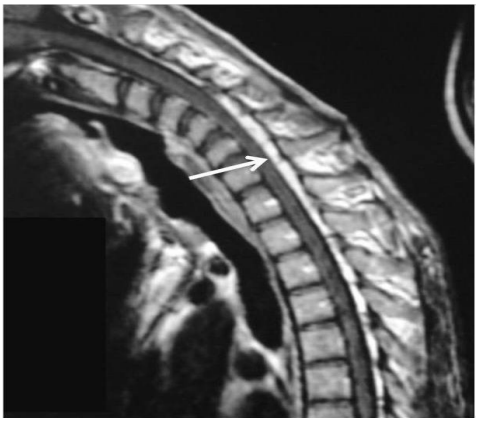

Fig. 4: Contrast
Intense homogenous enhancement of congested epidural venous plexus

Discussion

Hirayama disease is slowly progressive disease with attains stable phase later on. It is reported mainly in young males between the ages of 15 and 25 years of Asian population. High index of suspicion is required to perform exaggerated flexion MRI with contrast when spinal cord myelopathy is present in absence of canal narrowing. Insidious onset, weakness of unilateral upper extremity and atrophy of small muscles with no sensory or pyramidal tract involvement are main clinical presentations [1–5]. Although Hirayama et al first reported this disease in 1959 [2], pathologic study was not possible until 1982 [4], because of its benign course. Primary pathogenetic mechanism of Hirayama disease is presumed to be forward displacement of the posterior wall of the lower cervical dural canal in neck flexion which can be seen with MRI [6–9]. Disproportional length between the vertebrae and the dural canal was proposed by Kikuchi et al as mechanism of this anteriorly displaced dural canal in flexion[6]. The spinal dura mater is a loose sheath that is anchored in the vertebral canal by the nerve roots and by attachment to the periosteum in two places: one at the foramen magnum and the dorsal surfaces of C-2 and C-3, and the other at the coccyx [10].

In HD, the lower cervical cord moves forward in flexion and contacts the posterior surface of the vertebrae; the lower cervical cord becomes flattened at contact point. In addition, the posterior wall of the dural tube moves forward, the posterior epidural space expands forming a crescent shaped mass in the posterior epidural space formed by the congestion of posterior internal vertebral venous plexus [11] .This compression may cause microcirculatory disturbances in the territory of the anterior spinal

artery or in the anterior portion of the spinal cord. The chronic circulatory disturbance resulting from repeated or sustained flexion of the neck may produce necrosis of the anterior horns, which are most vulnerable to ischemia [12].

Conclusion

Hirayama's disease is a benign, non hereditary, non-progressive segmental spinal muscular atrophy. High suspicion is necessary for diagnosing this disease and early treatment. Decompression and fusion is the treatment of choice with some role of physiotherapy

Clinical Message

Hirayama disease is a very rare self-limiting clinical condition associated with weakness in the upper limb muscles. Early diagnosis is necessary as it is seen in younger people in their second and third decades. It is diagnosed only on flexion MRI of the cervical spine. It may be missed in many occasions as we usually advise just cervical spine MRI without mentioning flexion MRI. Use of a simple cervical collar to prevent neck flexion, has been shown to stop the further progression of the disease.

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