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HIRAYAMA DISEASE IN A YOUNG ADOLESCENT : A RARE CASE

Orthopaedics

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ABSTRACT

Hirayama Disease is a rare neurological condition also known as monomelic amyotrophy which commonly affects the distal upper limb extremities. It is self limited and commonly involve young male adults. There is focal amyotrophy of the distal upper limbs involving C7, C8, and T1segmental myotomes. The prevailing hypothesis postulates that the insufficient growth of the dura relative to the spinal column during puberty causes forward displacement of the dura in flexion with compression of the spinal cord which leads to the ischemia of the anterior horn cells at C8and T1. MR imaging in the flexion plays important role in the diagnosis. On MRI there is reduced bulk of the cervical cord with anterior displacement on flexion. The posterior laminodural space is with multiple veins within it which show enhancement on post contrast images. I report a rare case of 16yrs old male with similar imaging & clinical features.

KEYWORDS

monomelic, myotomes, weakness, flexion

INTRODUCTION

Hirayama Disease is a rare cervical myelopathy seen in the young male patients. It was initially described by Hirayama et al (1) in 1959 in a Japanese patient who had presented with unilateral atrophy of the distal upper limb. The disease was later coined as "juvenile muscular atrophy" of distal upper limb extremity(2) or "monomelic amyotrophy"(3). HD is characterized by an insidious-onset asymmetric atrophy with weakness of the distal muscles of the upper extremity with sparing of the brachioradialis muscle. It predominantly affects the C8–T1 segmental myotomes(4). In atypical presentation it may affect the lower limbs, however it is rarely seen (5). The commonly affected age group is second-to-third decades of life with age of onset of around 15–25 years (6). The male individuals are more commonly affected than females (6). The etiopathogenesis of this disease is debated , however the prevailing hypothesis postulates that the insufficient growth of the dura relative to the spinal column during puberty causes forward displacement of the dura in flexion with compression of the spinal cord which leads to the ischemia of the anterior horn cells at C8and T1(7).

I report a rare case with clinical and imaging features of Hirayama Disease in a young male presented to the department of radio-diagnosis at IGMC Shimla.

Case study

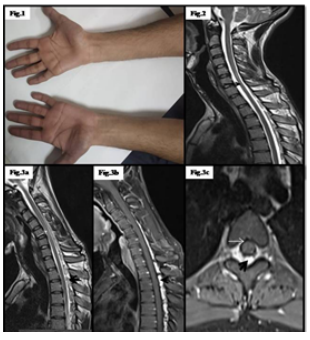
16 years old male patient presented with the complaints of progressive weakness of the bilateral distal upper limbs for the past 5 to 6 months. He had progressive worsening of the fine motor skills such as hand writing. Patient denied loss of sensation, autonomic dysfunction or any changes in the urinary or bowel habits. On examination there was asymmetrical atrophy of the bilateral thenar eminences and reduced bulk of the bilateral forearms (Fig 1). The tone was slightly reduced bilaterally with 3/5power in the right hand. Deep tendon reflexes were 2+ bilaterally. The sensation was intact bilaterally. Plantar reflex was normal.

For further workup MRI of the cervical spine was done.

Imaging features

MRI of the cervical spine was done in neutral position and flexion. There was reduced bulk of the cervical spinal cord at the level of C5-D1 vertebral levels (Fig 2). On flexion there was anterior displacement of the cervical cord which was compressed upon the cervical vertebrae (Fig 3a,3b&3c). There was widening of the posterior laminodural space on flexion which measures 3.4mm in neutral position and 7.6mm in flexion. Multiple flow voids were seen in the T2 & STIR images which were showing intense enhancement on post contrast images suggesting engorged veins (Fig 3b&3c).

Fig.1 is showing the reduced muscle bulk in the bilateral forearms (more on the right side) with atrophy of the bilateral hypothenar eminences. T2WI sagittal section in neutral position showing the hyperintense signal with reduced bulk of the cervical spinal cord at C5 to D1 vertebral levels (black arrow in Fig. 2).Sagittal T2WI (Fig 3a) & post-contrast T1WFatSat sagittal (Fig.3b) and axial sections (Fig.3c) in flexion show the anterior shift of the cervical cord (white arrow) & is anteriorly compressing upon the vertebrae. There is widening of the posterior laminodural space with multiple flow voids within it (arrowhead in fig.3a) which show intense post contrast enhancement of the veins in this region (arrowhead in fig 3b& 3c).



DISCUSSION

Hirayama disease is a rare neurological condition characterized by asymmetrical weakness and wasting of the distal upper limb predominantly involving the lower cervical cord C7, C8 &T1 myotomes. The young adults and adolescents ranging from 15 to 25yrs are predominantly affected. The initial presenting symptoms are slowly progressive hand weakness and fatigue, followed by cold paresis, tremors, and atrophy. Asymmetric distribution of symptoms and signs is characteristic, however bilaterally symmetric forms have also been reported (8).

The repeated or sustained flexion movements of the neck causes necrosis of the anterior horn cells of the lower cervical cord due to chronic microcirculatory changes in the anterior spinal artery territory, and is thought to be the possible etiopathogenesis of HD (9). Another possible cause could be the imbalance in growth between vertebral column and spinal canal contents, which leads to the compression of the anterior spinal cord against the vertebral column and detachment of the posterior dura, leading to widening of the laminodural space posteriorly. This ultimately causes microcirculatory disturbance and ischemic changes in the anterior spinal cord(10-13). In 2010, Ciceri et al (14) proposed that venous congestion in flexion might play an additional role in spinal cord ischemic changes. The venous engorgement is thought to be secondary to an impaired venous drainage toward the jugular veins during repeated or sustained neck flexion and an increased flow to the posterior internal vertebral venous plexus due to the negative pressure in the posterior epidural space because of anterior shifting of the dura. The imaging techniques play a very important role in the diagnosis of HD. Plain radiographs of cervical spine may only show loss of cervical lordosis, which is a very nonspecific finding. MR imaging is the most helpful technique in the diagnosis. The Conventional neutral position MR imaging may show an atrophied lower cervical cord and asymmetric cervical cord flattening with or without abnormal T2-weighted hyperintensities in the cervical cord, especially the anterior horn cells. However, MR imaging in neck flexion is more informative, which reveals classic

findings of posterior dural detachment with forward displacement of the posterior dural sac and loss of attachment between the posterior dural sac and the subjacent lamina, leading to widening of the laminodural space as seen in our case. Postgadolinium injection flexion MR images demonstrate moderate-to-intense enhancement of the engorged posterior epidural venous plexus, forming a crescent-shaped epidural mass, which might exerts a compression effect over the cord with or without flow voids(10,15). Lai et al (16) observed that forward shifting of the posterior dural sac was also seen in healthy subjects up to 46% , but without spinal cord compression. They further noted that the distance of such forward displacement was mild & was ranging from 1.0 to 4.2 mm with a mean of 1 mm, compared with those patients with HD in whom it ranged from 6.1 to 7.8 mm, with a mean of 6.7 mm. In our case the maximum forward displacement of the cervical spinal cord was 7.6mm. HD has a self-limiting course, and the management is usually conservative. Early recognition of HD is necessary because the patient can be advised to avoid or limit neck flexion movements, which helps to arrest the further progression of this disease. So, a high index of clinical suspicion is necessary to diagnose this entity because neutral position MR imaging may fail to diagnose it. Therefore, high clinical suspicion of HD is necessary in young patients with insidious onset of weakness of the hand and forearm muscles with muscle flaccidity. The flexion MR imaging sequences must be obtained to look for the widening of the laminodural space and anterior displacement of the posterior dura because it can be easily missed on conventional neutral position MR images.

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