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MRI FINDINGS OF HIRAYAMA DISEASE – A RARE CAUSE OF DISTAL UPPER LIMB WEAKNESS

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ABSTRACT

Hirayama disease, also termed non-progressive juvenile spinal muscular atrophy of the distal upper limbs, is a type of cervical myelopathy related to flexion movements of the neck.[1,3] It occurs mainly in young males between the ages of 15 - 25 years.[4] Here we report the MRI findings in two patients- a 16 years old young male presenting with insidious onset weakness, atrophy of left distal upper limb and another patient, a 23 years old male with bilateral distal upper limb weakness since 6 months. Both these young males had atrophy of distal upper limb and Hirayama disease as their clinico-radiological diagnosis. Magnetic resonance imaging (MRI) revealed mild short segmental atrophy and altered intramedullary signal of lower cervical cord in a neutral position. Anterior displacement of the posterior dura compressing the thecal sac and prominent dorsal epidural space with flow voids in flexion MRI study was observed. Both the patients were managed conservatively.

KEYWORDS: Hirayama disease, flexion MRI, cervical cord, dura.

INTRODUCTION

Hirayama disease also known as juvenile muscular atrophy of the distal upper extremity (JMADUE)[5] or monomelic amyotrophy (MMA)[6] and juvenile asymmetric segmental spinal atrophy.[7] It occurs mainly in young males between the ages of 15 - 25 years.[4] Typically presenting as insidious onset weakness and atrophy of distal upper limb. Although the etiopathogenesis of cervical myelopathy remains debatable, pathologic and radiologic findings suggest an abnormal compression or flattening of the anterior cord against the vertebral bodies during neck flexion, causing compression of the cervical cord and resulting in atrophic and ischaemic changes in the anterior horn of the cervical cord.

CASES

Case 1- sixteen years old male presented with insidious onset weakness and numbness in left hand since 2 months. The patient had complaints of poor grip with left hand. On examination atrophic changes were seen in the thenar, hypothenar, and interosseous muscles of the left hand and in the muscles of the left forearm. The deep tendon reflexes were symmetrically normal without Babinski sign. Sensory examination was normal. The right hand muscles were normal. There was no significant previous history of trauma or any chronic illness in this patient. None of his family members had the same symptoms.

Imaging findings

Patient underwent conventional plain cervical spine radiograph showing no definite abnormality. For further evaluation patient underwent MRI of cervical spine. Scan of the cervical cord on a 3 tesla MRI system was done. T1, T2 axial and sagittal images were obtained in neutral position. Scan showed mild atrophy of cervical cord from c5-6 to c6-7 with subtle altered medullary signal in this segment [figure: 1] these findings without any obvious other cause lead to further evaluation by scanning in flexion position. Prominence of dorsal epidural space is seen, predominantly in left aspect in flexion position showing flow voids on T2 sequence [figure: 2,4]. On extension MRI the epidural space got reduced and was the same as seen in scans on neutral scans [figure 3,5]. No contrast was given.

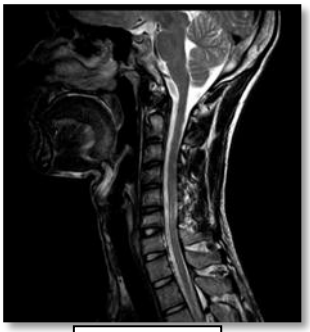
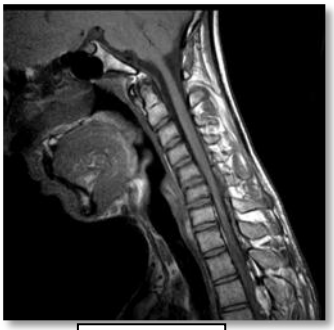


Figure 1



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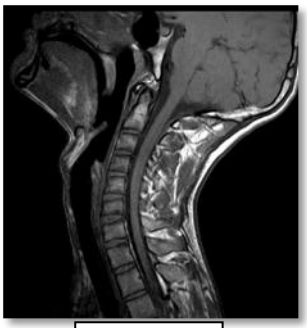

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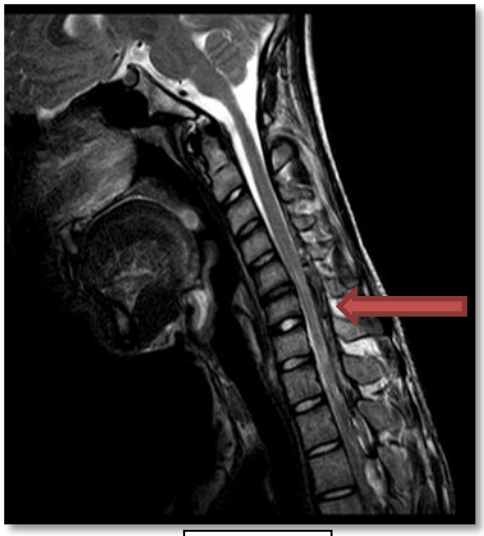

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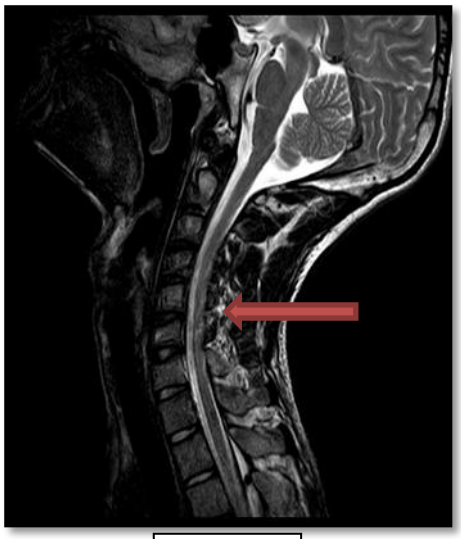


Figure 5

Figure 1: MRI sagittal section T2 weighted image in neutral position shows mild cord atrophy and intramedullary signal changes. Figure: 2, 4: MRI sagittal section T1 and T2 weighted image in flexion position reveals prominence of dorsal epidural space (arrow), showing flow voids on T2 sequence. Figure: 3, 5: MRI sagittal section T1 and T2 weighted image in extension position reveals, absence of dorsal epidural space (arrow).

Case 2- twenty three years old male presented with weakness of bilateral upper limbs, more on right side. Patient had complaints of difficulty in holding objects and limb tiredness. No sensory deficit was present. Deep tendon reflexes were normal. On examination bilateral thenar and hypthenar and forearm muscles were slightly atrophic.

Imaging findings

This patient underwent MRI cervical spine on a 3 tesla MRI system, pre and post contrast images were obtained in axial and sagittal planes. The scan revealed mild atrophy of cervical cord at the level C5 to C7 with signal changes in the cord in neutral position (figure:1,2). In flexion position, there was loss of attachment between the posterior dural sac and subjacent lamina (figure. 4). There was anterior shifting of the posterior dura causing compression of the lower cervical cord upon flexion of the neck. There was an enhancing epidural component in the lower cervical region in the flexion post-contrast scan. (figure:3,5).

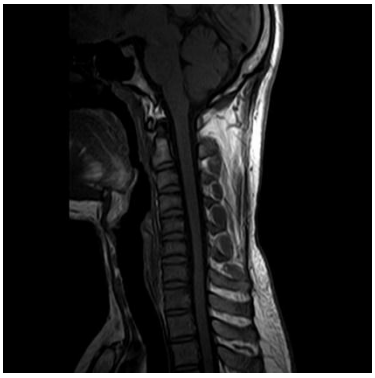
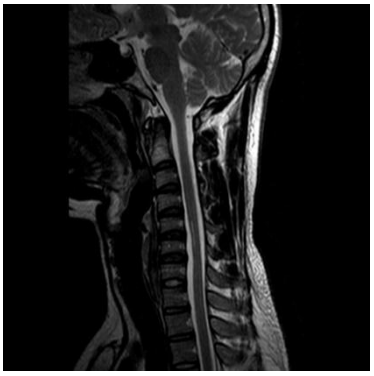


Figure 1



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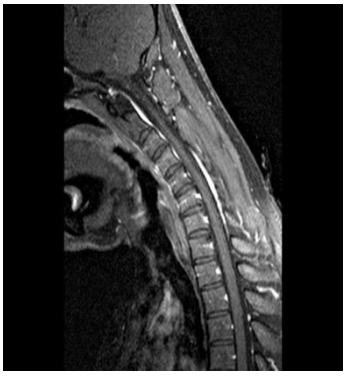

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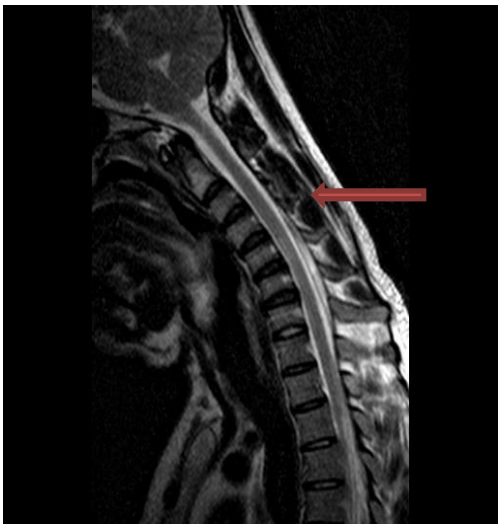

Figure 4

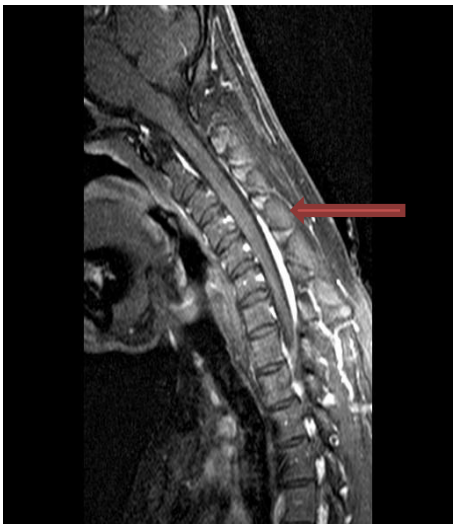

Figure 5

Figure 1, 2: MRI T1 and T2 sagittal images in neutral position revealing mild cord atrophy. Figure 4: MRI T2 sagittal scan in flexion position shows separation of dorsal dura (arrow). Figure 3,5: MRI T1 postcontrast images sagittal section reveals dorsal epidural space enchancement (arrow) and anterior shifting of the posterior dura causing compression of lower cervical cord.

DISCUSSION

Hirayama disease, also termed non-progressive juvenile spinal muscular atrophy of the distal upper limbs, is a type of cervical myelopathy related to flexion movements of the neck.[1,3] It occurs mainly in young males between the ages of 15 - 25 years.[4] Etiology and pathogenesis is still debatable however, the proposed pathophysiology is imbalanced growth between the patient's vertebral column and spinal canal contents will cause disproportional length between the patient's vertebral column and the spinal canal contents causing a tight dural sac. On neck flexion, the tight dural sac cannot compensate for the increased length of the posterior wall, which causes anterior shifting of the posterior dural wall and consequent compression of the cord against the posterior margin of adjacent vertebral bodies. This compression may cause microcirculatory disturbances in the territory of the anterior spinal artery in the lower cervical spinal cord. The chronic circulatory disturbance resulting from repeated or sustained flexion of the neck may produce gliosis and localized cord atrophy at the lower cervical region.[6,8]

Asymmetric cord flattening suggests another predisposing factor – "the posterior epidural ligament factor" – as put forth by Shinomiya *et al*. According to them, two kinds of ligaments between the posterior dura and the ligamentum flavum – one, fine ligaments; and the other, larger ligaments – contribute to resistance against separation of the posterior dura from the ligamentum flavum. Abnormal unequal distribution of the ligaments may be a cause of asymmetric cord compression.[7]

Imaging modalities like plain radiography is not contributory in diagnosing hirayama disease however, it can suggest other findings like straightning scoliosis or lordosis. Computed tomography myelography reveals asymmetrical cord flattening, with the epidural space seen as an area of low density behind the dural sac.[9]

As against these modalities, MRI is easy to perform and reveals various findings on neutral and flexion positioning. Localized lower cervical cord atrophy,

asymmetric cord flattening, parenchymal changes in the lower cervical cord, abnormal cervical curvature, loss of attachment between the posterior dural sac and subjacent lamina have been described.[10] Among these, localized lower cervical cord atrophy, asymmetric cord flattening and loss of attachment have an accuracy of 80% in identification of the disease; loss of attachment is the most valuable finding for diagnosing Hirayama disease in the neutral position.[10,11]

CONCLUSION

Hirayama is a rare disease of the young affecting men in the second to third decades of life. A high index of suspicion is required when imaging the spine in neutral position and can be confirmed with dynamic MRI in neck flexion giving characteristic findings. It is characterized by insidious onset and slowly progressive course followed few years later by static phase of unilateral or asymmetric atrophy of the hand(s) and forearm(s). Chronic microcirculatory changes in the territory of the anterior spinal artery supplying the anterior horns of the lower cervical cord is cause by repeated movement leading to cervical flexion myelopathy. Prompt diagnosis is important to institute early cervical collar therapy

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