Case Report – Mri Findings In Hirayama Disease Hirayama Disease----A Case Report

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Abstract
We report the MR findings in a case of Hirayama disease, a kind of cervical myelopathy related to flexion movements of the neck. In flexion MR studies, we can see the striking and pathognomonic picture of anterior shifting of posterior dura at the lower cervical spinal canal. In nonflexion studies, we find that asymmetric cord atrophy, especially at the lower cervical cord, though subtle, is highly suggestive of Hirayama disease. When it is seen, a flexion MR study is warranted to prove this diagnosis.
Hirayama disease, also termed nonprogressive juvenile spinal muscular atrophy of the distal upper limbs, is a kind of cervical myelopathy related to flexion movements of the neck (1–6). The pathogenetic mechanism of this disease is attributed to forward displacement of the posterior wall of the lower cervical dural canal when the neck is in flexion, which causes marked, often asymmetric, flattening of the lower cervical cord (1, 6–9). We report a case of Hirayama disease and describe the pathognomonic findings at flexion magnetic resonance (MR) imaging. We also discuss the mechanism behind this characteristic appearance and describe findings suggestive of Hirayama disease on routine nonflexion MR studies.
Keywords: *Hirayama disease, Flexion MR, Dynamic MRI, Cervical cord.*

I. Introduction

Hirayama Disease is a disorder of young adults of the age group twenty to thirty years.[1,2] It is an extremely uncommon disorder. Its other synonyms are juvenile muscular atrophy of the distal upper extremity (JMADUE)[3] or monomelic amyotrophy (MMA)[4] and juvenile asymmetric segmental spinal atrophy.[5] Although the cause of cervical myelopathy remains unclear, neuropathologic and neuroradiologic 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. We are reporting a case of Hirayama disease who presented in the neurology department with complaints of gradually increasing distal upper limb motor weakness and numbness.

II. Case Report

A 18-year-old boy had a 3-year history of slowly progressive weakness and thinning of the right hand and forearm that worsened in cold weather. More recently, he noted aggravated weakness of the right hand when he flexed his neck. His medical history was noncontributory; none of his family had the same symptoms. Neurologic examination revealed atrophic changes in the thenar, hypothenar, and interosseous muscles of the left hand and in the muscles of the left forearm, except the brachioradialis. Fine, irregular, and nonsynchronous tremulous movements in the left fingers were noted. The deep tendon reflexes were symmetrically normal without Babinski sign. Sensation to pin-prick, vibration, and joint position was intact. No extrapyramidal signs, Horner sign, or abnormalities in sweating and urination were noted

These findings were compatible with an anterior horn cell disorder of the cord. The plain cervical spine radiographs showed no definite abnormality. Nonflexion cervical sagittal MR images (1.5 T) revealed cord atrophy at the C4 -C6 vertebral level (Fig 1A). Axial T1- weighted and gradient-echo T2-weighted images showed evidence of cord atrophy, more obvious on the right anterior aspect (Fig 1B). Because the clinical presentation was reminiscent of Hirayama disease, a flexion cervical MR study was obtained. Sagittal and axial T1- and T2-weighted images showed anterior displacement of the posterior wall of the cervical dural canal below C-3, causing marked flattening of the cord (Fig 1C and D). An epidural mass, isointense with the cord on T1-weighted images and hyperintense on T2-weighted images, was noted at the posterior aspect of the lower cervical canal with some small flow void signals inside it (Fig 1C and D). After injection of contrast material, the epidural mass displayed strong and homogeneous enhancement (Fig 1E). This mass disappeared after the

patient returned to a nonflexion position, and it was considered to be engorged venous plexus due to dural shifting. The clinical presentation and the characteristic findings on flexion MR images led to the diagnosis of Hirayama disease. A neck collar was placed to prevent neck flexion, and the patient was doing well, with no further progression of symptoms, at the 3-month follow-up study.

Fig -1 Sag T2W MRI with neck in neutral position shows focal cervical cord atrophy at C4-C6 level without intramedullary signal abnormality. No extrinsic compressive lesion is seen.
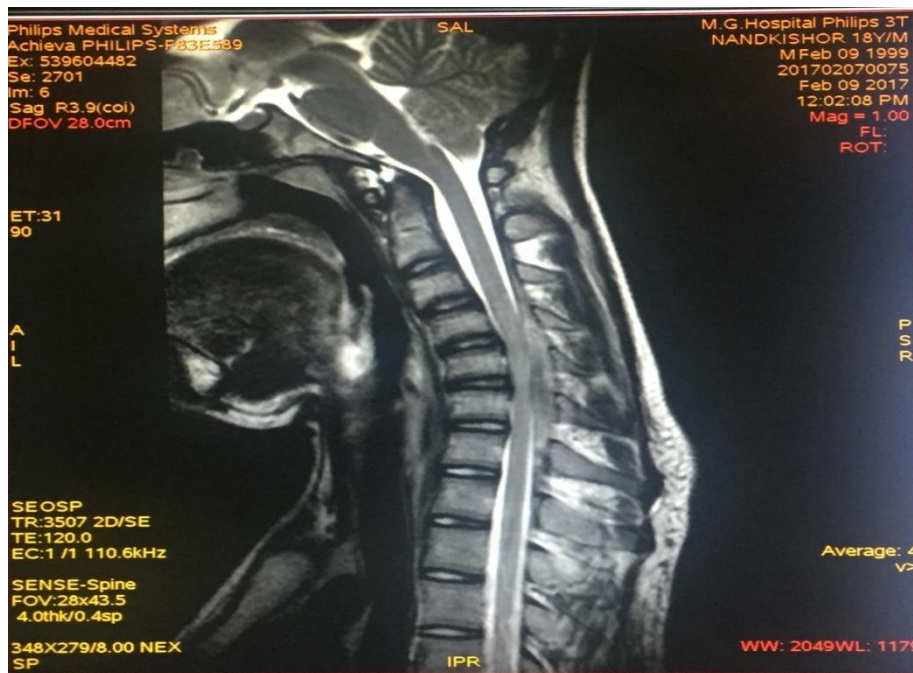

Fig – 2 Sag T2W MRI with neck in flexion position shows posterior wall of dural sac between C4-C6 vertebral level to shift anteriorly and anteriorly displaced cervical cord compressed over posterior surface of vertebral bodies**.**
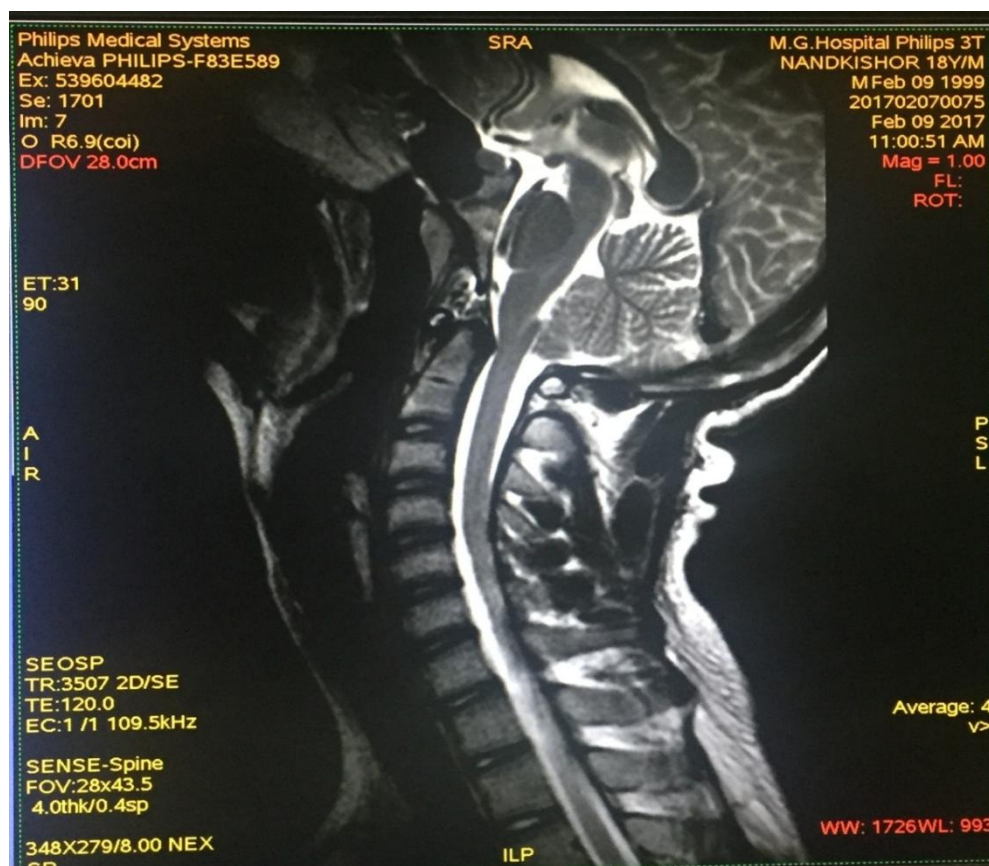
Fig – 3 Sag T2W MRI with neck in extension position shows repositioning of detached dura.

III. Discussion

First described by Hirayama *et al*. in 1959, sporadic juvenile muscular atrophy of the upper limb affects young men predominantly, and is characterized by an insidious unilateral or asymmetric bilateral muscular atrophy and weakness of the hand and forearm without sensory or pyramidal signs.[1]

HD is a benign disorder with a stationary stage after a progressive course for up to 6 or fewer years. It occurs mainly in young males between the ages of 15 and 25 years.[2,3]

Although the pathomechanism of the disease is debatable, Kikuchi *et al*. believe that an imbalanced growth causes disproportion in the lengths of the vertebral column and the spinal canal contents, resulting in a "tight dural sac." In the normal spine, the spinal dura is attached at two places – one at the foramen magnum, C2 and C3, and the other at the coccyx – and is anchored to the vertebral canal at the nerve root exits. In healthy subjects, this dura is slack and loosely suspended and consists of several transverse folds, which compensate for the increased length of the cervical canal in flexion. As against this, in Hirayama disease, a short dura cannot compensate for the increased length in flexion and so is displaced anteriorly, with resultant compression of the spinal cord.[3] Toma and Shiozawa proposed that the disproportionate shortening of the dural sac is accentuated during the juvenile growth spurt.[4]

The compression of the lower cervical cord by the posterior dural sac during chronic repeated flexion results in microcirculatory changes in the territory of the anterior spinal artery at the site of the most kyphotic level. Anterior horn susceptibility to ischemia accounts for the atrophy that follows, whereas the white matter is resistant.[2]. 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.[5]

Several conditions like syringomyelia, amyotrophic lateral sclerosis, cervical spondylosis associated with myelopathy, spinal cord tumor and traumatic myelopathy may cause localized amyotrophy of the distal arm, and should be differentiated from Hirayama disease by imaging modalities.[4] Conventional radiographic studies of the cervical spine are noncontributory and show only loss of cervical lordosis, straight alignment or scoliosis. Mild cord atrophy is noted on lateral myelograms, with forward movement of the posterior dural wall, reduction in the antero-posterior diameter of the dural sac and appearance of a lucent space behind the dural sac

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on flexion. However, myelography is cumbersome to perform as it is difficult to retain the contrast medium in the cervical subarachnoid space. Computed tomography myelography reveals asymmetrical cord flattening, with the epidural space seen as an area of low density behind the dural sac.[1,6]

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.[7] 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.[7,8]

On flexion MRI, forward migration of the wall of the dura mater is observed with an enlarged posterior epidural space.[1,9,10] A hyperintense, crescentic epidural mass showing curvilinear flow voids and uniform enhancement after administration of contrast is seen in the posterior epidural space.[10] Disappearance of this mass when the neck is in the neutral position suggests congestion of the posterior internal vertebral venous plexus.[3] A combination of three pathophysiological factors is responsible for this meningorachidian venous plexus engorgement. First, the anterior shift of the dural canal is responsible for negative pressure in the posterior spinal canal, with resultant increased flow to the posterior internal vertebral venous plexus.[8] Secondly, the anterior shift of the dura compresses the anterior internal vertebral venous plexus, with resultant increased burden on the posterior internal venous vertebral venous plexus, which leads to its subsequent dilatation.[10] Finally, the venous drainage of the jugular veins is reduced in neck flexion, which in turn impedes the venous return of the internal venous plexus.[8] A dynamic post contrast study as an important method for evaluation of suspected cases of Hirayama disease was stressed by Sonwalkar *et al.*[11] To ensure that this diagnosis is not missed, in patients presenting with focal wasting, after excluding motor neuron disease (MND), if the MRI otherwise looks normal, it should be repeated in flexion as well.

To conclude, we suggest the use of neutral and dynamic flexion MRI for the diagnosis of Hirayama disease. The flexion study aids diagnosis in patients of focal wasting if routine imaging is normal. Newer sequences such as 3D FIESTA could be further evaluated as an essential part of the screening protocol in patients with suspected Hirayama disease, thereby obviating the need for contrast administration and saving imaging time.

Conclusion

HD, a rare disease affecting young men in the second to third decades of life, 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) with sparing of the brachioradialis, characterized as oblique amyotrophy. It is thought to be a cervical flexion myelopathy related to repeated movements of the neck causing chronic microcirculatory changes in the territory of the anterior spinal artery supplying the anterior horns of the lower cervical cord. While dynamic contrast MRI is characteristic of HD, routine MRI has a high predictive value for diagnosis. Prompt diagnosis is important to institute early cervical collar therapy. Young adolescents with focal upper limb wasting should be evaluated to exclude HD or MMA, a benign condition amenable to collar therapy.

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