Role of dynamic MRI study in Hirayama disease

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Case Report

A 21-year-old male presented with slowly progressive weakness, thinning, and tremulousness of the left hand and forearm for 3 years. Neurological examination revealed moderate degree of wasting of forearm and small muscles of hand involving C7, C8, and D1 myotomes. Thenar, hypothenar, and introsseous muscles were weak. Deep tendon reflexes were normal in the upper limbs and brisk in the lower limbs with flexor plantar response. There were no long tract signs of objective sensory impairment. Electromyography (EMG) of affected muscles revealed features of chronic denervation with reinervation. Motor and sensory conductions were normal with no conduction block. Plain spinal radiograms showed abnormal straightening of the cervical vertebrae. Cervical MR images in the neutral position demonstrated focal atrophy of the cervical cord at the C5 and C6 vertebral levels. There was loss of dural attachment from subjacent lamina on left side [Figures 1a and b]. Dynamic flexion MRI showed the cervical cord to be displaced anteriorly and compressed over the posterior surface of the C 5–6 vertebral bodies with a prominent crescent shaped epidural mass with flow voids [Figure 1c]. The dynamic flexion MRI findings confirmed the clinical diagnosis of Hirayama disease.

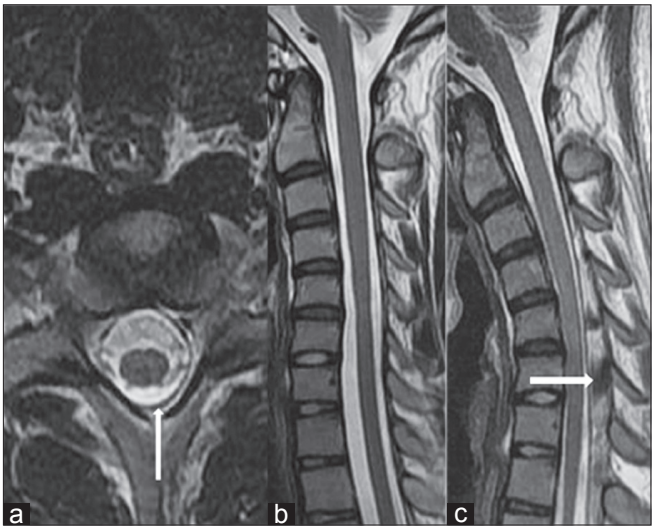

Figure 1: (a) Axial T2 weighted MRI in neutral position shows asymmetric cord atrophy with nearly 100% loss of attachment of dura from subjacent lamina on left side (arrow). (b) Sagittal T2 weighted MRI in neutral position show cord atrophy in lower cervical cord. (c) Dynamic flexion noncontrast T1 weighted sagittal MRI illustrates the characteristic anteriorly displaced cervical cord, flattened against vertebra with loss of subarachnoid space. Passive crescent shaped epidural venous congestion seen with prominent flow voids (arrow)

Discussion

Hirayama disease is the eponym for juvenile muscular atrophy of the distal upper extremity, also variably called as nonprogressive juvenile spinal muscular atrophy of the distal upper limbs, brachial monomelic amyotrophy, benign focal amyotrophy, juvenile muscular atrophy of the unilateral upper extremity, and juvenile asymmetric segmental spinal muscular atrophy.[1,2] The different names highlight the clinical features of the disease which predominantly involves unilateral upper extremity in adolescent males causing weakness without sensory or pyramidal tract involvement and cord atrophy.[1,3,4] Hirayama disease is a benign disorder with a stationary stage after an initial progressive course.[1,4] Due to the benign nature of the disease, only two autopsies have been reported. At pathologic examination, ischemic changes were found in the anterior horn cells most marked at C7 and C8 segmental levels distinctly differing from other degenerative diseases like motor neuron disease.[4]

Neuroradiological findings include straightening of spine/kyphosis, focal asymmetric lower cervical cord atrophy and/or signal alterations, loss of attachment between the posterior dural sac and lamina in neutral neck position.[2,5,6]

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The etiopathogenesis of the disease remains unclear; the Japanese authors have stressed the role of disproportionate growth between the vertebral column and the dural sac during the pubertal growth period to underlie this phenomenon.[2] The length of cervical canal is smallest during extension and the dural sac is slack. This cervical canal length increases during flexion when the dural sac is stretched, normally the slack of dura compensates for this increase. In Hirayama patients with disproportionate vertebral column and dural sac length, the dural sac is already stretched tight and the only possible movement for this tight dural sac is to follow the vertebral curvature and get anteriorly displaced, stripping the dural sac from subjacent lamina. However, this anterior shift of the tight dural sac has unwanted repercussion of pressing the cervical cord against the vertebrae causing ischemic atrophy of the anterior horn cells.[2,4]

The electrophysiological studies show worsening of findings during neck flexion.[7] The above discussion brings to the fore the importance of dynamic compression in Hirayama disease and not surprisingly the neutral position MRI has just 49% sensitivity for diagnosis of Hirayama disease, being lower than neutral position myelography and CT-myelography.[7] Dynamic evaluation of the cervical spine is needed to capture this compression, sagittal MRI in flexion and neutral position with or without contrast is required to diagnose the disease.

Dynamic flexion MRI shows the characteristic findings of anterior shifting of posterior wall of cervical dural sac, stretching and tightening of dural sac, flattening of the lower cervical cord against vertebra, effacement of subarachnoid space, enlargement of posterior epidural space which forms a crescent shaped mass, prominent epidural flow voids suggestive of epidural venous congestion and enhancement of the epidural space on postcontrast studies.[2,5-7] Unfortunately, neutral position MRI is the position routinely employed for cervical myelopathies, this fact highlights the need for increased awareness among radiologists and the clinicians about Hirayama disease who can then direct the patient for dynamic flexion MRI, which has a sensitivity of 87%. In doubtful cases full flexion CT-Myelography may be done which has sensitivity of 94%.[7]

The disease remits spontaneously in 5 years in 85% of cases; however conservative management with cervical collar has been shown to decrease the severity of myelopathy and duration of disease. Surgical options like anterior fixation have also been tried with variable success.[7]

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