INTRODUCTION

Hirayama disease is a form of cervical myelopathy related to flexion movement of neck. The pathogenetic mechanism of this disease is attributed to forward displacement of 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. We report a case of Hirayama disease and describe the pathognomonic findings at flexion magnetic resonance imaging and the possible pathogenetic mechanism of this disease.

CASE REPORT

A previously healthy 19 year old male presented with insidious onset of wasting and weakness in the left hand and forearm for the past 2 years. He also noticed twitching of muscles over that area. He denied history of weakness in his right upper limb. There was neither history of trauma nor family history of neuromuscular disease. Neurological examination revealed severe wasting and moderate weakness of left hand forearm (Figure -1). Fasciculations and polymyoclonus were present. Deep tendon reflexes were preserved. Sensation was intact. There was no extra pyramidal sign, Horner's syndrome or abnormalities in sweating noted. EMG showed neurogenic pattern in left abductor pollicis brevis suggestive of active denervation change. Motor nerve conduction was normal. Findings were suggestive of anterior horn cell involvement of C7—T1. Non flexion cervical MRI (Sagittal T2)image revealed cord atrophy at the C7—T1 vertebral level (Figure-2). Non flexion cervical MRI (Axial T2)image revealed asymmetrical cord atrophy with hyperintense signal (Figure-3). Flexion cervical MR imaging was done because of suspicion of Hirayama disease. It revealed anterior displacement of posterior wall of cervical dural canal causing flattening of the cord (Figure-4). The clinical presentation and flexion MR images are diagnostic of Hirayama disease. A neck collar was placed to prevent neck flexion. On follow up, no further progression of symptoms noted.

DISCUSSION

Hirayama disease is a benign disorder which occurs mainly in young males between the ages of 15 and 25 years. The clinical features include insidious onset, predominantly unilateral upper extremity weakness and atrophy, no sensory or pyramidal tract involvement. Cervical flexion MRI show forward displacement of the posterior wall of the lower cervical dural canal which is presumed to be a primary pathogenetic mechanism of Hirayama disease. This has been explained by a tight dural canal in flexion, caused by a disproportional length between the vertebrae and the dural canal. The imaging findings in our case is consistent with this hypothesis. 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 fora...
men magnum and the dorsal surfaces of C-2 and C-3, and the other at the coccyx. The remainder of the dura mater is only suspended and cushioned in the spinal canal by the epidural fat, venous plexus, and loose connective tissues. In neck extension, the dura mater of the cervical spine is slack and thrown into accordion like transverse folds. In neck flexion, the dura becomes tighter, because the length of the cervical canal increases as the neck moves from extension to flexion. Normally, the slack of the dura can compensate for the increased length in flexion; therefore, the dura can still be in close contact with the walls of the spinal canal without anterior displacement. In Hirayama disease, the dural canal is no longer slack in extension, because of an imbalance in growth of the vertebrae and the dura mater. Therefore, a tight dural canal is formed, which cannot compensate for the.

resulting from repeated or sustained flexion of the neck may produce necrosis of the anterior horns, which are most vulnerable to ischemia. In our case, we found that asymmetry is one of the most characteristic findings of this disease, both clinically and radiologically. Thus, in cases of adolescent onset of distal upper limb weakness, the finding of asymmetric cord atrophy on routine non flexion MR studies, especially at the lower cervical cord, should raise the suspicion of Hirayama disease. When this sign is seen, a flexion MR study should be performed to confirm the diagnosis.

Though Hirayama disease is self-limiting, early diagnosis is still necessary, because placement of a cervical collar will prevent neck flexion, which has been shown to stop disease progression. But in some cases, anterior cervical decompression with fusion is done if conservative treatment fails to arrest progression of the disease even after 5 years.

Fig 2: Non flexing cervical MRI (Sagittal T2) showing asymmetrical cord atrophy with hyper intense signal.

Fig 3: Non Flexion cervical MRI (Axial T2) showing asymmetrical cord atrophy with hyper intense signal.

Fig 4: Flexion cervical MRI (Sagittal T2) showing anterior displacement of posterior wall of cervical dural canal causing flattening of the cord.

REFERENCES


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