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An Intersting Case of Myelopathy – A Case Report

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Abstract

Hirayama disease, also known as monomelic amyotrophy (MMA), is a rare cervical myelopathy that manifests itself as a self-limited, asymmetrical, slowly progressive atrophic weakness of the forearms and hands predominantly in young males. The forward displacement of the posterior dura of the lower cervical dural canal during neck flexion has been postulated to lead to lower cervical cord atrophy with asymmetric flattening. We report a case of Hirayama disease in a 30-year-old man presenting with gradually progressive asymmetrical weakness and wasting of both hands and forearms.

Keywords : Hirayama's disease, Posterior Dural sac, MRI cervical spine

Introduction

Hirayama's disease is a rare benign disorder, also referred to as monomelic amyotrophy (MMA), Juvenile non progressive amyotrophy. It is a focal, lower motor neuron type of disease. Mainly young males in their second and third decades of age are most commonly affected. It is seen most commonly in Asian countries like India and Japan. In majority of people cause of this disease is unknown. MRI of cervical spine in flexion will reveal the cardinal features of Hirayama disease.

Case Report

A 30 Year old male admitted with the history of weakness and wasting of the distal fore arm muscles and small muscles of the hand for the past 8 years.h/o **wasting and weakness** of both **forearm and hand** for the past 8 years **insidious in onset** first started in the left hand **gradually progressed** to the left distal forearm .Six months later he noticed **similar complaints** in the right hand which **gradually progressed** to the right fore arm (distal > proximal). **After four years there is no further progression of weakness and wasting .**h/o difficulty in mixing the food & writing.h/o difficulty in buttoning and unbuttoning shirt. h/o muscle cramps and no fasciculations. h/o coarse tremors in both hands. h/o head ache and neck pain on and off for the past two years. He had no history of sensory disturbances, bladder and bowel disturbances. He had no history of trauma or fever.not a k/c/o diabetic, hypertensive, asthma, TB

On Examination

o/e Patient

conscious , oriented , afebrile

No pallor/cyanosis/clubbing/ pedal edema/jaundice/ generalised lymphadenopathy

VITALS :

BP – 130/80 mmhg PR -87/MIN RR- 16/MIN SBC > 30 TEMP – NORMAL

HEIGHT NECK RATIO - 12.5

HIGHER MENTAL FUNCTIONS - NORMAL

MOTOR SYSTEM

BULK :

INSPECTION : wasting of both distal fore arm and hand muscles clawing of hands +

MEASUREMENTS	**RIGHT (cm)**	**LEFT (cm)**
MID ARM	25	25
MID FORE ARM	14	14
MID THIGH	35	35
MID LEG	29	29

TONE	RIGHT	LEFT
ELBOW	NORMAL	NORMAL
WRIST & HAND	DECREASED	DECREASED
LOWER LIMB	NORMAL	NORMAL

POWER :

UPPER LIMB	RIGHT	LEFT
neck ,shoulder jt and scapula	5/5	5/5
Elbow - flexion & extension	5/5	5/5
Wrist -flexion & extension	3/5	3/5
Small muscles of hand	3/5	3/5
Hand grip	weak	weak
LOWER LIMB	**RIGHT**	**LEFT**
HIP	5/5	5/5
KNEE - flexion & extension	5/5	5/5
ANKLE	5/5	5/5
Small muscles of foot	5/5	5/5

TRUNK & ABDOMINAL MUSCLES - 5/5

Superficial and deep tendon reflexes – normal
Sensory system – intact
Cerebellum - normal
ANS – normal
Cranial nerves - normal
Extrapyramidal - no involuntary movements
Meninges - no neck rigidity , kernigs sign - absent
Spine and cranium – normal
Carotid - equally felt on both sides ,no bruit

Other Systems

CVS - s1 s2 + , no murmur
RS - b/l nvbs + , no added sounds
PA - soft

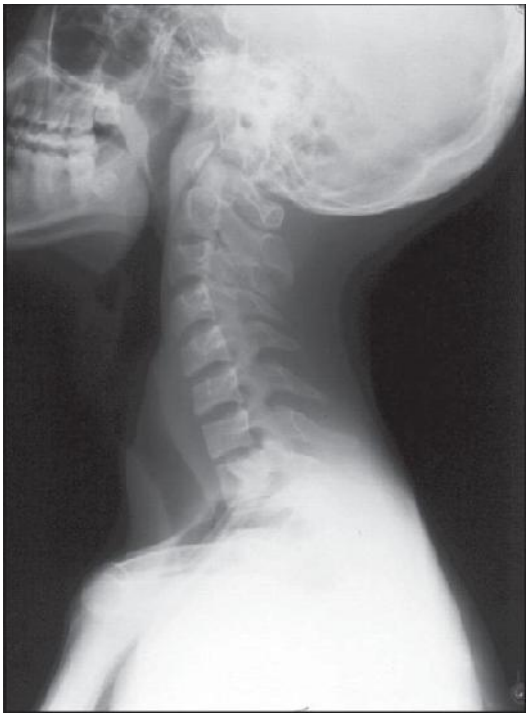
Investigations :

cbc
renal function tests
liver function tests
thyroid function tests
} within normal range

serum electrolytes: sodium - 142 meq/l ,magnesium - 1.8mgs ,potassium - 4.3 meq/l

calcium - 8.2 mgms/dl
CPK : 96 mcg/l
ESR : 26 mm/hr CRP : negative

Plain cervical spine X - ray was normal .



- Magnetic resonance imaging (MRI) of the cervical spine in neutral position showed thinning of cervical cord from C6 to C7 level, suggestive of cord atrophy [Fig - 1].

- MRI of the cervical spine in flexion shows flattening and forward displacement of posterior dural sac during flexion. Mild prominence of posterior epidural space at C6 to D3 space. Abnormally prominent enhancing epidural veins in upper dorsal canal when neck is flexed (fig – 2)

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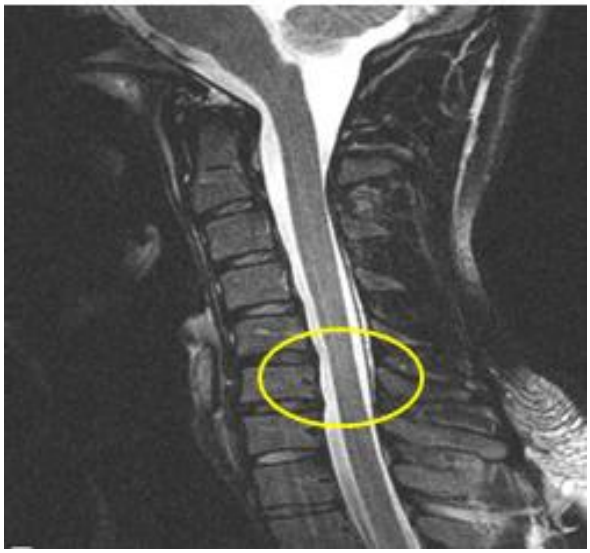
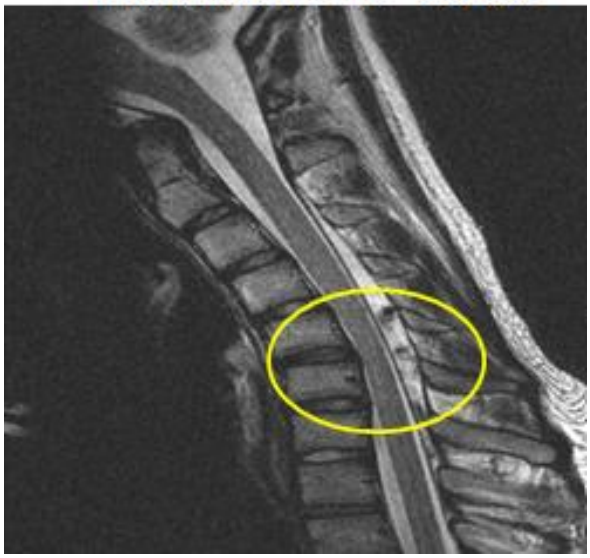
MRI CERVICAL SPINE – NEUTRAL



MRI CERVICAL SPINE - FLEXION



Discussion

Hirayama disease is characterised by focal amyotrophy with unilateral or asymmetric bilateral weakness and wasting of muscles innervated by C7, C8, and T1. It's an insidious onset, chronic, often self-limiting disorder with male preponderance, seen between the ages of 15 and 25 years. Hirayama et. al first reported this disease in the year 1959 , but pathologic study was not done till 1982 , because of its benign course. At pathologic examination, these authors found the lesions only in the anterior horns of the spinal cord from C-5 to T-1, particularly marked at C-7 and C-8 . Most commonly seen in Japan and other Asian countries like India and Malaysia .

The pathogenesis of the disorder is unknown – probable causes suggest that an imbalanced growth between the patient's vertebral column and spinal canal contents. This imbalanced growth will cause disproportional length between the patient's vertebral column and the spinal canal contents, which will cause a "tight dural sac" or "overstretch of the cord" in the neutral position and an anteriorly displaced posterior dural wall when the neck is flexed [9]. Current neuroradiologic techniques are able to show forward displacement of the posterior wall of the lower cervical dural canal in neck flexion, which is presumed to be a primary pathogenetic mechanism of Hirayama disease . In neck extension, the dura mater of the cervical spine is slack and thrown into 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. The difference in length between extension and flexion from T-1 to the top of the atlas is 1.5 cm at the anterior wall and 5 cm at the posterior wall . 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 increased length of the posterior wall during flexion. This causes an anterior shifting of the posterior dural wall, with consequent compression of the cord. This compression may cause microcirculatory disturbances in the territory of the anterior spinal artery or in the anterior portion of the spinal cord. The chronic circulatory disturbance resulting from repeated or sustained flexion of the neck may produce necrosis of the anterior horns, which are most vulnerable to ischemia.

In patients with Hirayama disease, conventional X ray of the cervical spine usually show no specific abnormalities except straight alignment or scoliosis. Myelography may show the forward movement of the posterior dural wall when the neck is flexed ; however, myelography is difficult to perform, as it is difficult to retain the contrast medium in the cervical subarachnoid space when the neck is flexed, regardless of the patient's position. MRI studies in neck flexion, which are easy to obtain, will show the forward displacement of the posterior wall and a well-enhanced crescent-shaped mass in the posterior epidural space of the lower cervical canal . This mass is thought to represent congestion of the posterior internal vertebral venous plexus rather than vascular malformations or tumors, because it vanishes once the neck returns to a neutral position . MRI shows atrophy of the lower cervical cord in a neutral position and there will be abnormal cervical curvature (straight or kyphotic) and loss of attachment

between the posterior dural sac and subjacent lamina, which is a most valuable in Hirayama disease.

Conclusion

In our patient initial work up was towards motor neuron disease. But the history of weakness of distal fore arm and hand muscles at the age of 22 which was insidiouds in onset gradually progressive and pontaneous arrest of weakness and wasting made us think of hirayama diasease.we took dynamic MRI study which showed atrophy of the lower cervical cord and post epidural space enhancing.

Even though Hirayama disease is a rare self-limiting disease, early diagnosis is necessary. Use of a simple cervical collar to prevent neck flexion, has been shown to halt the progression of the disease. Diagnosis of Hirayama disease is mainly based on flexion MRI of cervical spine. Asymmetry is one of the most characteristic findings of this disease, both clinically and radiologically. Thus, in cases of adolescent onset slowly progressive distal upper limb weakness followed by stabilization, with neurogenic changes in the EMG and the findings of asymmetric cord atrophy on regular nonflexion cervical spine MRI studies, especially at the lower cervical cord, one should keep in mind the diagnosis of Hirayama disease. When this sign is seen, a flexion MRI study should be performed to confirm the diagnosis.

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