



MANAGEMENT OF HIRAYAMA DISEASE - A CASE REPORT

Neurology

Dr Rajendra Pradhan	DNB Internal Medicine, Institute Of Neurosciences, Indraprastha Apollo Hospitals – 110076.
Dr. Pushendra N Renjen*	Sr. Consultant Neurologist & Academic Coordinator, Institute of Neurosciences, Indraprastha Apollo Hospitals- 110076. *Corresponding Author
Dr. Dinesh Chaudhari	Associate Consultant, Institute Of Neurosciences, Indraprastha Apollo Hospitals – 110076.
Dr. Abhas Kumar	DNB Neurology, Institute Of Neurosciences, Indraprastha Apollo Hospitals – 110076.
Dr. Charchit Gupta	DNB Neurology, Institute Of Neurosciences, Indraprastha Apollo Hospitals – 110076.
Dr. Anjali Mishra	FNB Critical Care, Max Super Speciality Hospital, New Delhi.

ABSTRACT

Hirayama's disease is a rare benign disorder, also referred to as monomelic amyotrophy (MMA), Juvenile non progressive amyotrophy, Sobue disease. It is usually seen during periods of rapid growth (15-25 years). It is a focal, lower motor neuron type of disease. Distal upper extremities characteristically show weakness and atrophy. Hirayama disease 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. In this paper a case is presented of this rare self-limiting disease. A 26-year old male patient presented with complaints of weakness of left little finger, insidious in onset, gradually progressive involving the other fingers of left hand followed by weakness of right upper limbs for 6 months. He also complaints of loss of muscle mass in left upper limb. MRI revealed the cardinal features of Hirayama disease. This case report presents information about the differential diagnosis, detailed symptoms and treatment methods of a Hirayama patient.

KEYWORDS

INTRODUCTION-

Hirayama disease (HD), a rare neurological condition, is a sporadic juvenile muscular atrophy of the distal upper extremities, which predominantly affects the lower cervical cord. It mainly develops in the late teens and early twenties with a male preponderance. The typical clinical features include insidious onset and slow progression of unilateral or bilateral muscular atrophy with weakness of the forearms and hands. It is seen most commonly in Asian countries like India and Japan [1,2], characterized by gradual onset of muscular dystrophy in distal part of the upper limbs related to flexion movements of the cervical spine [3,4-8]. There is no sensory loss, or the motor movement is delayed with respect to symptomatic findings. Muscle strength is also known as cold paralysis with poor prognosis with exposure to cold [12,13].

In Hirayama patients, movement tremor is seen, triggered by light mobilization on the fingers while resting [14]. The pathogenic mechanism in Hirayama's disease is due 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 [3, 8-11].

Hirayama disease is uncommon and the cause is unknown. Different theories have been postulated but the most accepted hypothesis is cervical myelopathy induced by function [15-16]. MRI cervical spine in flexion will reveal the cardinal features of Hirayama disease. We report a case of Hirayama's disease involving both the upper limbs and describe the characteristic MR imaging. Early diagnosis is necessary as it is seen in younger people in their second and third decades

CASE REPORT-

A 26 years old gentleman came with a history of insidious onset of weakness in both the hands left side followed by right side of 6 months' duration. He noticed weakness in the left little finger, progressively involved the other fingers of the left hand gradually progressed to the forearm muscles. Within next 1 month he noticed similar complaints in the right hand also, which was gradually progressed to the forearm

muscles. He also noticed loss of muscle mass of left hand and forearm which was gradually progressive in nature. He did not have any pain, loss of sensation, diplopia, dysphagia, ptosis, muscle cramps, fasciculations, headache or neck pain. There was no history of trauma, febrile illness, poliomyelitis or exposure to toxins or heavy metals in the past. There was no family history of similar complaints or neuromuscular disease.

On examination he was conscious and well oriented. General vital parameters were normal. His cranial nerves and sensory examination was normal. Motor examination showed bilateral complete clawing of fingers with gross atrophy of left upper limb more than right upper limb, distal weakness of left and right upper limb, superficial abdominal reflexes absent, DTR +++ except triceps which was absent. Coordination and gait were normal.

Blood routine investigations, KFT, LFT, TFT and CPK were within normal limits. Plain cervical spine X-ray was normal. EMG showed evidence of denervation in the form of fibrillation and fasciculations in both upper limb muscles. There was no involvement of the lower limb muscles. Magnetic resonance imaging (MRI)[Fig: 1,2,3] of the cervical spine in neutral position showed thinning of cervical cord from C4 to C7 level, suggestive of cord atrophy. On forward flexion of the cervical spine, When the neck was flexed, the cervical cord was displaced anteriorly and was compressed over the posterior surface of the C 5-6 vertebral bodies.

Based on the characteristic findings on flexion MR images and clinical findings diagnosis of Hirayama disease was made. He was operated with all the essential pre-operative work-up. Operative findings – engorged epidural veins, tense dura, thin cord. C3 – T1 laminoplasty along with duroplasty done using fascia lata, dura and para spinal muscle. Surgery was uneventful and patient was stable postoperatively. At the end of 6-month follow-up the patient was doing well, with no further progression of symptoms.



Fig: 1- MRI C Spine Neutral position showing thinning of cervical cord from C4 to C7 level, suggestive of cord atrophy



Fig: 2 - MRI C Spine Flexion showing anterior displacement of the posterior dura from C4 to T1 levels with maximal shift at C6 and C7



Fig:3-MRI image of hinging cervical involvement

DISCUSSION-

Hirayama disease is characterized 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 usually between the ages of 15 and 25 years [17,18-20]. Hirayama et. al first reported this disease in the year 1959 [18], but pathologic study was not done till 1982 [20], 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 [20]. probable causes of the disease suggest that an imbalanced growth between the patient's vertebral column and the 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 [21]. In neck extension, the dura mater of the cervical spine is slack and thrown into transverse folds [22]. 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 [22]. 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.

Scarcity of the disease and several atypical reported cases pose a diagnostic challenge; Tashiro et al. [23] recently outlined the criteria requirements for diagnosis of HD:

- (1) Distal predominant muscle weakness and atrophy in forearm and hand
- (2) Involvement of the unilateral upper extremity almost always all the time
- (3) Onset between the ages of 10 to early 20s
- (4) Insidious onset with gradual progression for the first several years, followed by stabilization
- (5) No lower extremity involvement
- (6) No sensory disturbance and tendon reflex abnormalities
- (7) Exclusion of other diseases (e.g., motor neuron disease, multifocal motor neuropathy, brachial plexopathy, spinal cord tumors, syringomyelia, cervical vertebral abnormalities, anterior interosseous, or deep ulnar neuropathy)

Apart from these features, many authors [24] report sparing of brachioradialis muscle, giving the impression of an "oblique atrophy."

The differential diagnosis of HD includes the distal form of spinal muscular atrophy, amyotrophic lateral sclerosis (ALS), post-polio syndrome, multifocal motor neuropathy with conduction block, and toxic neuropathy as well as structural lesions of the cervical cord (syringomyelia). These clinical entities can be identified by their characteristic clinical, radiological, and electrophysiological features [25].

There is no cure for MA. Treatment is conservative and with early detection aims to slow the progression of muscle wasting. A cervical collar worn in the early stages of disease has been shown to halt progress of the disease in some cases, as it prevents neck flexion. Muscle strengthening exercises and hand coordination training can also be helpful. patients should be evaluated for surgery on an individual basis, and surgery should probably be limited to the most severe cases that have progressed quickly.

CONCLUSION -

- Hirayama disease is a self-limiting disease however early diagnosis is necessary because a cervical collar may arrest the progression of the disorder.
- Physiotherapy is also helpful in preventing complication.
- MA is not life threatening but can cause a social disability in those with a complete loss of hand function.
- Early surgical intervention can minimize progression of disease.

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