Hirayama Disease Involving Bilateral Distal Upper Limbs of a Student: A Case Report.

Ushnish Mukherjee1, Rachit Gulati1, Pankaj Kumar Mandal2, Sunny Malik3

1Post Graduate Trainee, Department of Physical Medicine and Rehabilitation, R.G. Kar Medical College, Kolkata, West Bengal (India)
2Professor and Head of the Department, Department of Physical Medicine and Rehabilitation, R.G. Kar Medical College, Kolkata, West Bengal (India).
3Consultant Pain Physician, Department of Anaesthesiology, Max Super Speciality Hospital, Patparganj, New Delhi.

ABSTRACT

Hirayama disease is a rare neurological disorder characterized by initially progressive asymmetric muscular weakness and wasting of the distal upper limb(s) in predominantly young men, probably due to sustained or repeated neck flexion causing ischemic changes in the cervical anterior horn. Dynamic MRI (Neutral and flexed) of cervical spine is the mainstay for confirming diagnosis. A 16-year-old boy presented with insidious onset, gradually progressive difficulty in writing and other fine tasks, weakness, tremulousness of fingers, cold paresis and wasting of both hands and medial aspects of forearm (Right & Left) excluding brachioradialis for last 3 months without any trauma, pain, hypopigmented patch or thickened nerve and bladder or bowel involvement. No significant family, past medical or surgical, addiction history were found, but he had been studying for approximately 12 hours/day for last one year in a faulty posture on his bed suggestive of prolonged and repetitive neck flexion. General, sensory system and cranial nerves examination showed no abnormality. MMT was normal in both lower and upper limbs except for intrinsic muscles of hands. Signs of UMN or cerebellar lesions were not seen. Dynamic MRI of cervical spine revealed forward displacement of the posterior cervical dural sac at C5-C7. EMG, NCV of both upper limbs suggested denervated and reinnervated potential in C7-T1 muscles. Patient was advised to use Hard cervical collar and counselled regarding maintaining proper posture during studying in a properly configured chair-table. After 3 months, improvement in handwriting was noted without any signs of disease progression.

Keywords: Hirayama Disease; Neurology.

INTRODUCTION

Hirayama disease was first reported and described by Dr. K. Hirayama in 1959 in Japan.[1] It is a rare neurological disorder characterized by initially progressive asymmetric muscular weakness and wasting of the distal upper limb(s), more in right side.[2] About two-thirds of cases do not deteriorate clinically after about 5 years of disease onset.[2] Predominantly affecting men during their teenage and early twenties with a peak at 15-17 years.[2,3] It seems to be caused by dynamic compression of the lower cervical cord resulting from sustained or repeated neck flexion.[2-5] Its relatively benign course and characteristic appearance of oblique amyotrophy differentiate it from motor neuron disease.[2,4] As the Dynamic MRI (Neutral and flexed) of cervical spine is the mainstay for confirming clinical diagnosis, it is frequently underdiagnosed.[4-6]

CASE REPORT

A 16-year-old boy attended Physical Medicine and Rehabilitation OPD of R G Kar Medical College, Kolkata, with complaints of insidious onset, gradually progressive difficulty in holding pen while writing and other fine tasks, weakness, tremulousness of his fingers, cold paresis and thinning of both hands (Right>Left) for last 3 months. No history suggestive of trauma, neck/upper limb pain, sensory or bladder/bowel involvement. No significant family history, past medical or surgical history, addiction history was obtained. The boy had been studying for approximately 12 hours/day for last one year in a faulty posture on his bed suggestive of prolonged and repetitive neck flexion.

Physical Examination

General, sensory system and cranial nerves examination showed no abnormality. No hypopigmented patch or thickened nerve was seen. Wasting noted in the medial aspect of the forearms and hands (Right>Left) with bilateral preservation of brachioradialis. [Figure 1]
Manual Muscle Testing (MMT) was normal in both lower and upper limbs except for diminished power noted in intrinsic muscles of hands. Both power and pinch grip strength were diminished bilaterally when these were measured with Jammer Hand Dynamometer (Lafayette Hand Dynamometer, Model-J00105) and Pinch Gauge (Lafayette Pinch Gauge, Model-J00111) respectively. Cerebellar signs or signs of UMN lesion were not seen.

**Diagnostic Assessment**

MRI cervical spine: Contrast enhanced T1 sagittal in flexed position showed loss of dorsal dural attachment and anterior shifting with enhanced posterior epidural space from C4-7 which was not seen on neutral position MRI of cervical spine - only showed reversal of spine lordosis.

Nerve Conduction studies showed reduced Compound Muscle Action Potential with normal Sensory Nerve Action Potentials, EMG revealed occasional large-amplitude Motor Unit Action Potentials and reduced recruitment suggestive of denervated and reinnervated potential in C7-T1 muscles of both upper limbs.

All other routine investigations were within normal limit.

**Differential Diagnosis**

- C8-T1 radiculopathy
- Amyotrophic lateral sclerosis
- Syringomyelia
- Spinal cord tumors

**Treatment Given**

Hard cervical collar was given to the patient. He was counselled regarding proper posture and seating arrangements in chair-table during his study time.

After 3 months, the patient reported a subjective improvement in his hand-writing and better handgrip strength, though there was no significant change in MMT. [Figure 3]

![Fig.- 3. Hand writing – (A) Before & (B)After](image)

Most importantly, there was no further deterioration both in terms of power and muscle atrophy.

**DISCUSSION & CONCLUSION**

Typical history and clinical findings in this student pointed towards the diagnosis of Hirayama disease and confirmed by characteristic Dynamic MRI findings. Prolong and repetitive neck flexion was probably the precipitating factor for developing this disease as the boy was preparing for his 10th standard final exam for last 1 year in faulty posture. Though the etiopathogenesis of this disease is unclear, but most probably during the growth spurt, disproportionate growth of the spinal canal compared to dura leads to a tight dural sac causing anterior displacement of posterior dural wall and cord compression on sustained or repeated neck flexion. This leads to rise in intramedullary pressure causing chronic changes in microcirculation of anterior spinal artery territory supplying the anterior horns of the lower cervical cord resulting in ischemic changes in the cervical anterior horn involving mainly the C7, C8 and T1 myotomes.

The plausible aetiology of cold paresis is active denervation leading to conduction block of the muscle fibre membrane in reinnervating muscles.

Once the patient started using hard cervical collar with maintaining proper posture during studying in a properly configured chair-table, further cervical flexion injury was restricted resulting not only a halt in the disease progression but also showing some subjective improvement. It is important to confirm the diagnosis of Hirayama Disease by dynamic MRI at the earliest, as the application of hard cervical collar & posture correction during the early phase can halt the progression of the disease.

**Outcome And Follow-Up**

**Informed consent**
Informed consent was obtained from the guardian of the patient.

REFERENCES


Source of Support: Nil, Conflict of Interest: None declared