ORIGINAL RESEARCH PAPER

Medicine

KEY WORDS: unilateral upper limb atrophy, cervical collar, forward migration of spinal cord

DISTAL MUSCLE WASTING OF UPPER LIMB- A RARE ENTITY

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ABSTRACT

A 21-year-old male came with insidious onset of weakness of both hands, initially right followed by left of 3 years duration. On examination he had wasting of both hands, reduced power in both wrist and reduced hand grip. Lower limbs had no abnormality. MRI spine dynamic films showed forward migration of the spinal cord from C3-T1 noted on flexion, thinning of the cord from C4-C7 suggestive of cord atrophy. With these findings' diagnosis of juvenile sporadic atrophy of distal upper extremities (Hirayama disease) was made. Patient was prescribed cervical collar and he is under regular follow up.

INTRODUCTION:

Hirayama disease is also called juvenile muscular atrophy of unilateral upper extremity. Usually affect young men in second and third decade with male: female ratio 20:1. Patient presents with chronic wasting of one or both hands and forearms without sensory changes or long tract signs. The damage in this disease is from intermittent compression of lower cervical cord and gradual deterioration of lower cervical cord and gradual deterioration of the motor neurons in the anterior grey matter. The most seeming reason for this anterior horn cell disease is circulatory comprise secondary to forward displacement of tight dura leading to dynamic compression on flexion. What is most important about this process is the degree of recovery afforded by ligamentous sectioning and by similar surgical approaches that accomplish decompression of the lower cervical cord. The application of hard cervical collar during the early phase of illness can halt the progression of illness.

CASE PRESENTATION

21-year-old male who works at a garage came with complaints of weakness of both hands which was insidious in onset initially affecting only the 4th and 5th fingers of right hand. Patient at that time found it difficult to hold the water hose while washing cars, he used just his thumb, index finger and middle finger to do regular activities. The weakness was progressive and maximum after 3 months and static then. 6 months back he developed weakness of lateral 3 fingers first and then noticed similar pattern of weakness in his left hand. No weakness of lower limbs, sensory disturbances, bowel and bladder incontinence. No symptoms suggestive of cranial nerve and cerebellar involvement. No history of exposure to heavy metals, neck pain, trauma or electrocution. No significant family history. The patient was conscious oriented, vitals were stable. His cranial nerves and sensory examination was normal. Motor examination showed wasting of whole forearm and hand-thenar, hypothenar and lumbricals (right > left), clawing of right hand. No wasting of brachioradialis noted. Power was reduced in flexion and extension of wrist bilaterally, other muscles of upper limb and normal limb were normal. Superficial and deep reflexes were preserved.

INVESTIGATIONS

All routine blood investigations done were normal. Vitamin B12 and thyroid function tests were normal.

MRI Spine dynamic films (with neck flexed) showed atrophy of the spinal cord at C4-C7 levels, anterior displacement of spinal cord from C3-T1.

Nerve conduction study showed bilateral motor radiculoneuropathy of upper limbs. Nerve conduction velocity was preserved in bilateral median nerve and left ulnar nerve, reduced in right ulnar nerve. The amplitude was reduced along all the nerves and absent in the right ulnar nerve.

DIFFERENTIAL DIAGNOSIS

Motor neuron disease is a very close differential diagnosis of Hirayama, but UNLIKE Motor neuron disease this disease progresses initially and is followed by spontaneous arrest several years after the onset.

TREATMENT

This patient was explained about the nature of the disease and he was put on hard cervical collar to prevent neck flexion.

The patient is under regular follow up, he is doing well with no further worsening of symptoms.

DISCUSSION

Hirayama disease, a rare neurological condition, is a sporadic juvenile muscular atrophy of distal upper extremities, which predominantly affects the lower cervical cord. It mainly develops in the late teens and early twenties with a male preponderance.

One of the arguments regarding pathogenesis is that wasting and weakness associated with the disease is because of imbalanced growth between the patient's vertebral column and spinal canal contents. The imbalanced growth causes a tight Dural sac and displaces posterior Dural wall anteriorly when the neck is flexed. Normally, the spinal dura mater is loosely anchored to the vertebral canal by the nerve roots and the periosteum at Foramen magnum and dorsal surface of C2 and C3, Coccyx.

The dura mater is slack enough to adjust to the increased length of cervical spine in flexion.

In patients with Hirayama disease, however, the tight Dural sac separates the posterior Dural sac from its subjacent lamina and on neck flexion, cannot compensate for the increased length of the posterior wall. The posterior Dural wall shifts anteriorly, and the cervical spinal cord gets compressed against the posterior margin of adjacent vertebral bodies. This compression affects the anterior spinal artery and causes microcirculatory disturbances in its territory in the lower cervical cord. The anterior horn cells which are vulnerable to ischemia begin to

degenerate, resulting in localized cord atrophy of the lower cervical region, weak and wasted hands and forearms.

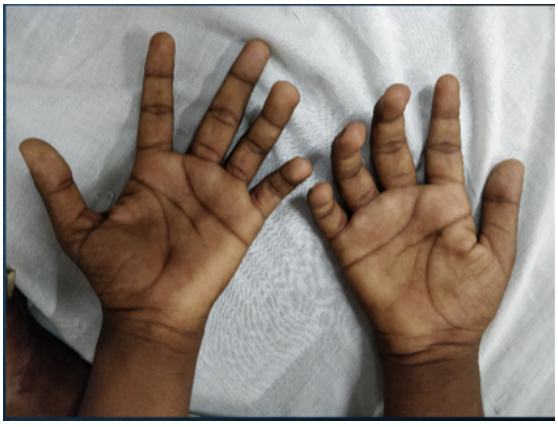
Hirayama disease is also associated with raised levels of serum IgE and several allergic disorders. Blood often stagnates in the compressed cervical cord allowing platelets to aggregate and causes histamine to be released, factors that cause arterial spasm and microcirculatory disturbances.

The key to diagnose this disease during MRI scanning is to obtain images when neck is flexed. MR studies in flexion not only show the anterior displacement of the posterior wall but also a well-enhanced crescent shaped lesion in the posterior epidural space of the lower cervical canal. This lesion is thought to represent congestion of the posterior internal vertebral venous plexus.

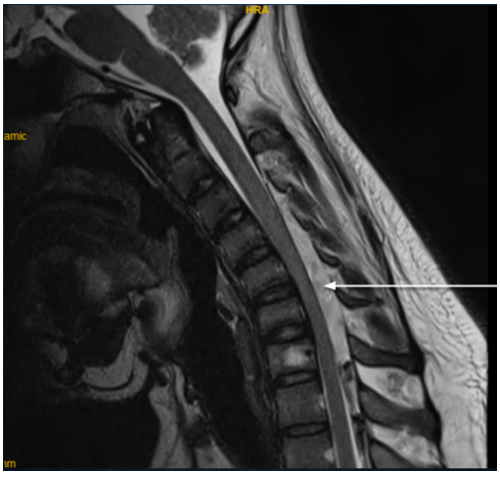
Although Hirayama disease is self-limiting disorder, early diagnosis is necessary because a cervical collar, by preventing neck flexion, may halt the progression of the disorder.

LEARNING POINTS

- *Hirayama Disease is a rare neurological disorder predominantly affecting young males*
- *Hirayama Disease should be suspected in patients presenting predominantly with unilateral distal upper extremity weakness sparing brachioradialis*
- *The key to diagnose this condition is flexion MRI of cervical spine.*
- *Early diagnosis is important as application of hard cervical collar which will prevent neck flexion, has been shown to stop the progression of the disease like in our patient.*



Bilateral Forearm Wasting



MRI of spine on flexion showing forward migration of spinal cord from C3-T1.
Atrophy of spinal cord noted from C4-C7 level.

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