Hirayama Disease: A rare case report with involvement of cervical spine

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Abstract

Introduction: Hirayama disease is a type of cervical myelopathy, also termed non progressive juvenile spinal muscular atrophy mainly involves forearm and hand means distal part of upper extremities. It's mainly affects the young male.it is benign motor neuron disease because mainly become stationary within 5 years of onset. Diagnosis is based on clinical findings and dynamic flexion MRI images showing anterior displacement of the posterior wall of dura mater and venous congestion in the epidural space. The patient of hirayama disease is mainly treated conservatively. Surgery is required only in severe cases.

Patient: A 19 year man came with a history of insidious onset of weakness in both the hands, left side followed by right of 4 years duration. He noticed weakness in the left hand muscles which was gradually progressed to the fore arm muscles. Within 6 months he noticed similar complaints in the right hand also, which was progressed gradually to the fore arm muscles. He had also noticed atrophy of muscles of hand and fore arm which was gradually progressive in nature. We present a cases meeting both clinical criteria and dynamic radiological criteria which suggests diagnosis of Hirayama disease. Patient has stabilized spontaneously over the course, and MRI scans show that typical changes have disappeared. On follow up no progression of disease occurred and condition remained stable.

Conclusions: Our case suggest that the condition of the patient with Hirayama stabilizes naturally; the natural course of the disease seems to recommend conservative treatment, for conservative treatment, a cervical collar is used which avoids neck flexion and prevents worsening of disease and provides remission. Surgical treatment is limited to patients with severe progressive disease.

Keyword: Hirayama disease, Cervical spine, Monomelic amyotrophy.

Introduction

Hirayama disease was first defined in Japanese in 1959, and in English in 1963, 1 as unilateral focal amyotrophy of one of the upper limbs; being a benign motor neuron disease showing ischemic necrosis of anterior horn of spinal cord. Other disease which shows similar clinical picture like this are Acute and chronic inflammatory demyelinating polyneuropathy in HIV, amyotrophic lateral sclerosis in physical medicine and rehabilitation, brainstem gliomas, chronic inflammatory demyelinating poly radiculoneuropathy, congenital muscular dystrophy, congenital myopathies.

It is mainly seen in young individuals of age 20-30 years mostly males. It is also known as monomelic amyotrophy but it also involves both upper extremity. Amyotrophy is not always monomelic; instead, it tends to be bilateral. Fasciculation's are infrequent. It mainly involves distal part of upper extremities including forearm and hands (C6-C7); with typical sparing of brachioradialis muscle (dermatome C5).Atrophy sparesT1 myotome. No signs of pyramidal tract involvement or sensory disturbance is noted.

The pathogenesiss of Hirayama disease comes from ischeamic changes of anterior horn cells of lower cervical spinal cord.

Surgical treatment is rarely used as the condition of the patient stabilizes after 2-3 years of progression.

Patient

Patient: 19-year-old male.

Medical History: The patient presented with weakness and atrophy of fourth and fifth finger of both upper extremities since last 3 to 4 years. It was associated with atrophy of hypothenar muscles but no pain or sensory disturbance was noted in both distal extremities. The disease progression was mainly seen in first year of onset.

Physical Examination: Examination revealed severe amyotrophy of the dorsal interosseous muscles of the right hand and muscle atrophy in the first, third, and fourth interosseous spaces and muscles of forearm (Fig. 1). The patient showed weakness of 4^{th} and 5^{th} digits of both hands with preserved sensation and mainly showed atrophy of muscle mass. Other neurological examination including reflexes was normal. 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.

Fig. 1: Muscle atrophy of muscles of forearm

Complementary Tests

Results from an electromyography study (EMG) suggested severe C7-8 radiculopathy and active denervation. The electroneurography (ENG) study showed evidence of chronic motor neuronopathy affecting C7-8 suggestive of hirayama disease. Electromyogram showed evidence of denervation in the form of fibrillation and fasciculation's in C7 and C8 distribution in both upper limb muscles. Left side was more predominant compared to right. Both Brachioradialis were spared. A cervical MRI scan in the neutral position did not show any major abnormality or intramedullary signal intensity changes (Fig. 3). MRI images with neck flexion shows anterior migration of dura mater compressing the spinal cord in C5 and C6 level. It also shows venous congestion in the affected area with decreased diameter of spinal cord at that region.

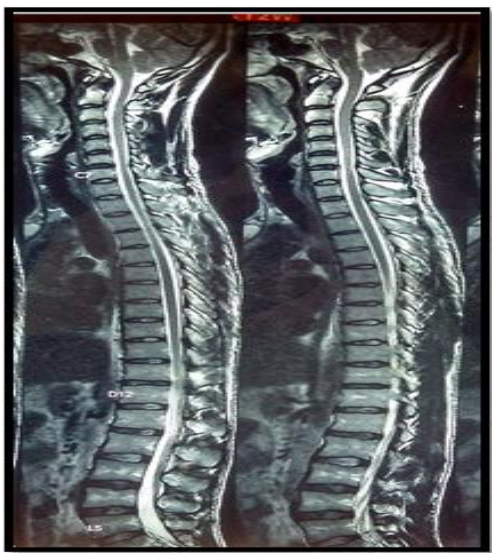

Fig. 2: Cervical MRI scan in the neutral position showing no gross abnormality

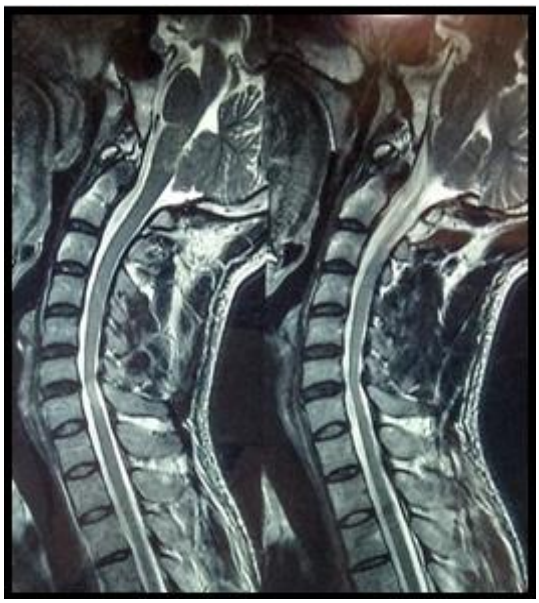

Fig. 3: Flexion cervical MRI scan (T2 and T1) showing marked dura mater shifting from the posterior lamina and accumulation of liquid and engorgement of the posterior epidural venous plexus. There is forward displacement of spinal cord at C5-C6 level

Discussion

The case presented here displayed typical symptoms of Hirayama disease: bilateral involvement of distal extremities typically sparing brachioradialis. The disease progressed insidiously and then became stable.

The main diagnosis of hirayama disease depends on MR images particularly in neck flexion; a neck flexion of 35 degrees has the maximum sensitivity of diagnosis. In neck flexion MR images shows forward displacement of dura mater and epidural venous plexus congestion in case of hirayama disease particularly in lower cervical spine region however it is not always diagnostic. Clinical correlation of the disease and electromyographic studies are utmost important to confirm the diagnosis.

The point to discuss in hirayama disease is the mode of treatment whether conservative or surgical; as disease stabilizes in 2-3 years. For conservative treatment, a cervical collar is used which avoids neck flexion and prevents worsening of disease and provides remission.

As the incidence of Hirayama disease is as low as 1/1 lac population and its natural course of progression, surgical treatment is rarely done. Surgical treatment is only required for rapid progressive disease.

Conclusion

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.

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