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Case Report

Hirayama Disease

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ABSTRACT

Hirayama Disease (HD), also known as Monomelic Amyotrophy is a cervical myopathy which is characterized by progressive muscular wasting and weakness of distal part of the upper limb in young men, followed by spontaneous arrest within several years. HD specifically affects the lower motor neurons of nerve roots C7, C8 and T1 myotomes. Males are more commonly predisposed in comparison to females. Symptoms of the disease typically begin between the ages of 15-25 years old. The disease is most common reported in Asian regions especially in Japan, China and India.

Here, we report a case of 17-year-old patient presenting with 8-month duration of graduate left hand and forearm weakness and muscle wasting. The clinical and radiological findings confirmed the diagnosis.

KEYWORDS: Monomalic amyotrophy, Amyotrophic lateral sclerosis, Oblique amyotrophy, Juvenile muscular atrophy

INTRODUCTION

Hirayama disease is a rare neurological disorder of juvenile muscular atrophy causing unilateral or asymmetrical bilateral weakness and muscular wasting which predominantly affects the lower cervical cord. Muscle atrophy involves muscles innervated by C7, C8 and T1 myotomes. The typical clinical features are insidious in onset and slowly progressive in nature that commonly affects young males between the ages of 15 – 25 years[1]. Sensory disturbances and autonomic involvement are rare. The overall incidence of Hirayama disease is low with most prevalence in Asian countries such as Japan and India.

CASE REPORT

A 17-year-old male presented with history of progressive weakness and wasting of the muscles of hand and forearm of the left upper limb from the past 8 months. Patient first noticed inadequate power in 4th and 5th fingers of his left hand followed by progressive inability to lift heavy objects over the period of time. These symptoms limited several daily activities especially during lifting heavy objects. Patient also complaints of aggravation of symptoms during forward bending (Flexion) of the neck and during cold weather where his left hand would be very cold or "Frozen". There are no associated complaints of neck pain, loss of sensation, tremors and muscle weakness in other parts of the body, radiculopathy or fasciculation. There is no relevant past history of medical or surgical instance, trauma or exposure to toxins. None of his family members have similar complaints.

Physical examination revealed relatively weak and wasted muscles of medial aspect of left forearm and muscles of hand (Thenar, Hypothenar and Interosseous muscles) [Fig.1]. Patient had no tremors, fasciculations or involuntary movements. Routine laboratory investigations such as Complete Blood Count, Thyroid Function Test, Renal Function Test and Liver Function Test were within normal range.

MRI of cervical spine was performed using T1W, T2W & STIR sequence in multiple planes in both flexion & neutral position using 1.5 Tesla Siemens MRI machine.

The MRI study revealed increased posterior epidural space on the flexion of the cervical spine with anterior

displacement of dorsal dura [Fig.2 & 3], most prominent at C4-C5 to C6-C7 levels with resultant reduced canal anterior-posterior diameter (7.3 mm).

Based on his clinical and radiological findings the patient received the diagnosis of "Hirayama disease".

Fig1. Relatively weak and wasted muscles of medial aspect of left forearm and muscles of hand.

examinations usually remain normal.

The exact etiopathogenesis of HD remains unknown, pathological study conducted by Hirayama et al[4] showed lesions in anterior horn cells of the spinal cord particularly marked at C7-T1 level with some degree of chronic vascular insufficiency along with shrinkage, necrosis and degeneration of large and small nerve cells with mild gliosis. With the

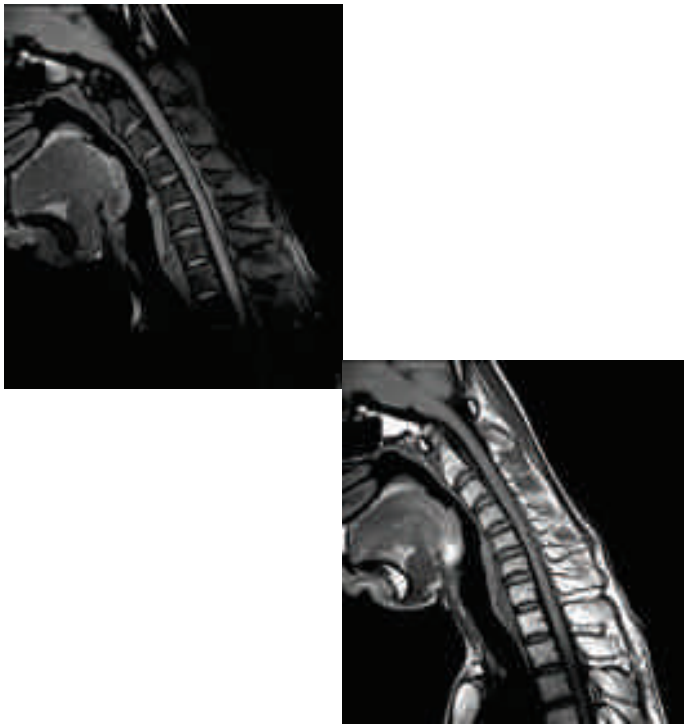

Fig 2 & 3. –Flexion MR sagittal T1W image (B) and T2W (C) image shows enlarged posterior epidural space with anterior displacement of dura.

CASE DISCUSSION

The disease was first described by Hirayama-et-al[2] in the year 1959 under the name of "Juvenile muscular atrophy of unilateral upper extremity" and in the year 1984 Gouriedevi etal[3] coined the term Monomelic Amyotrophy. Hirayama disease is cervical myopathy which affects C7, C8 and T1 segmental myotomes with sparing of the Brachioradialis muscle, which gives characteristic appearance of "Oblique amyotrophy" along with sparing of proximal muscles of the upper limbs. It manifests with gradual unilateral or asymmetrical bilateral muscle weakness and atrophy of the aforementioned myotomes. The disease has typical male predisposition in the age group of 15-25 years of age. It is a self-limited disease which usually progress for a few years and is considered benign in its nature because of its progressive course followed by stationary stage. Patient may complain of aggravation of symptoms on exposure of cold as called "Cold Paresis". The sensory system and reflexes and cranial nerve

current advancement in neuroradiology techniques and equipment recent studies also favor the theory of chronic ischemic changes to the anterior horn cells of lower cervical spine due to compression by increased laxity of dura mater[5]. Thus, repeated neck flexion causes chronic trauma and ischemia to the spinal cord ultimately leading to myelopathy which can be diagnosed on MRI cervical scans.

Normal routine MRI done in neutral head and neck position often render normal [Fig.4] or only minor changes in the region such as increased signal intensity in spinal cord, mild cord flattening or crescent shaped lesion in the posterior epidural space. It is only with specialized flexion MRI scans this disease can be appropriately diagnosed, specifically an anterior shift of the cervical dural sac causing spinal cord compression. The increase in lamino-dural space with presence of cervical spinal cord flattening during flexion [Fig.5] is essential for diagnosis of Hirayama disease[5].

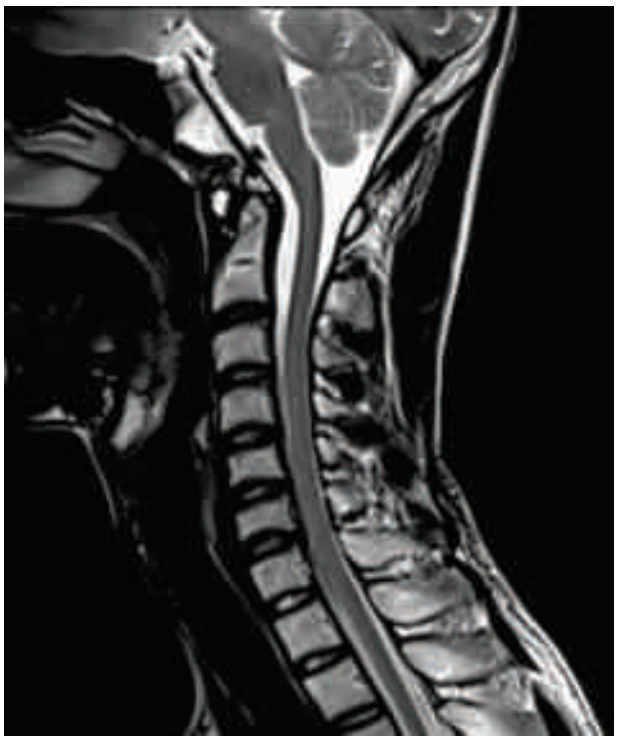

Fig4. Neutral position neck shows normal T2W MRI Cervical scan.

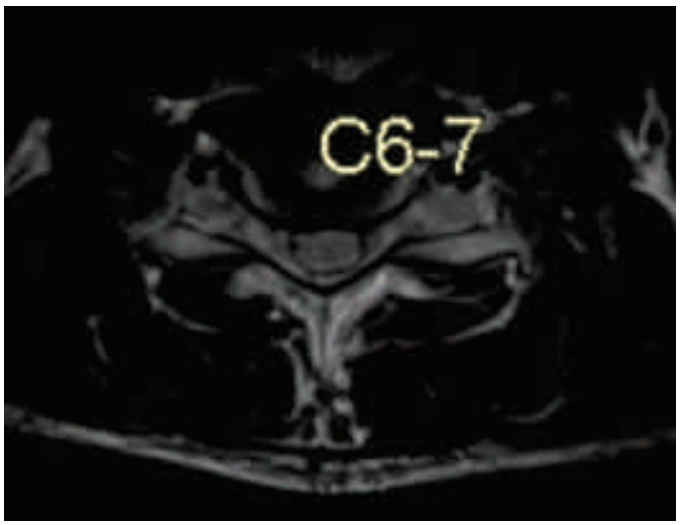

Fig5. Flexion MRI axial T2W images shows cord compression at level C6-C7

The differential diagnosis of Hirayama disease includes-

A.Amyotrophic lateral sclerosis,

B.Myotonic dystrophy,

C.Syringomyelia,

D.Post-polio syndrome.

Although Hirayama disease is a self-limiting disease, early diagnosis and conservative treatment with cervical collar may limit further damage by limiting neck flexion. Physiotherapy may also help.

In conclusion Hirayama disease is a rare neurological case with typical clinical and radiological findings and should be suspected in young male adults with weakness and atrophy in muscles of hand and forearm.

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