Case Report

Hirayama Disease – A case report

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ABSTRACT

Hirayama Disease is a non-progressive juvenile spinal muscular atrophy of the distal upper limb. The flexion movement results in cervical myelopathy which affects the dorsal aspect of the cervical cord. It is a benign motor neuron disorder and its initial course is progressive which subsequently becomes stationary. These findings are quite sensitive to magnetic resonance imaging in flexion position which sometimes may be missed in neutral position. We hereby report a case an adolescent boy who presented with asymmetrical weakness of both hands and his magnetic resonance imaging in flexion revealed changes suggestive of Hirayama disease.

***Keywords**: Hirayama disease, spinal muscular atrophy, cervical myelopathy, MR Imaging*

Hirayama disease or juvenile muscular atrophy of the distal upper limb is a rare condition and was first described by Keizo Hirayama in 1959. Histopathological confirmation could be done in 1982 which suggested the anterior horn cell involvement [1]. Hirayama disease characteristically affects adolescents and young adults and presents as weakness and atrophy of the distal part of the upper limbs. Weakness is usually unilateral or asymmetrically bilateral but can rarely present as symmetrical weakness also. Anterior shifting of the posterior dura of the lower cervical dural canal during neck flexion resulting in lower cervical cord atrophy with asymmetric flattening is the hallmark of the entity [2]. Hirayama disease with bilateral involvement has been reported rarely in the literature, thus highlighting the significance of imaging in such patients. Awareness of its natural history, careful examination, and use of magnetic resonance imaging (MRI) studies of the cervical spine, especially in flexion help making an accurate diagnosis of Hirayama disease.

CASE REPORT

A 16 years old male adolescent boy presented with the history of asymmetric atrophy, weakness and cold paresis of both the hands and forearms. Weakness initially started in the left hand and gradually involved the right hand also. Though pain was not the main complaint, patient reported to the pain clinic because of the weakness associated pain in both hands. These findings were progressive for the last three years and now stationary for the last two months. There was no history of trauma and his past medical history was also not significant. There was no family member affected by similar type of illness. Physical examination revealed atrophy of the thenar, and hypothenar muscles of both hands, more on the left side along with the mild atrophy of the muscles of both the forearms. Deep tendon reflexes and sensations were preserved. Power of the proximal muscles of both upper limbs was normal. Examination of the lower limbs, cranial nerves and cerebellar functions revealed no abnormality. Rest of the systemic examination was also normal.

Routine blood investigations including complete blood count, liver function test, renal function tests, serum electrolytes and muscle enzymes e.g.

creatinine kinase and lactate dehydrogenase were within normal limits. The motor nerve conduction velocities of the median and ulnar nerves were normal, but electromyography showed increased polyphasic waves along with fibrillations in the affected muscles sparing brachioradialis. There was no sensory involvement. These findings suggested involvement of the anterior horn cells of the lower cervical levels, and lead to a suspicion of diagnosis of Hirayama disease.

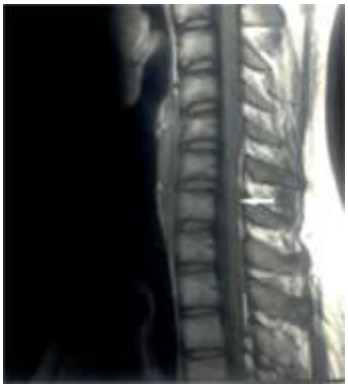

Figure 1 - T1W Sagital image of cervical spine in neutral position shows subtle thinning of cord at C7 vertebral level (white arrow).

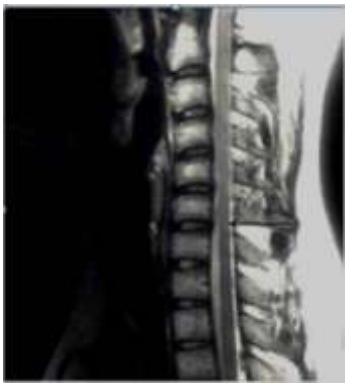

Figure 2 - T2W Sagital image of cervical spine in neutral position shows somewhat thinning of cord at C7 level with fuzzy outline (black arrow).

The patient was subjected to MRI cervical spine in both neutral and flexion positions. T1W and T2W imaging have shown increase in the epidural space at the level of C7-T1 [Figure1 and 2]. The cervical cord was thinned at this level. Contrast enhanced T1W and T2W flexion images were taken which confirmed the diagnosis of Hirayama disease [Figure 3 and 4]. There were multiple flow voids seen in the epidural space at the C7-T1 level and the cord has shown myelomalacia with atrophic changes [Figure 3]. There was a homogenous enhancement of this region in contrast images.

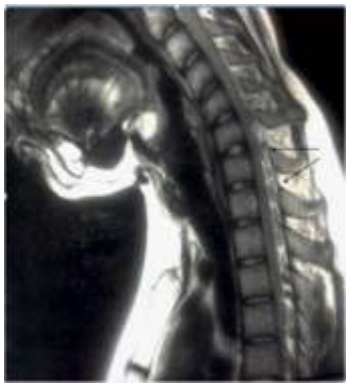

Figure 3 - T2W Sagital in flexion position. There are multiple flow voids in the posterior enlarged epidural space with thinning of the cord at the same level (arrows).

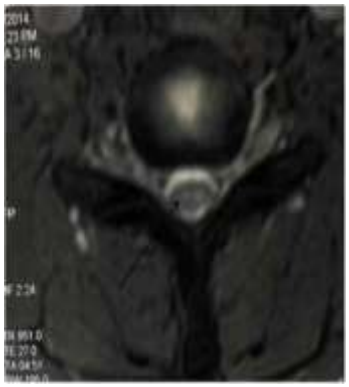

Figure 4 - T2W axial image shows thinning of the cord at C7 level (arrow).

On the basis of the clinical and radiological findings, diagnosis of Hirayama disease was made and the patient was advised to avoid flexion movements of the neck and cervical collar was prescribed. After 6 months of follow up there was no progression of the disease.

DISCUSSION

Hirayama disease predominantly affects the males in the age group of 15-25 years. It is insidious in onset causing predominantly unilateral upper limb

weakness and atrophy without any sensory or pyramidal tract involvement. It gives the picture of oblique amyotrophy as brachioradialis is spared. This is usually is unilateral but this can present as bilateral asymmetrical weakness [3]. In our case, it was bilateral asymmetrical as both the upper limbs were involved with different levels of atrophy and weakness [4]. In rare cases, there can be bilateral symmetrical involvement also. The underlying aetiopathology is due to the chronic microcirculatory changes in the region of anterior spinal artery because of strains due to repeated flexion. This leads to the necrosis of the anterior horn cells of the inferior cervical cord.

MR images in flexion show forward migration of the posterior wall of the duramater causing increase in the epidural space [5]. The cord at this level shows atrophic changes. Multiple flow voids can also be seen in this enlarged epidural space. The contrast images show homogenous enhancement of this region. This is because of engorged venous complex due to dural shifting and it disappears in the neutral position [6-7].

Hirayama disease is a self limiting disease; however, the earlier diagnosis can lead to prevention of unwanted flexion movements of the neck. The progression can be checked by advising neck collar as a first-line treatment. Surgical interventions such as cervical decompression and fusion have also shown some promising results and may be considered for selective patients [8].

CONCLUSION

Hirayama disease should be suspected in young patients presenting with upper limb muscle weakness and atrophy. Awareness of its natural history, careful examination, and dynamic flexion MRI studies of the cervical spine are the key for the accurate diagnosis of Hirayama disease.

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