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HIRAYAMA DISEASE – CASE REPORT

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ABSTRACT

Purpose - *Hirayama disease is an progressive disease caused by cervical neck flexion compressing the anterior horns of cervical spinal cord, primarily seen in young males in Asian descend and this condition is increasingly being encountered internationally.*

Material and Methods - *We present a young male with progressive hand and forearm weakness. We discuss the clinical presentation, appropriate investigations and management of this condition*

Results - *Our patient underwent MRI cervical spine with whole spine screening which revealed anterior translation of posterior Durameter at C5- C7 level. Successfully treated with surgery - C4-5 and C5-6 discetomy with C5 corpectomy was done with caging and fixation done with cervical soft collar post operatively.*

Conclusions - *This case illustrates typical presentation, diagnostic investigations and treatment of Hirayama disease. This will alert clinicians of this condition and optimize the management of affected individuals.*

KEYWORDS*: Hirayama disease; magnetic resonance imaging; muscular atrophy, spinal; spinal cord*

Article History

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INTRODUCTION

Hirayama disease is a benign, self-limiting cervical myelopathy first brought to attention by Hirayama (1) in 1959. Researches have established this disease as a new entity that differs from motor neuron disease because of spinal cord compression by the posterior dural sac during neck flexion. Male is predominant, clinically characterized as progressive course of distal upper-limb muscular weakness and atrophy. This new clinical entity was further consolidated by following reports of 20 patientsin 1963 by Hirayama K, Toyokura Y, Tsubaki T, et al. (2) and 38 patients in 1972 by Hirayama K. (3). This may be caused by congestion of the dilated venous plexus in the posterior epidural space (4, 5). We describe the symptoms, characteristic images, treatment, and clinical course of this case.

CASE PRESENTATION

18-year-old male patient with a history of weakness of bilateral upper limbs since a 6- months, it was insidious in onset and progressive in nature. He was aware of difficulty with raising both arms when changing clothes, he also experienced difficulty in lifting heavy substances . Weakness in both arms gradually progressed over the course of 6 months for which he was admitted in our hospital Neurological examination revealed muscular weakness and atrophy in the bilateral thenar and hypothenar muscles, predominantly affecting the right side as seen **fig 1.and fig 2.** He was alert and cranial nerves

were intact. His sensory system revealed no abnormalities. . Tendon reflexes were bilateral mildly decreased in the biceps, brachioradialis andtricep reflex. Cerebellar function and gait were normal.

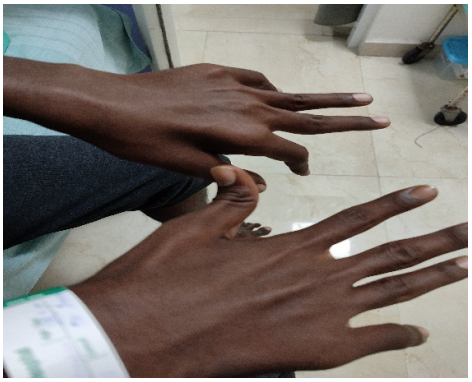

Figure 1

Figure 2

Complete blood counts, blood chemistry, and urinalysis revealed normal results. Nerve conduction studies were normal.

MRI Cervical spine with whole spine screening was taken and on sagittal T1 and T2-weighted cervical MRI in the neutral position, curve of the cervical was kyphotic at the C5–C7 vertebral level. Onflexion imaging there is detachment and anterior translation and of the posterior dura with prominence of the enlarged epidural space with prominent flow voidsseen in **fig3 and fig 4**

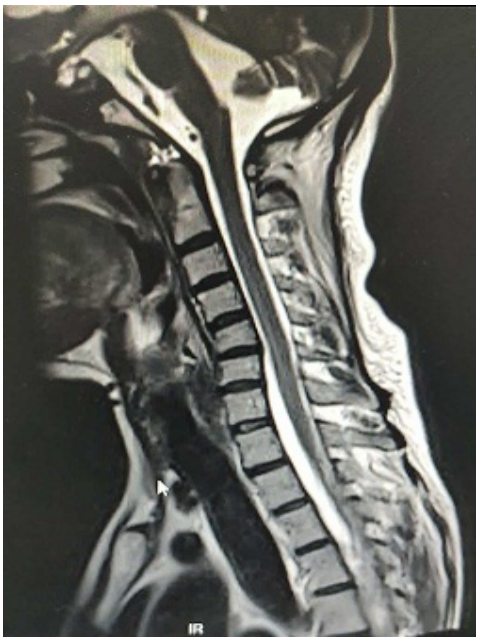

Figure 3

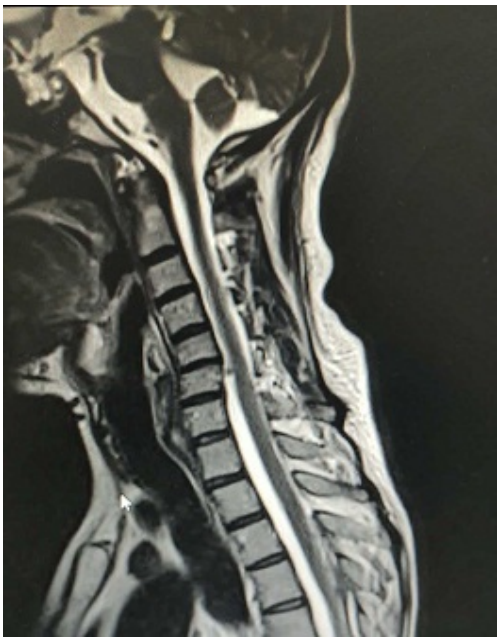

Figure 4

Elective surgery was done procedure – right sided approach with transverse incision was given , anterior cervical discectomy C5 –C6 and C6-C7 and corpectomy of C5 with caging and fixation was done as seen in **fig 5.**post operatively , patient was advised to wear soft cervical collar for 6-8 weeks and provided guidance in avoiding neck flexion exercises. We have conducted follow-up examinations every 3 months. After 12 months, his symptoms were stable with some mild improvement.

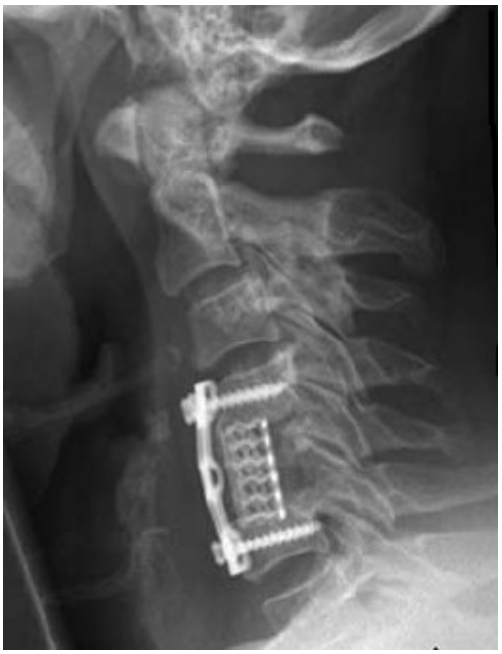

Figure 5

DISCUSSIONS

Our case presented with muscular weakness and atrophy in the distal muscles of upper limbs bilaterally at the C5–C6 myotome level. . In the flexion position of the neck, the dura mater shifted forward with an enlarged posterior epidural space, including homogeneous gadolinium enhancement of the epidural space. Neuro radiological findings were consistent with Hirayama disease. Similar cases have been described, showing juvenile muscular atrophy of the distal upper extremities caused by a pathogenesis similar to Hirayama disease. To the best of our knowledge, six such cases have been reported, including the present patient (6- 10).These cases with "proximal-type Hirayama disease" were characterized by spinal cord involvement at the C5–C6 vertebral level in young men. Cervical kyphosis may produce a relatively higher level involvement of the spinal cord at cervical flexion position, resulting in mid cervical segmental myelopathy.

Several reports have described the usefulness of surgical therapy with cervical spinal fusion (11). Use of a cervical collar is recommend to avoid neck flexion. We successfully treated our case by anterior cervicalC4-5 and C5-6 discectomy and corpectomy of C 5 with caging and fixation and wearing a neck soft collar. His symptoms were stable with some mild improvement.

CONCLUSIONS

According to our case and previous reports, a subtype of Hirayama disease presents with distal involvement of the upper extremities. The patho mechanism of proximal and distal involvement is speculated to be due to an unusual spinal column alignment as so-called "cervical kyphosis," involving a difference in the apex vertebral level in cervical flexion, resulting in symptoms different from typical Hirayama disease.

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