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Case Study

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# FLEXION MRI IN HIRAYAMA DISEASE

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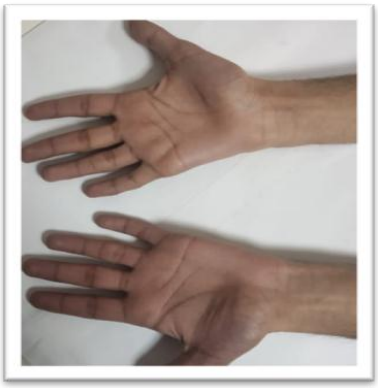
**KEYWORDS:** Hirayama disease; Flexion MRI; Cervical spine.

## INTRODUCTION

Hirayama disease is also known as juvenile muscular atrophy of distal upper limb or monomelic amyotrophy. It is a rare disorder characterized by asymmetric distal weakness and muscle wasting mainly affecting cervical segmental myotomes. The disease most commonly affects young adults in the age group of 15-25 years, with a male predominance. It is mostly sporadic however few familial cases have been reported in literature.

## CASE REPORT

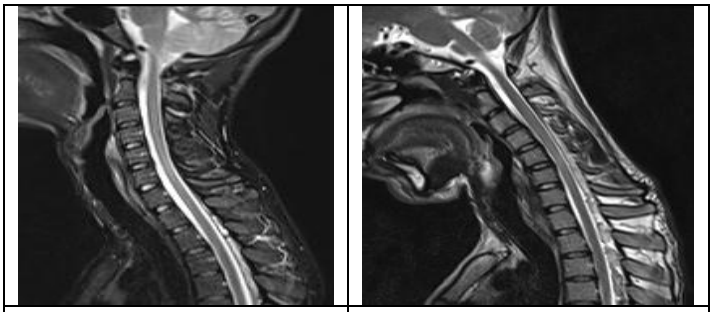
Case report of a 16 year old male patient who presented with history of progressive weakness and muscle wasting in bilateral upper limbs for past 5 months. There was no history of trauma in past. On clinical examination, there was reduced bulk of forearm muscles and atrophy of thenar muscles bilaterally. Based on history and clinical examination diagnosis of Hirayama disease was kept and MRI cervical spine was requested.



Clinical photograph showed reduced bulk of thenar muscles bilaterally.

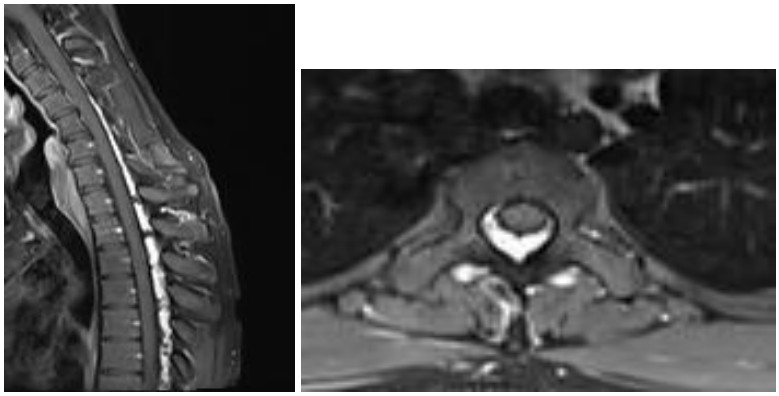
## IMAGING FEATURES

Images were acquired on 1.5 Tesla scanner (Avanto, Siemens).



**(A) Neutral position** **(B) Flexion**

(A) In neutral position, there was loss of cervical lordosis with long segment T2 signal intensity in cord.
(B) Flexion MRI showed increase in laminodural space with compression of spinal cord.



Post contrast images showed marked enlargement and enhancement of posterior epidural plexus with forward displacement of spinal cord.

## DISCUSSION

Hirayama disease is an uncommon entity which differs from motor neuron disorders as prognosis in this condition is favorable; prompt recognition allows early intervention and stops disease progression.

Kikuchi et al suggested that a disproportion in length between vertebral column and the spinal canal content results in tight dural sac. During flexion cervical cord increases in length upto 3 cm. However the dura, attached to foramen magnum cranially and coccyx caudally; can not compensate for this increased length of cord. Therefore dura displaces anteriorly and compresses spinal cord.

On neutral MRI, findings might be subtle and difficult to identify. MRI with flexion sequence is highly specific in diagnosis of Hirayama disease. Pathognomonic findings of this disorder are symmetric spinal cord atrophy with widening of lamino-dural space which are best visualized on flexion MRI.

## CONCLUSION

Radiologist should maintain a high level of suspicion for Hirayama disease in young male patients with asymmetric upper limb weakness and muscle atrophy. Inclusion of flexion sequence to routine neutral MRI increase the detection rate of this disorder.

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