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Case Report

Radiology Section

Hirayama Disease Diagnosed by Dynamic MRI before the Onset of Clinical Muscle Wasting: A Case Report

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ABSTRACT

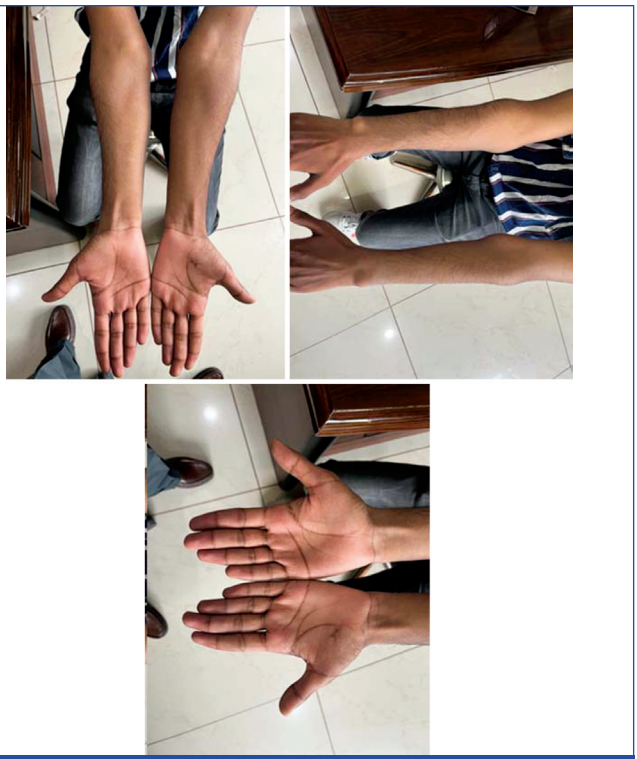
Hirayama Disease (HD) is a rare, flexion-induced cervical myelopathy classically characterised by distal upper-limb weakness and muscle wasting. Muscle atrophy is widely regarded as a hallmark diagnostic feature, and all previously reported cases describe varying degrees of amyotrophy at presentation. A 21-year-old male presented with intermittent neck pain and a completely normal neurological examination. Neutral-position Magnetic Resonance Imaging (MRI) revealed a short-segment T2-weighted hyperintensity at the C5-C6 level without spinal cord atrophy. Dynamic flexion MRI demonstrated anterior displacement of the posterior dura mater from C5 to T1, with associated posterior epidural venous plexus engorgement findings diagnostic of Hirayama disease. Dynamic flexion MRI plays a crucial role in identifying early or subtle disease before irreversible anterior horn cell loss occurs.

Keywords: Cervical myelopathy, Magnetic resonance imaging, Monomelic amyotrophy, Neck pain, Upper-limb weakness

CASE REPORT

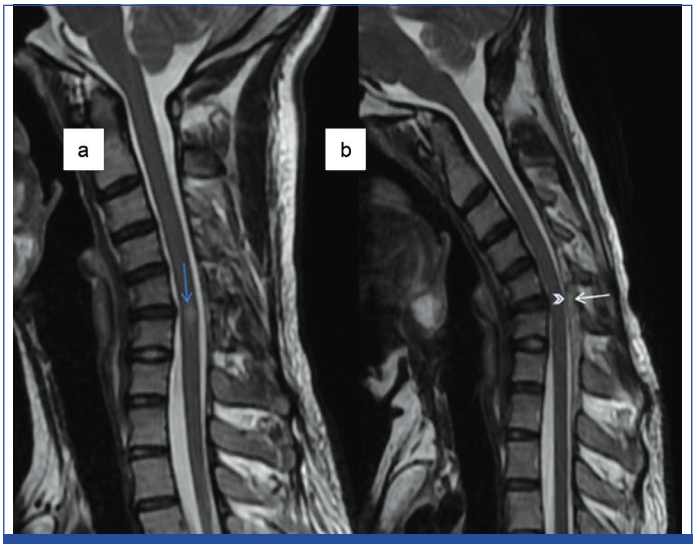
A 21-year-old male presented with intermittent neck pain for three months. The pain was episodic, non radiating, and not associated with weakness, numbness, tremors, cold paresthesias, or functional limitations. There was no history of trauma, systemic illness, or prior neurological symptoms.

Neurological examination was entirely normal. Motor strength was full (Medical Research Council grade 5/5) in all upper and lower limb muscle groups. There was no evidence of muscle wasting, fasciculations, or asymmetry involving the forearms or intrinsic hand muscles [Table/Fig-1]. Deep tendon reflexes were normal and symmetrical, and sensory examination was unremarkable.



[Table/Fig-1]: Clinical photographs of the patient's upper limbs showing symmetrical forearms and hands with preserved muscle bulk. There is no evidence of distal muscle wasting, fasciculations, or asymmetry involving the thenar, hypothenar, or interosseous muscles, supporting the absence of clinical amyotrophy at presentation.

Routine cervical spine MRI in the neutral position demonstrated a short-segment T2-weighted hyperintensity within the cervical spinal cord at the C5-C6 vertebral levels, without associated cord atrophy. Dynamic flexion MRI revealed anterior displacement of the posterior dura mater extending from C5 to T1, accompanied by posterior epidural venous plexus engorgement, confirming the diagnosis of Hirayama disease despite the absence of muscle wasting [Table/Fig-2]. A mild disc-osteophyte complex was noted at the C5-C6 level, causing minimal thecal sac indentation without significant canal stenosis.



[Table/Fig-2]: MRI findings in Hirayama disease; a) Sagittal T2-weighted MRI of the cervical spine in the neutral position demonstrates a short-segment T2 hyperintensity within the cervical spinal cord at the C5-C6 level (blue arrow), without associated cord atrophy; b) Sagittal T2-weighted MRI of the cervical spine obtained in flexion shows anterior displacement of the posterior dura mater (arrowhead), resulting in effacement of the dorsal thecal sac and formation of a prominent posterior epidural space (white arrow), consistent with engorgement of the epidural venous plexus, characteristic of Hirayama disease.

Based on the combined clinical and radiological findings, a diagnosis of early-stage (pre-atrophic) Hirayama disease without muscle wasting was established. The patient was managed conservatively with avoidance of excessive cervical flexion, activity modification, and the use of a cervical collar. At 12-month clinical follow-up, the patient remained neurologically stable, with no progression of symptoms or development of distal upper-limb weakness, muscle atrophy, or other neurological deficits. Given the stable clinical course

and absence of new symptoms, a repeat MRI was not performed, and conservative management was continued.

DISCUSSION

The Hirayama disease (HD) is an uncommon form of cervical flexion myelopathy that predominantly affects adolescent and young adult males, typically between 15 and 25 years of age [1]. The classical clinical presentation includes insidious-onset, asymmetric distal upper-limb weakness, hand muscle atrophy, tremors, and cold paresis. The underlying pathophysiology is attributed to abnormal forward displacement of the posterior dura mater during neck flexion, resulting in repetitive dynamic compression of the lower cervical spinal cord, chronic microcirculatory compromise, and ischemic injury to the anterior horn cells [2].

In comparison with our patient, who presented with isolated neck pain and a normal neurological examination, the case series reported by Singh A et al., from a tertiary care centre in eastern India evaluated 13 patients with clinical suspicion of Hirayama disease on a 3-Tesla scanner. The age range in that series was 16-26 years, and 100% of the cohort demonstrated anterior shifting of the posterior dural sac, cord flattening, abnormal cervical curvature, and an enhancing epidural component on dynamic imaging. Notably, nine of those 13 patients had already developed localised cord atrophy and intramedullary T2 hyperintensities, placing them at a more advanced disease stage than the present case [3].

Kalekar T et al., reported 13 Indian patients. All patients demonstrated excessive forward shifting of the posterior dura on flexion, with resultant cord compression; however, 92% had already developed chronic myelomalacia with abnormal cord hyperintensity and atrophy [4]. Similarly, Sonowal TN et al., characterised dynamic MRI findings in patients from North-East India, reinforcing anterior dural displacement and epidural flow voids as characteristic features [5]. A more recent case series of 10 patients confirmed anterior displacement of the dural sac in all cases and posterior epidural venous plexus engorgement in nine, with conservative cervical collar therapy successful in eight of 10 patients [6].

In contrast to these series, in which overt amyotrophy was a consistent clinical finding, our patient demonstrated no detectable motor deficit or muscle wasting. Boruah DK et al., emphasised the importance of measuring the laminodural space during flexion cervical MRI to detect subtle disease, an approach that complements our diagnostic reasoning [7]. The present case is therefore unusual in that classic clinical hallmarks were absent, yet pathognomonic flexion-induced imaging changes were present.

The insidious nature of HD frequently mimics other conditions; Kumar S et al., reported two cases from India in which antecedent trauma initially misled clinicians, resulting in delayed recognition of the underlying myelopathy [8]. A mild C5-C6 disc–osteophyte complex was also identified on neutral MRI; however, it produced only minimal thecal sac indentation without significant spinal canal stenosis or static cord compression and was therefore considered an incidental age-related degenerative finding. Importantly, this lesion does not explain the characteristic dynamic anterior displacement of the posterior dura mater and posterior epidural venous plexus engorgement observed on flexion MRI, which are considered hallmark imaging features of Hirayama disease. Thus, the diagnosis was based on dynamic flexion-induced abnormalities rather than on incidental degenerative changes.

Conservative management with cervical collar immobilisation functions by mechanically limiting neck flexion, thereby preventing repeated dynamic compression of the lower cervical cord and mitigating the microcirculatory compromise that drives anterior horn cell injury. Tokumaru and Hirayama provided the seminal evidence supporting this approach in a series of 38 patients, demonstrating that cervical collar therapy can induce early arrest of disease progression, with improvement most likely in patients with shorter illness duration and mild cord atrophy in the neutral position [9]. Building on this Fu Y et al., documented the outcomes of collar therapy in 73 patients, noting that 77.8% wore the device for less than six months due to factors such as cosmetic concerns, inconvenience, and a perception that the illness had stabilised [10].

CONCLUSION(S)

The present case illustrates a rare pre-atrophic presentation of Hirayama disease in which characteristic radiological abnormalities precede neurological deficits. Dynamic flexion MRI is essential for early diagnosis and timely intervention, potentially preventing progression to irreversible anterior horn cell injury and long-term disability.

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