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Unmasking distal upper limb atrophy – Hirayama disease

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Abstract

Hirayama disease is an uncommon, non-progressive juvenile amyotrophy that predominantly affects young Asian males. It typically presents with insidious onset distal upper limb weakness and muscle wasting without sensory involvement. Early recognition is important, as timely diagnosis can prevent disease progression. We report the case of a 28-year-old male with gradually progressive weakness and wasting of the distal upper limb muscles, exacerbated by cold exposure, and without sensory deficits. Flexion magnetic resonance imaging revealed a long-segment posterior dural detachment extending from C5 to D3 with associated cervical cord compression. Electrophysiological studies demonstrated denervation changes with preserved sensory conduction. The patient was managed conservatively with a cervical collar, following which his clinical condition stabilized. This case highlights the clinical and radiological variability of Hirayama disease. It reinforces the importance of flexion MRI in establishing an early diagnosis and supports conservative, non-surgical management in clinically stable cases.

Keywords: Hirayama disease; monomelic amyotrophy; flexion MRI

INTRODUCTION

Hirayama disease is a rare flexion-induced cervical myelopathy primarily affecting young Asian males.[1] It presents with gradual distal upper limb weakness and wasting, unilaterally or asymmetrically bilateral. Preservation of the brachioradialis muscle produces the characteristic oblique amyotrophy involving the C7, C8, and T1 myotomes.[2] Flexion MRI is the diagnostic cornerstone, demonstrating anterior displacement of the posterior dura, cervical cord compression, and flattening.[3] Electromyography (EMG) demonstrates denervation changes consistent with anterior horn cell involvement, while nerve conduction studies (NCS) confirm preserved sensory conduction.[4] This case highlights radiological variability of Hirayama disease, emphasizing early diagnosis using flexion MRI.

CASE REPORT

A man aged 28 years came to the outpatient clinic with a complaint of gradually progressive weakness in his right upper limb resulting in difficulty in fine motor tasks like writing, buttoning clothes, and holding objects. This weakness aggravated with exposure to cold temperatures with gradual thinning of the right hand and forearm. He denied sensory disturbances, cranial nerve or truncal involvement, lower limb weakness, bowel or bladder dysfunction, or similar family history. On neurological examination, bilateral atrophy of the medial forearm musculature was seen; the brachioradialis muscle remained unaffected. Weakness of wrist flexion, extension, and intrinsic hand muscles was noted, more pronounced on the right, resulting in asymmetric

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involvement. Figure 1 illustrates the prominence of the brachioradialis muscle.

Reflexes were preserved, and cranial nerve, sensory, and lower limb examinations were normal. Cervical spine Magnetic Resonance Imaging (MRI) revealed posterior dural detachment with associated anteroposterior cord flattening extending from C5 to D3 segment. The flexion and sagittal view of the MRI of the cervical spine is illustrated in Figures 2 and 3.

EMG showed denervation potentials in the affected muscles and NCS showed absent responses in the right median and ulnar nerves; sensory conduction preserved. He was treated conservatively with a cervical collar. On further visits, he demonstrated clinical stability with no further progression of weakness and reported improvement in performing activities of daily living.

DISCUSSION

Hirayama disease is an uncommon non-progressive lower motor neuron disorder first described in Japan, with a prevalence estimated at 1 per 30,000 in Japanese surveys.[1] It is most prevalent in young Asian males aged 15–25 years and is thought to arise from repeated ischemic injury to the anterior horn cells triggered by neck flexion.[5] A mismatch between spinal column growth and dural sac expansion is believed to produce a narrowed dural canal. During neck flexion, the dura posteriorly becomes excessively taut and fails to accommodate to the elongated cervical canal. Consequently, the posterior dural wall shifts forward, compressing the spinal cord and compromising its microcirculation.[6]

While classically unilateral, bilateral asymmetric nature has been seen in roughly 10-30% of cases, often with one limb being dominantly affected.[7] This pattern may result from more extensive or bilateral compression during neck flexion. In our patient, both forearms showed comparable wasting, but the intrinsic hand muscles were more prominently wasted on the right, illustrating this asymmetric pattern. Sparing of the brachioradialis muscle bilaterally maintained the classical oblique amyotrophy sign. Cold paresis, defined as transient worsening of weakness upon exposure to cold temperatures, is a recognized manifestation, though it may complicate the differential diagnosis by mimicking neuromuscular junction disorders or multifocal motor myelopathy.[5] Its pathophysiology remains unclear, but may be related to impaired neuromuscular transmission or altered membrane excitability in compromised anterior horn cells.

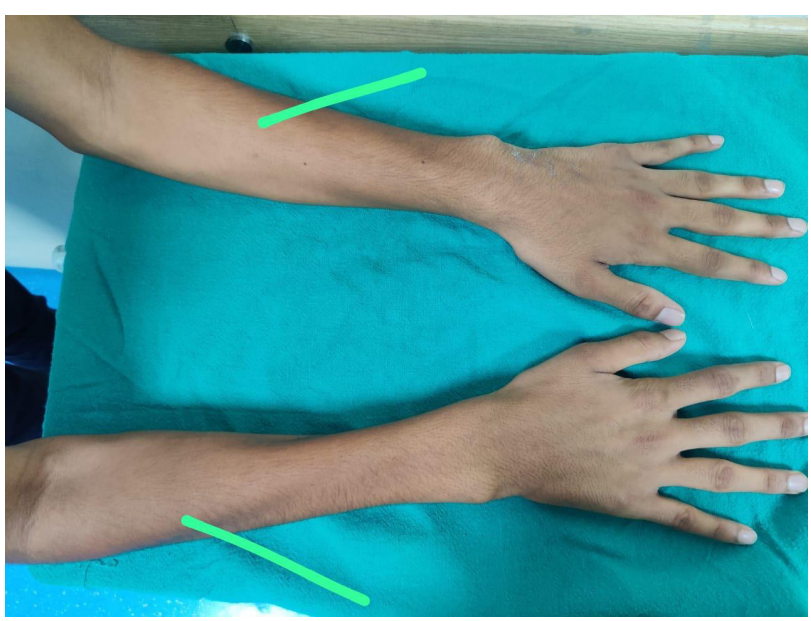

Figure 1. Atrophy of the muscles of the forearm except the brachioradialis muscle. The highlighted portion shows the sparing of the brachioradialis muscle in both forearms.

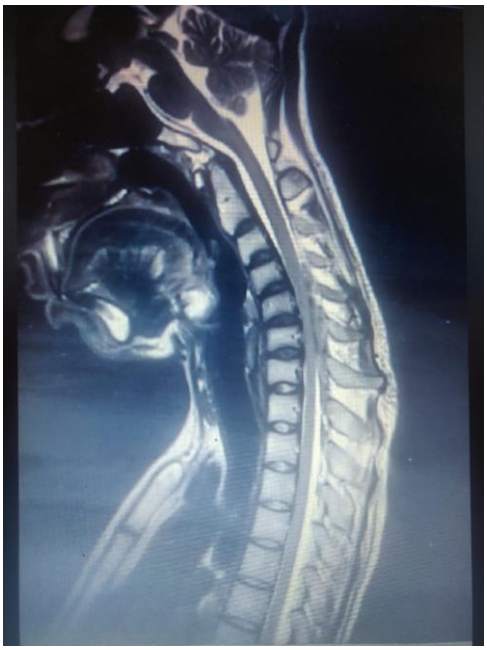

Figure 2. MRI cervical spine – flexion view

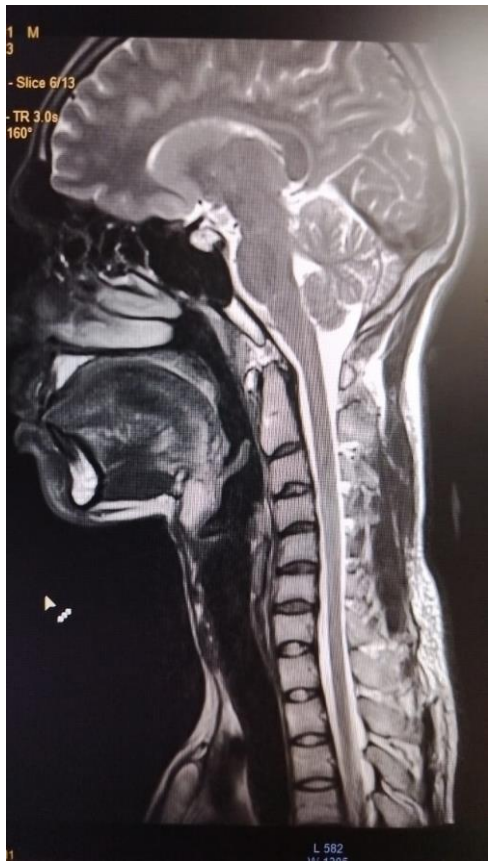

Figure 3. MRI cervical spine – sagittal view

MRI in the neutral position is often unremarkable but may show subtle findings such as lower cervical cord atrophy (commonly C5–C7), asymmetric pear-shaped cord flattening, loss of cervical lordosis, and posterior dural detachment from the laminae. In some patients, intramedullary T2 hyperintensity may also be observed, indicating chronic myelomalacia. Flexion MRI characteristically demonstrates shifting of the posterior dura anteriorly, cervical cord flattening, and a crescent shaped enhancing epidural space posteriorly, sometimes accompanied by epidural flow voids.[3] Boruah *et al*. reported a laminodural gap ranging from 3 to 9.8 mm during maximal dural shift, with a mean of 5.99 mm.[8]

Over the last few years, attempts have been made to streamline and standardize the diagnostic process for Hirayama disease. The Huashan diagnostic criteria, proposed in 2021, integrate clinical, imaging, and electrophysiological features to classify cases as definite or probable.[9] Applying these criteria, our patient fulfills the definition of definite Hirayama disease. A comparison of the Huashan criteria with the findings in this case is presented in Table 1.

In our patient, Flexion MRI demonstrated a relatively long segment of dural detachment and cord flattening from C5 to D3 an uncommon demonstration compared to the typical C5–C7 distribution [5,9]. Electrophysiological findings supported the diagnosis and excluded amyotrophic lateral sclerosis, distal hereditary motor neuropathies, and chronic inflammatory demyelinating neuropathy.

Given its general self-limiting nature, Hirayama disease is most commonly managed with a cervical collar to prevent further spinal cord compression. It restricts neck movement, prevents compression injury to the spinal cord and halts disease progression. Early use of this intervention, particularly during the initial 3–4 years of disease progression, has been recommended and is associated with favourable outcomes.[10] However, surgical options like anterior cervical discectomy and fusion (ACDF), posterior fixation, and laminoplasty could be recommended in situations where the disease progresses despite collar therapy.

In conclusion, this case underscores the clinical and radiological heterogeneity of Hirayama disease, including the less common long segment dural detachment and bilateral

Table 1: A comparison of the Huashan criteria with the findings in this case

Huashan diagnostic criteria (2021)	Findings in our case
Clinical manifestations	
Onset in adolescence/young adult, male predominance	28-year-old male
Insidious distal upper limb weakness and wasting, typically affecting hand/forearm muscles	Progressive wasting of intrinsic hand and medial forearm muscles
Unilateral or asymmetrically bilateral involvement	Bilateral involvement with more severe intrinsic hand muscle wasting on the right
Sparing of cranial nerves, lower limbs, and sphincters	No cranial, sensory, or sphincter involvement
Imaging manifestations	
Lower cervical cord atrophy or flattening; anterior shifting of posterior dura on flexion MRI	Cord flattening and loss of posterior dural attachment from C5–D3 on flexion MRI
Electrophysiological manifestations	
Neurogenic changes on EMG; preserved sensory conduction on NCS	EMG: denervation potentials; NCS: absent motor responses with preserved sensory conduction

muscle involvement. Early recognition through flexion MRI is critical to prevent diagnostic delays and initiate appropriate conservative intervention. The outcomes in this patient reinforce the efficacy of cervical collar therapy in stabilizing disease progression and improving function. Clinicians should maintain a high degree of suspicion for Hirayama disease in males of younger age presenting with wasting of upper limbs distally, especially in the presence of cold-induced paresis and characteristic imaging findings.

DISCLOSURE

Ethics: Approved by the Institutional Ethics Committee of ESIC Medical College & Hospital, Sanathnagar, Hyderabad (protocol code ESICMC/SNR/IEC-SO494/02-2025, date of approval: 25 February 2025). A full and detailed consent from the patient/guardian has been taken.

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Conflict of interest: None

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