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Unmasking Hirayama Disease with Dynamic 3.0 Tesla Flexion and Contrast-Enhanced MRI: A Case Report

Dr. Arpit Kushwaha[1], Dr. Harguneet Kaur[2], Dr. Madan Manmohan[3]

[1]Department of Radiodiagnosis, Dr. D. Y. Patil Medical College, Nerul, Navi Mumbai, India
Corresponding Author Email: *arpitkushwaha97[at]gmail.com*

[2]Department of Radiodiagnosis, Dr. D. Y. Patil Medical College, Nerul, Navi Mumbai, India
Email: *harguneetkaurgulati[at]gmail.com*

[3]Department of Radiodiagnosis, Dr. D. Y. Patil Medical College, Nerul, Navi Mumbai, India

Abstract: ***Hirayama disease is an uncommon, self-limiting flexion myelopathy that typically affects young males and may mimic motor neuron disease when routine neutral-position cervical MRI is non-diagnostic. We report a 21-year-old male who presented with one year of insidious difficulty in lifting weight with the left hand, with no sensory symptoms and no history of trauma or surgery. On clinical suspicion of a motor neuron disorder, riluzole had been started before MRI evaluation. Dynamic cervical spine MRI performed on a 3.0 Tesla scanner demonstrated subtle T2 hyperintensity and focal atrophy of the lower cervical cord on neutral imaging. Flexion MRI revealed anterior translation of the posterior dura, loss of dural attachment, development of an enlarged posterior epidural space measuring approximately 6.3 mm, bowstringing and compression of the lower cervical cord at C6-C7, and heterogeneous post-contrast enhancement of the posterior epidural space, representing engorged epidural venous plexus. The posterior epidural abnormality significantly reduced on extension, confirming its dynamic nature. This case highlights the value of dynamic flexion MRI, with contrast-enhanced imaging as a useful adjunct, in establishing the diagnosis and preventing misclassification as progressive motor neuron disease.***

Keywords: Hirayama disease; dynamic MRI; flexion myelopathy; posterior epidural venous plexus; cervical cord atrophy

1. Introduction

Hirayama disease, also known as juvenile muscular atrophy of the distal upper extremity, monomelic amyotrophy, or flexion myelopathy, is an uncommon, sporadic lower motor neuron disorder that predominantly affects adolescent and young adult males. It classically presents with insidious, asymmetric weakness and wasting of the distal upper limb, especially involving the forearm and intrinsic hand muscles. Sensory symptoms, pyramidal signs, and sphincter dysfunction are typically absent, which helps distinguish it from several compressive, inflammatory, and degenerative neurological disorders [1,2].

The clinical significance of Hirayama disease lies in its potential to mimic progressive motor neuron disease, cervical radiculopathy, syringomyelia, or brachial plexopathy. Conventional neutral-position MRI may be normal or may show only subtle lower cervical cord atrophy and intramedullary T2 hyperintensity. The pathognomonic abnormality is dynamic and becomes apparent during neck flexion, when the posterior dural sac shifts anteriorly, the posterior epidural venous plexus enlarges, and the lower cervical cord becomes compressed against the posterior vertebral bodies [3-5].

This case report highlights the role of dynamic flexion-extension MRI and contrast-enhanced imaging in confirming Hirayama disease in a young male who was initially suspected to have a motor neuron disorder.

2. Literature Survey

Hirayama and colleagues first described juvenile muscular atrophy of the unilateral upper extremity in 1963 [1]. Subsequent studies established that the disease is related to a mismatch between growth of the vertebral column and the dural canal, producing a relatively tight dural sac. During neck flexion, the shortened posterior dura cannot accommodate the increased length of the cervical canal and therefore shifts anteriorly. This produces repetitive compression of the lower cervical cord and chronic ischemic injury of anterior horn cells, particularly at C7-T1 myotomes [2,3].

MRI is central to diagnosis. Neutral-position findings may include loss of cervical lordosis, focal lower cervical cord atrophy, asymmetric cord flattening, and intramedullary T2 hyperintensity. Dynamic flexion MRI demonstrates anterior displacement of the posterior dural wall, loss of dural attachment from the lamina, crescentic enlargement of the posterior epidural space, flow voids or enhancement related to the epidural venous plexus, and reversible anterior bowstringing of the cord [3-5]. Contrast-enhanced images are useful in equivocal cases because the posterior epidural crescent enhances due to venous engorgement rather than solid tumor or abscess [5].

3. Problem Definition

Young patients with distal upper limb wasting are often investigated for motor neuron disease or peripheral nerve disorders. If cervical MRI is performed only in the neutral

position, Hirayama disease can be overlooked. Delayed recognition may lead to unnecessary anxiety, prolonged neurological work-up, and inappropriate therapeutic assumptions. The present case illustrates this diagnostic pitfall and demonstrates how dynamic MRI can convert a near-non-specific neutral study into a confident radiological diagnosis.

4. Methodology / Approach

Patient information and clinical history

A 21-year-old male presented to the neurology outpatient department with an approximately one-year history of insidious difficulty in lifting weight with the left hand. There was no associated tingling or numbness and no history of trauma or surgery. Clinical documentation described weakness of hand grip, and a trial of riluzole had been started on suspicion of a motor neuron disorder before MRI evaluation. Detailed neurological grading, reflex charting, and electrophysiological assessment were not available for inclusion in this report.

MRI technique

MRI of the cervical spine was performed on a 3.0 Tesla scanner. Neutral-position sagittal and axial sequences were followed by dynamic sagittal acquisitions in flexion and extension. Post-contrast T1-weighted images were obtained after intravenous gadolinium administration, including flexion post-contrast imaging to assess the posterior epidural space and venous plexus. Images used in the manuscript were anonymized.

5. Results and Discussion

Neutral-position MRI findings

Neutral-position MRI demonstrated reversal of cervical lordosis centered at C5-C6 with mild right-sided scoliosis. Small anterior osteophytes were seen from C3 to C7, and Modic type III endplate changes involved the postero-superior endplates of C4-C7. The spinal canal appeared mildly capacious, with an AP diameter of approximately 11.3 mm. No significant fixed cord compression was seen. However, subtle T2 hyperintensity was noted in the cord from C5 to C7, with reduction in the AP width of the cord to approximately 4.5 mm, suggestive of focal lower cervical cord atrophy.

Diffuse disc bulges at C5-C6 and C6-C7 indented the anterior subarachnoid space and mildly narrowed the neural foramina, without definite nerve root compression. Residual spinal canal diameters measured approximately 8.4 mm at C5-C6 and 8.8 mm at C6-C7 at the intervertebral disc levels. These degenerative findings did not explain the dynamic flexion-dependent cord compression pattern.

Dynamic flexion-extension and contrast-enhanced findings

On flexion, there was anterior translation of the dorsal dura with loss of attachment of the posterior dural sac from the subjacent lamina. A prominent posterior epidural space developed and measured approximately 6.3 mm in maximum AP width. This space appeared hyper- to isointense on T1-weighted and T2-weighted images. The lower cervical cord appeared bowstrung over the posterior aspect of the lower cervical vertebral bodies and was compressed by the enlarged posterior epidural space, predominantly at C6-C7, where the minimum cord width measured approximately 3.4 mm. On extension, the posterior epidural space became significantly thinned and poorly appreciable, confirming the dynamic nature of the abnormality.

On post-contrast flexion imaging, the posterior epidural space demonstrated heterogeneous enhancement, corresponding to engorgement of the posterior epidural venous plexus. The combination of anterior dural displacement, loss of dural attachment, dynamically enlarged enhancing posterior epidural space, reversible cord bowstringing, and lower cervical cord atrophy was diagnostic of Hirayama disease.

Table 1: Dynamic MRI observations and measurements

Parameter	Neutral position	Flexion / extension findings
Spinal canal AP diameter	Approximately 11.3 mm; mildly capacious	Not used as diagnostic dynamic parameter
Cord signal and caliber	Subtle T2 hyperintensity from C5-C7 with focal cord AP width approximately 4.5 mm	Cord bowstringing and compression at C6-C7; minimum width approximately 3.4 mm
Posterior epidural space	Not prominent on neutral imaging	Prominent on flexion; maximum AP width approximately 6.3 mm; significantly thinned on extension
Posterior dural attachment	No definite detachment	Loss of dural attachment with anterior translation of the posterior dura
Post-contrast behavior	No fixed enhancing epidural mass	Heterogeneous enhancement of posterior epidural space, consistent with engorged venous plexus

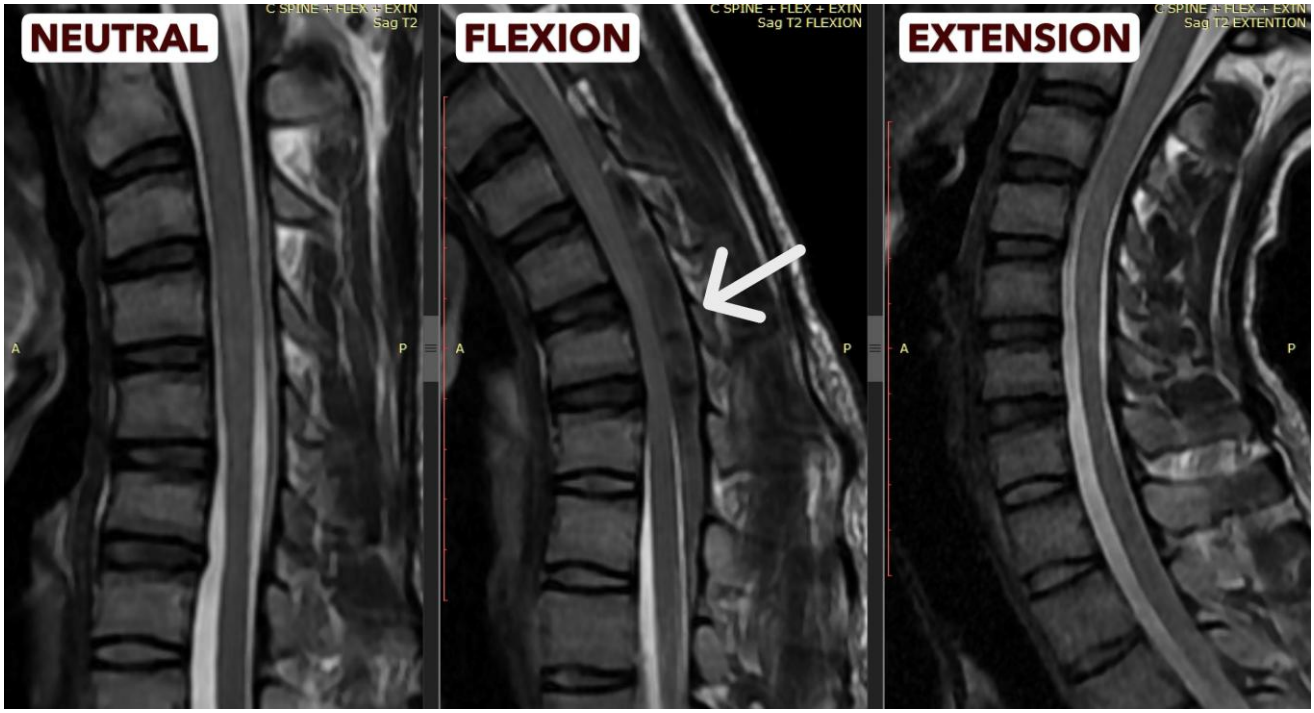

Figure 1: Sagittal T2-weighted neutral, flexion, and extension MRI. The flexion image demonstrates anterior displacement of the posterior dura, enlargement of the posterior epidural space, and anterior bowstringing of the lower cervical cord. The extension image shows reduction of the posterior epidural abnormality, confirming its dynamic nature.

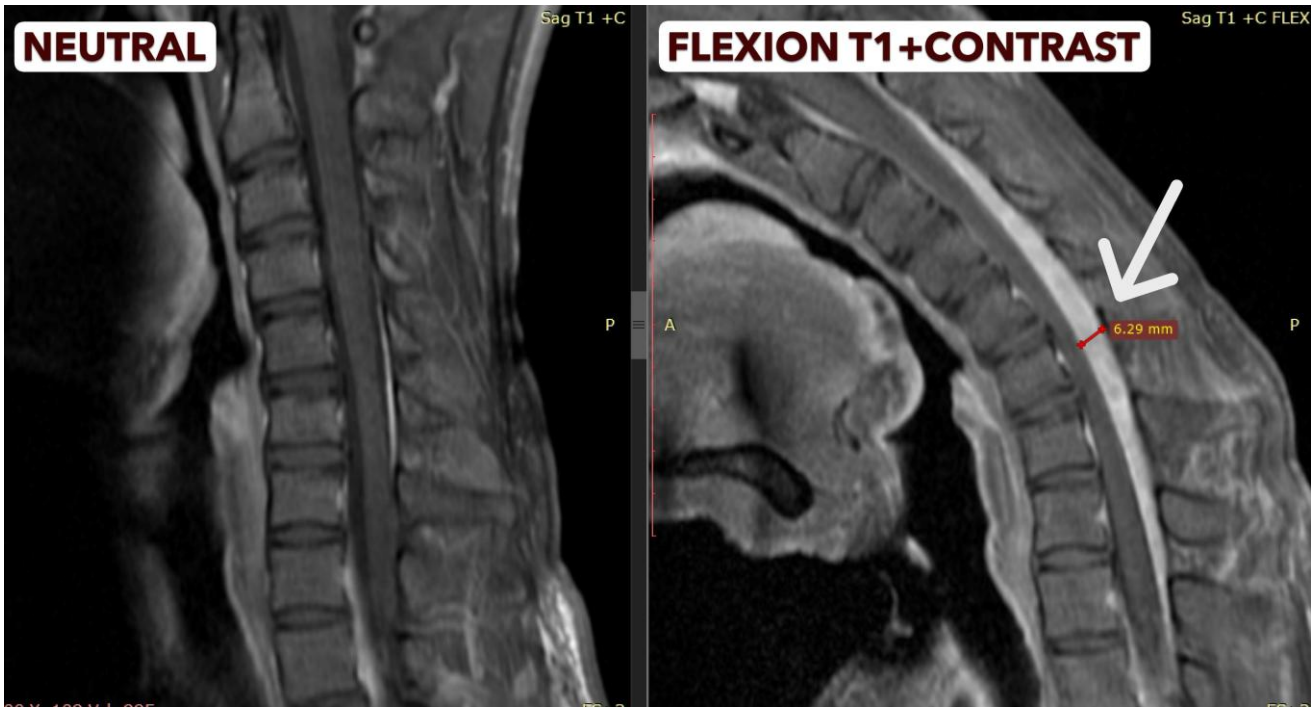

Figure 2: Sagittal post-contrast T1-weighted images in neutral and flexion. Flexion post-contrast imaging shows an enlarged posterior epidural space measuring approximately 6.3 mm, with heterogeneous enhancement representing engorged posterior epidural venous plexus.

Mechanism of disease

The pathophysiology is best explained by an imbalance between the growth of the vertebral column and the dural canal. During flexion, a relatively tight posterior dura shifts anteriorly and detaches from the lamina. The posterior epidural venous plexus becomes engorged, and the lower cervical cord is compressed against the posterior vertebral bodies. Repeated flexion-related compression is believed to produce chronic microcirculatory ischemia of anterior horn cells, resulting in focal lower motor neuron weakness and wasting [2-5].

Table 2: Important differential diagnoses and distinguishing features

Differential diagnosis	Why considered	Features favoring Hirayama disease in this case
Amyotrophic lateral sclerosis / motor neuron disease	Distal hand weakness and wasting in a young patient can raise concern for motor neuron disease.	Young age, focal unilateral distal upper limb involvement, absence of documented upper motor neuron or bulbar features, and pathognomonic dynamic MRI findings.
Syringomyelia	Can cause hand wasting due to anterior horn cell involvement.	No syrinx was seen; the dominant abnormality was flexion-dependent posterior epidural venous engorgement and reversible cord compression.
Compressive cervical radiculopathy	C5-C6 and C6-C7 disc bulges were present.	No definite nerve root compression; symptoms and MRI pattern were better explained by dynamic dural shift and cord bowstringing.
Multifocal motor neuropathy / distal spinal muscular atrophy	Can present with focal lower motor neuron weakness.	Dynamic cervical MRI demonstrated the structural mechanism typical of Hirayama disease; electrophysiology was unavailable.
Epidural mass or vascular malformation	Enhancing posterior epidural space may mimic other epidural pathology.	The epidural abnormality appeared only/enlarged during flexion and thinned on extension, favoring dynamic venous engorgement rather than a fixed lesion.

Diagnostic and management implications

This case demonstrates why neutral MRI alone may be insufficient. In the neutral position, the findings were limited to loss of lordosis, subtle cord signal abnormality, and focal cord atrophy. The diagnostic features became evident only on flexion imaging. The enhancing posterior epidural crescent on contrast-enhanced flexion images supported the venous nature of the lesion and helped distinguish it from a fixed epidural mass.

Management of Hirayama disease is usually conservative, particularly when diagnosed early. Avoidance of sustained neck flexion and use of a cervical collar are intended to prevent further flexion-induced cord injury. Surgical options such as anterior cervical fusion or duraplasty may be considered in selected progressive or refractory cases, but most patients are managed non-operatively [2,4,5]. In the present case, detailed post-imaging treatment records and longitudinal clinical or imaging follow-up were not available.

Strengths and limitations

The strength of this report is the clear demonstration of the complete dynamic imaging pattern of Hirayama disease on a 3.0 Tesla MRI study, including neutral, flexion, extension, and post-contrast flexion findings. The case also illustrates a real diagnostic pitfall, as a motor neuron disorder had been clinically suspected before dynamic MRI.

The limitations are the absence of electrophysiological confirmation, lack of detailed neurological grading, and unavailability of longitudinal follow-up. Electromyography and nerve conduction studies could have strengthened the exclusion of motor neuron disease and documented chronic denervation in C7-T1 myotomes. Nevertheless, the diagnosis is strongly supported by the compatible clinical profile and characteristic flexion-dependent MRI findings.

6. Conclusion

Hirayama disease should be considered in young males with insidious, asymmetric distal upper limb weakness and wasting, especially when sensory symptoms and upper motor neuron signs are absent. Neutral cervical MRI may show only subtle cord atrophy or signal change and can miss the diagnosis. Dynamic flexion MRI is the key investigation, demonstrating anterior displacement of the posterior dura, loss of dural attachment, posterior epidural venous engorgement, and reversible lower cervical cord bowstringing. Contrast-enhanced flexion MRI is a valuable adjunct because it confirms enhancement of the engorged epidural venous plexus. Early recognition prevents misdiagnosis as progressive motor neuron disease and supports timely conservative flexion-limiting management.

7. Future Scope

Future reporting of Hirayama disease should include standardized dynamic MRI protocols, axial flexion images where feasible, objective cord and epidural space measurements, electrophysiological correlation, and clinical follow-up after flexion-restricting treatment. Larger radiology-centered case series may help define practical imaging thresholds and improve early recognition of this underdiagnosed entity.

8. Learning Points

- Hirayama disease can mimic motor neuron disease when clinical presentation is limited to distal upper limb lower motor neuron weakness.
- Neutral cervical MRI may be near-normal or show only subtle lower cervical cord atrophy and T2 hyperintensity.
- Dynamic flexion MRI is essential for demonstrating anterior dural displacement, loss of dural attachment, posterior epidural space enlargement, and reversible cord bowstringing.
- Heterogeneous enhancement of the flexion-induced posterior epidural space supports engorged venous plexus rather than a fixed epidural mass.
- Early clinico-radiological diagnosis can redirect management toward neck-flexion restriction and avoid unnecessary labeling as progressive motor neuron disease.

Ethical Statement

Written informed consent was obtained for the use of de-identified clinical and imaging data for academic publication. All patient identifiers were removed from the manuscript images. Institutional ethical standards were followed.

Conflict of Interest
The authors declare that they have no conflict of interest.

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Author Contributions
All authors contributed to case conception, image interpretation, manuscript drafting, critical revision, and approval of the final version for submission.

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