VER
Vascular & Endovascular Review

Hirayama Disease: Early Detection Through Dynamic Cervical Mri In A Young Male

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ABSTRACT

Hirayama disease, also known as cervical flexion-induced myelopathy, is a rare self-limiting cervical myelopathy predominantly affecting young males. Early diagnosis is essential because timely cervical collar therapy can halt disease progression. We present a 17-year-old male with progressive hand weakness whose dynamic MRI revealed characteristic flexion-induced cervical cord compression. This case highlights the importance of flexion MRI in diagnosing Hirayama disease.

KEYWORDS: Flexion Position MRI (Diagnostic), Role of Dynamic MRI

How to Cite: Nelluri Krishna Sai, Rajendra V Mali,Santosh D Patil., (2025) Hirayama Disease: Early Detection Through Dynamic Cervical Mri In A Young Male, Vascular and Endovascular Review, Vol.8, No.17s, 213-216

INTRODUCTION

Hirayama disease (HD) is a benign focal lower motor neuron disorder first described by Keizo Hirayama in 1959. It is characterized by unilateral or asymmetric distal upper limb weakness, predominantly affecting C7–T1 myotomes. The disorder is associated with disproportionate growth of the spinal canal and dura during puberty, leading to anterior displacement of the posterior dura during neck flexion and subsequent spinal cord ischemia.

Dynamic cervical MRI remains the cornerstone of diagnosis.

Case Presentation

A 17-year-old male presented to the outpatient department of KLE's Dr Prabhakar Kore Hospital, Belagavi with:

- Progressive *clawing* of the right hand for 1 year
- Generalized weakness of the distal right upper limb
- No significant birth, developmental, or family history

Clinical Examination

- Marked wasting and weakness of intrinsic hand muscles
- Clawing deformity suggestive of C7–T1 involvement
- No sensory deficits
- No signs of upper motor neuron involvement
- Non-progressive course after the initial period of deterioration

A clinical suspicion of Hirayama disease led to dynamic cervical spine MRI (neutral, flexion, and extension).

Imaging Findings

Neutral Position MRI

- T2 hyperintensities in the spinal cord from C3 to C7
- Cord atrophy particularly involving anterior horn cell regions
- Loss of normal cervical lordosis

Flexion Position MRI (Diagnostic)

- Anterior displacement of the posterior dura at C4–C5, C5–C6, and C6–C7
- Engorged posterior epidural space forming a crescent-shaped high-signal region

- Compression and flattening of the cervical cord between the displaced dura posteriorly and vertebral bodies anteriorly

These findings are characteristic of Hirayama disease**.**

Extension MRI

- Capacities of the spinal canal restored
- No neural foraminal stenosis
- No abnormal cord compression

Discussion

Hirayama disease is a flexion-induced myelopathy caused by:

1. Forward shifting of the posterior dural sac during neck flexion
2. Resultant compression and ischemia of the anterior horn cells
3. Disproportionate growth of vertebral column vs. spinal canal during adolescence

The disease is typically self-limiting but can cause permanent disability if not recognized early.

Epidemiology

- Predominantly affects young males
- Presents mainly in the teens or early 20s
- Rare familial cases (but not considered hereditary)
- Male preponderance attributed to greater pubertal growth spurts

Role of Dynamic MRI

Dynamic (flexion) MRI is the single most crucial diagnostic tool.

Findings include:

- Forward dural shifting
- Engorged posterior venous plexus
- Crescent-shaped hyperintense epidural space
- Cord flattening
- Segmental T2 hyperintensities and atrophy (late stage)

These correlate strongly with electromyographic and clinical findings.

Genetic Insights

Recent studies suggest variants in CEP126 (KIAA1377) and C5orf42 genes may be associated.

Management

The mainstay of treatment is:

1. Cervical Collar Immobilization
   - Prevents neck flexion
   - Reduces repeated microtrauma
   - Helps halt disease progression in early stages
2. Physiotherapy
   - Strengthening exercises
   - Preventing contractures
3. Surgical Options (For Progressive Cases)
   - Anterior cervical fusion
   - Dural tenting
   - Laminoplasty

Our patient was advised cervical collar therapy and physiotherapy. As symptoms had plateaued, surgery was not indicated.

CONCLUSION

This case highlights the importance of recognizing Hirayama disease early, especially in young males presenting with progressive hand weakness. Dynamic flexion-extension MRI is essential for diagnosis and treatment planning. Early collar therapy can significantly slow or halt disease progression, preventing long-term disability.

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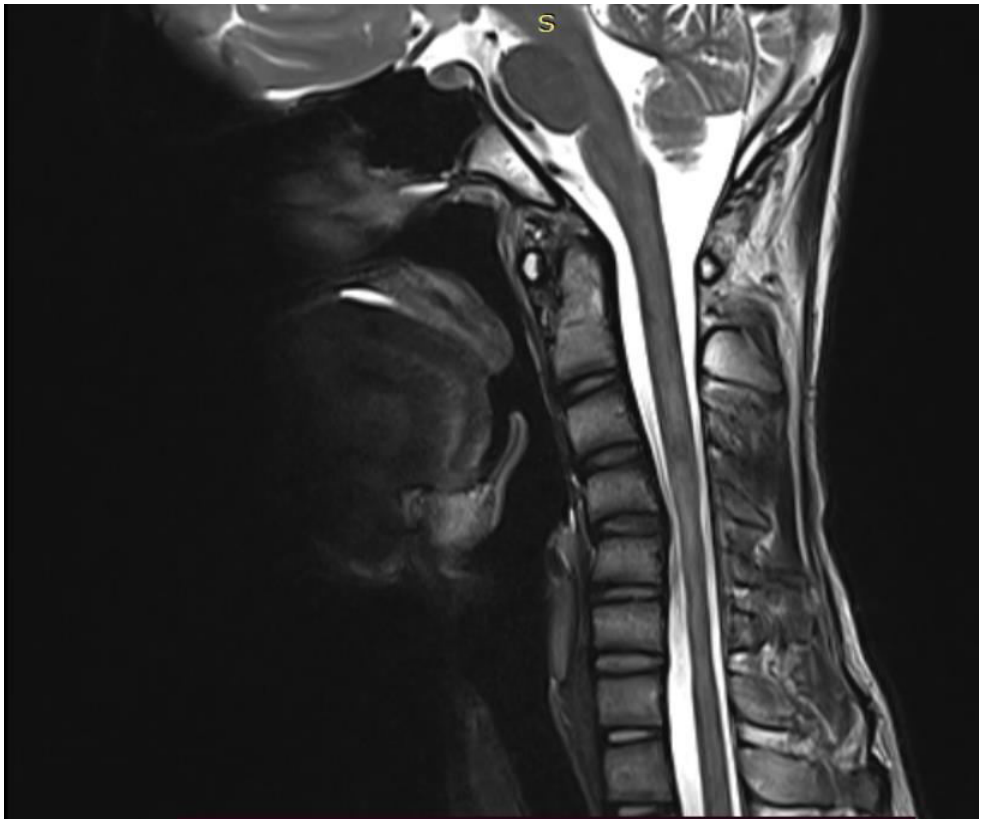
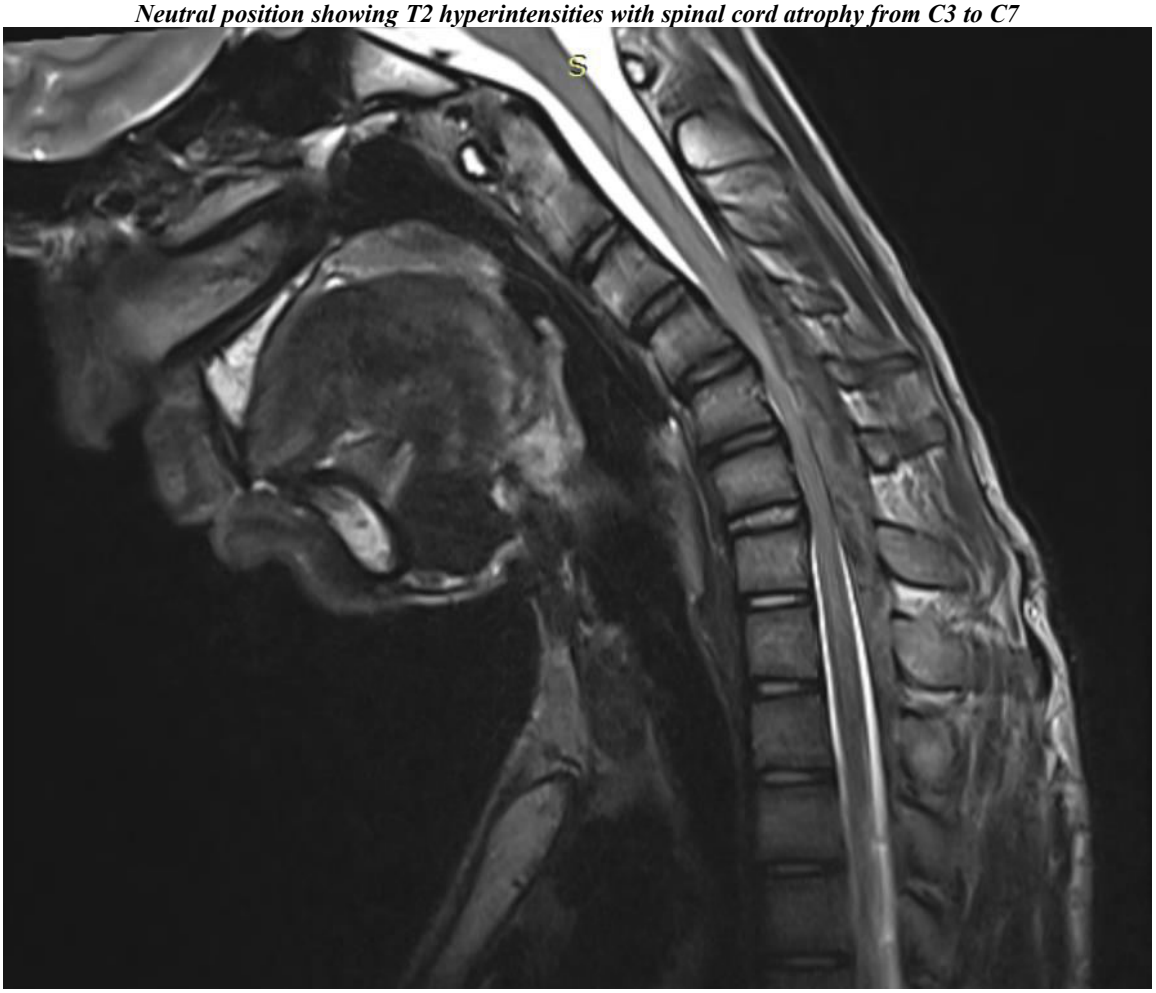
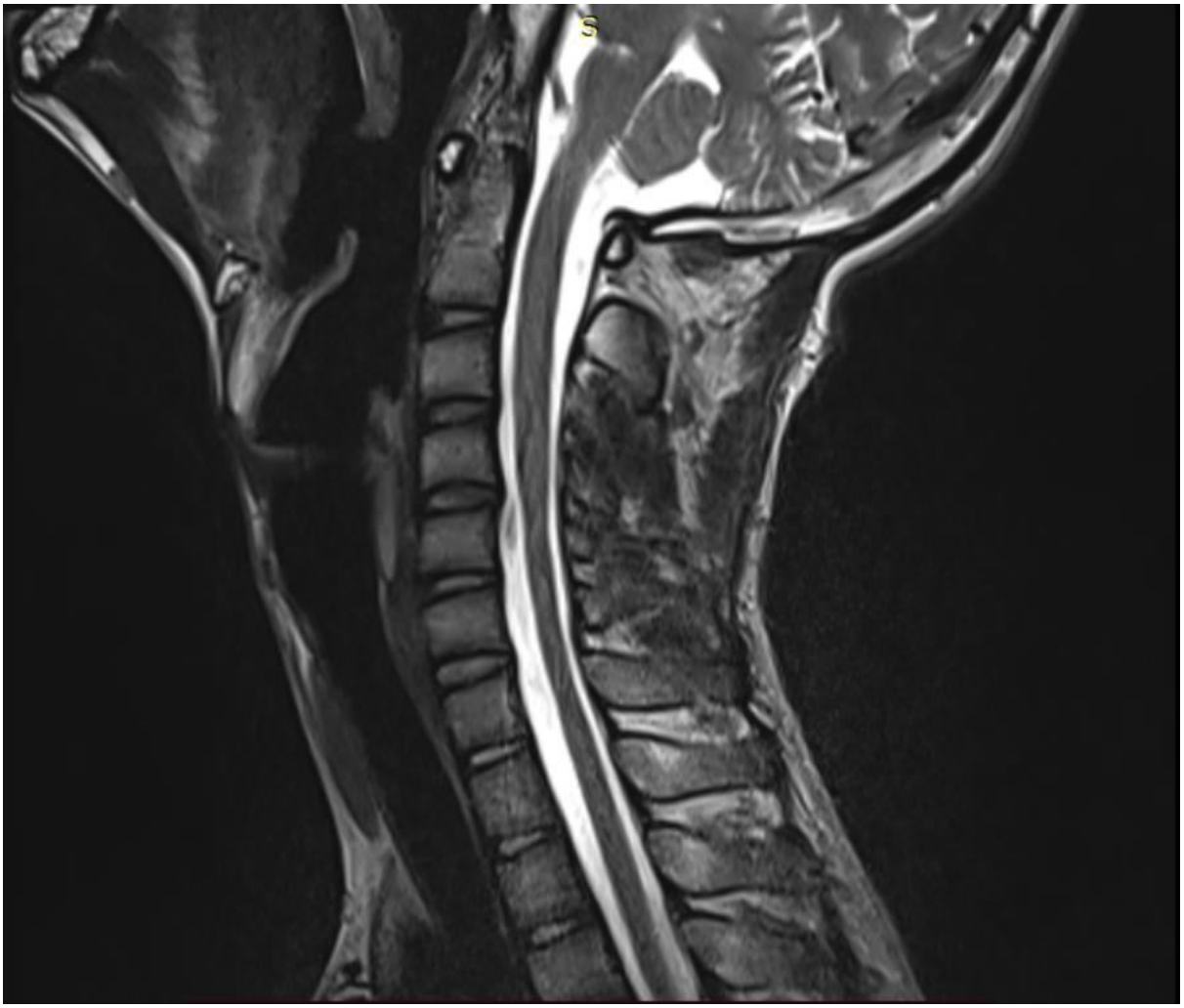


Neutral position showing T2 hyperintensities with spinal cord atrophy from C3 to C7



Flexion position showing anterior displacement of posterior theca compressing the cord anteriorly



Extension showing spacious canal