



## Case Report

# Hirayama disease – Early MRI diagnosis of subacute medullary ischemia: A case report

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## ABSTRACT

**Background:** Hirayama disease (HD) is a rare, benign, and self-limiting motor neuron disorder that results in selective motor impairment of the C7-T1 myotomes. It is characterized by progressive, unilateral, or bilateral asymmetric muscle atrophy of the distal upper extremities and myelopathy.

**Case Description:** A 23-year-old male presented with bilateral atrophy of the thenar/hypothenar eminences/intersosseous muscles, plus left-hand weakness. The cervical MRI documented subacute ischemic damage of the distal cervical cord. To rule out a tumor and reduce questionable cord compression, the patient underwent a C5–C6 anterior cervical discectomy and fusion (ACDF) immediately followed by a laminectomy with durotomy and to obtain a spinal cord biopsy. When the histology confirmed focal cord ischemia consistent with HD, it was clear that both operations were unnecessary.

**Conclusion:** Establishing the diagnosis of HD is based on clinical findings and MRI/flexion MR features which include the demonstration of an increased T2-weighted intramedullary cord signal, enlargement of the posterior epidural space, and segmental spinal cord atrophy. The presence of HD should be recognized as a “nonsurgical entity,” and conservative nonsurgical management should be employed.

**Keywords:** Amyotrophy, Hirayama disease, Magnetic resonance imaging

## INTRODUCTION

Hirayama disease (HD) is a rare, benign, and self-limiting motor neuron disorder that typically involves selective motor impairment in the C7-T1 myotomes (e.g., involving the hands and forearms and related to cervical flexion maneuvers). It is attributed to chronic spinal cord damage occurring during repeated cervical flexion maneuvers and is characterized by progressive subacute unilateral or bilateral distal upper extremity myelopathy (i.e. painless amyotrophy).<sup>[2]</sup> Dynamic cervical MRI studies show (1) a high intrinsic cord signal, (2) asymmetric cord flattening in the neutral position with enlargement of the posterior epidural space, and (3) spinal cord compression on flexion studies.<sup>[6]</sup> Here, we present a 23-year-old male with distal cervical HD who unnecessarily underwent a C5–C6 anterior cervical discectomy and fusion (ACDF) and laminectomy/duroplasty with cord biopsy.

## CASE REPORT

### Clinical presentation

A 23-year-old Italian male presented with a 2-year history of progressive left greater than the right-hand weakness/myelopathy. He exhibited left greater than the right-sided atrophy of the thenar/hypothenar eminences, and interosseous muscles. Motor nerve conduction/electromyographic studies showed left greater than the right-sided C7-T1 denervation, while sensory conduction studies were normal.

### MRI findings

The patient underwent both neutral and flexion. A 1.5 Tesla cervical magnetic resonance imaging (MRI) showing diffuse lower cervical spinal cord swelling (C4-C7) most severe at C5-C6 with mild spondylosis and a high intrinsic T2-weighted cord signal (e.g. with punctate foci of restricted diffusion) [Figures 1a-d and 2a]. A 30° flexion noncontrast MRI demonstrated posterior extradural space expansion due to an engorged epidural venous plexus, with anterior displacement at the C4-C6 level [Figure 2b].

The contrast the MR showed enhancement within the cord at C5-C6 level [Figure 1d and 2c].

### Surgery

The patient first underwent a C5-C6 ACDF to achieve anterior decompression. To exclude an intrinsic cord tumor, a cervical laminectomy with durotomy allowed for an intramedullary cord biopsy. The latter documented focal cord ischemia consistent with intrinsic HD.

### Follow-up

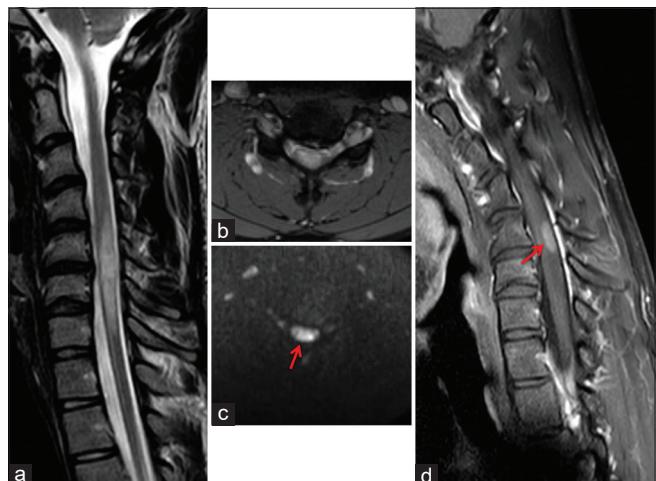
One year later, the cervical MRI demonstrated a reduction of the ischemic C5-C6 cord focus, and the patient clinically remained unchanged. Over the next 2 years, the subsequent MRI scans showed a mild progressive focal atrophy of the cord at the C5-C6 level with selective involvement of the anterior horns [Figure 3].

## DISCUSSION

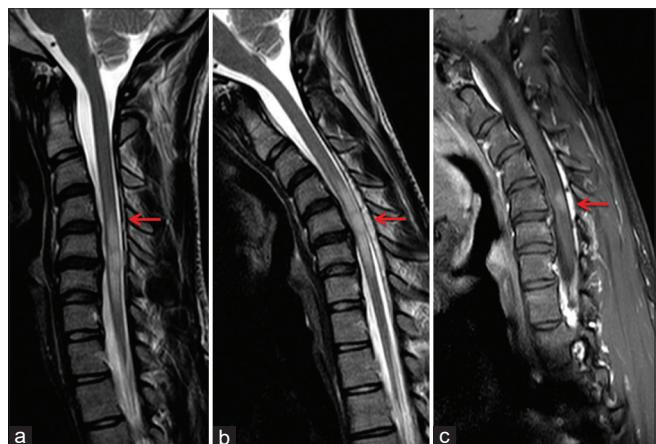
### Etiology

HD, also called “Juvenile muscular atrophy of the distal upper extremities,” is a rare myelopathy involving the lower cervical spine characterized by an insidious onset of atrophy involving the C7, C8, and T1 myotomes, with sensory sparing.<sup>[2,3]</sup>

HD may be attributed to: (1) a disproportionate growth between the spinal column and cord or (2) repeated neck flexion with microcirculatory changes involving the anterior spinal artery result in ischemia.<sup>[5]</sup>



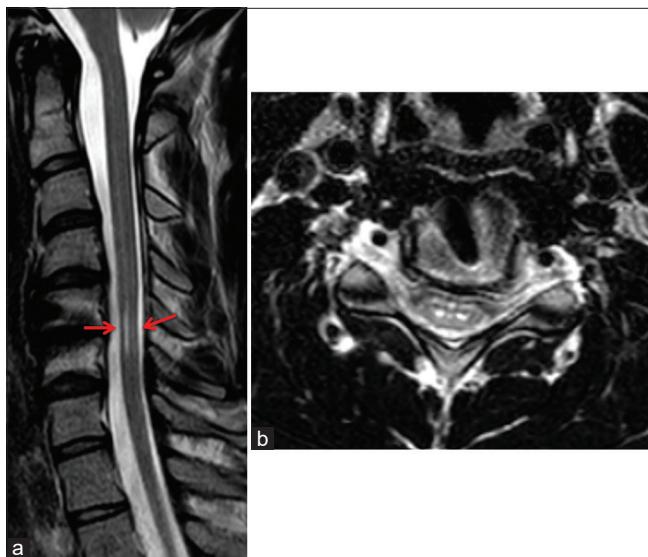
**Figure 1:** (a) Sagittal T2-weighted FSE magnetic resonance imaging of the cervical spine in neutral position. Segmental intramedullary swelling with high signal intensity at C5 to C7 level and loss of cervical lordosis. (b,c) Neutral position axial T2-weighted GRE and axial DWI sequence at C5-C6 disc level showing segmental spinal cord hyperintensity with some foci of restricted diffusion (arrow). A median-paramedian disc herniation at the same level was also evident. (d) Sagittal contrast-enhanced T1-weighted FSE acquisition on neck flexion showing a partial enhancement of the spinal cord lesion at C5-C6 level (arrow).



**Figure 2:** (a) Sagittal T2-weighted FSE image in neutral position with evidence of posterior detachment of dural sac (arrow). (b) Flexion sagittal T2-weighted FSE images confirming anterior displacement of the posterior dura from C4 to C7 levels with spinal cord flattening and prominence of the posterior epidural space (arrow). (c) Sagittal contrast-enhanced T1-weighted FSE in flexion position with evidence of enhancement of the enlarged posterior epidural space (arrow).

### MRI findings

Dynamic cervical MRI scans play a critical role in the diagnosis of HD. The neutral MRI usually shows (1) localized lower cervical cord atrophy/asymmetric cord flattening, (2) a high intrinsic cord signal on T2-weighted images, and (3) an



**Figure 3:** Two-year MRI follow-up after surgery on neutral position. (a) Sagittal T2-TSE-weighted image depicting segmental spinal cord atrophy at C5–C6 level (arrows). (b) Axial T2-GRE-weighted acquisition at C5 level with evidence of bilateral medullary hyperintensity of the anterior horns.

abnormal cervical curvature with loss of attachment between the posterior dural sac and subjacent lamina.<sup>[6]</sup> Flexion MRs (30–40°) show (1) anterior displacement of posterior wall of the cervical dural sac and (2) dorsal flattening of the lower cervical cord due to enlargement of the posterior epidural space attributed to a congested epidural venous plexus<sup>[6]</sup>. With gadolinium, there is a homogeneous enhancement of the epidural space<sup>[6]</sup>.

#### Etiology of ischemia

The etiology of spinal cord ischemia is potentially related to passive dilation of the posterior epidural venous plexus, contributing to chronic ischemic damage of the anterior horns<sup>[4]</sup>.

#### Surgery

There is no role for surgery in HD<sup>[7]</sup>. Only rarely, should anterior or posterior cervical surgery be considered to rule out the tumor and stem deterioration due to cord swelling.<sup>[1]</sup> Here, the C5–C6 ACDF and laminectomy, intramedullary cord biopsy, and duroplasty were unnecessary. Such unwarranted surgical intervention should be avoided by others who should immediately entertain HD among the differential diagnostic considerations.

## CONCLUSION

HD should be suspected in young males presenting with a slowly progressive myelopathy. Based on the MRI findings, the diagnosis of HD, a neurodegenerative disorder (i.e., intrinsic cord changes without significant extrinsic compression), should be considered averting unnecessary surgery.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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#### Conflicts of interest

There are no conflicts of interest.

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