Case Report

Magnetic resonance imaging features of “Proximal” hirayama disease: Case report and literature review

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Introduction

Proximal “Hirayama” disease (PHD) is characterized by proximal upper extremity atrophy. It is unlike classical Hirayama disease (HD) which involves the distal acral part of the upper limb. Magnetic resonance (MR) findings include posterior cervical epidural soft-tissue lesions resulting in cord compression/glialosis along with kyphosis. Here, we present a 17-year-old male with cervical PHD who refused surgery, and was successfully treated with a collar for 1 year.

Case Presentation

A 17-year-old adolescent male presented with 3 months of gradually progressive right upper limb weakness (3/5) and atrophy (i.e., involving the muscles of arm and forearm muscles) [Figure 1]. Lab studies were normal.

Electromyelography (EMG)

The needle EMG of the muscles of the right arm and forearm showed polyphasic motor unit action potentials with increased amplitudes and delayed latencies.
MR

The cervical MR showed cord compression, flattening, and atrophy with kyphosis from C2 to C6 maximum at C4-C5 apex [Figure 2a]. Axial T2W scans demonstrated hyperintense signals in the anterior horn cell region at C4 (characteristic "snake eyes" sign) [Figure 2b-d]. The sagittal T2W MR imaging (MRI) in flexion demonstrated a posterior epidural soft-tissue mass with prominent flow voids from C3 to C6 compressing/displacing the cord anteriorly; this mass markedly enhanced with contrast [Figures 3-5].

Diagnosis of PHD

Based on the clinical presentation and dynamic MRI findings, a diagnosis of PHD was established. The patient refused to undergo surgery. Therefore, he was managed with a cervical collar for 1 year during which time his deficit and MR findings stabilized.

DISCUSSION

HD is a form of cervical myelopathy that commonly occurs in young adolescents predominantly males (M: F = 3:1).\[2\] HD is often seen in Asians although a few cases have been observed in other regions. The disease most commonly affects the C7-T1 levels, and result in slow progression of a unilateral or asymmetric bilateral muscular amyotrophy. However, as seen in this case, HD may involve the C4 and C5 levels.\[4\]

MR findings of PHD

MRI is the preferred technique for diagnosis of HD.\[6\] Cervical MRI in the neutral position may show kyphosis and an atrophied/flattened lower cervical cord (i.e., the anterior

Figure 1: Photograph of the patient showing muscle wasting of the right proximal upper limb predominantly arm and forearm as compared to the left upper limb.

Figure 2: (a) Sagittal T2W magnetic resonance imaging (MRI) in neutral position image demonstrating loss of normal lordotic curvature with focal kyphotic angulation of the cervical spine with increased signal intensity in the cervical spinal cord at C4-C5 intervertebral level. Degeneration of intervertebral disks noted at multiple levels. (b) Axial MRI T2W image demonstrating typical snake eye hyperintense signals of the cervical spinal cord at C4-C5 level suggestive of subacute spinal cord ischemia. (c) Enlarged (zoomed in) Axial MRI T2W image demonstrating typical “snake eye” hyperintensity in the anterior horn cells of the spinal cord. (d) Diagrammatic representation of atrophied and gliosed anterior horn cells demonstrating snake eye appearance.

Figure 3: Sagittal T2W magnetic resonance imaging in flexion position demonstrating prominent posterior epidural soft-tissue component (arrow) displacing the posterior dural sac anteriorly and causing compression of the spinal cord with increased signal intensity(myelomalacia/subacute infarct) at C4 and C5 vertebral levels. Note: the apex of compression at C4 and C5 levels in the spinal canal causing maximum cord compression due to the kyphotic curvature.
Table 1: Differentiating imaging features between PHD and classical HD.

| Imaging Findings                              | Proximal type               | Classical distal or acral type |
|-----------------------------------------------|-----------------------------|-------------------------------|
|                                               | Present case report         | Paeng et al.⁷                  | Yukote et al.⁸                  | Tsuzuki et al.⁹                  |
| Level of cervical cord hyperintensity with cord flattening | C4–C5 level                | C4–C5 level                   | No cord signal abnormality      | C5–C6 and below                  |
| Level of extension of posterior epidural soft tissue | C3–C6 level                | C3–C6 level                   | C3–C6 level                     | C4–C6 level                      |
| Presence of predisposing factors:             | Loss of lordotic curvature | Focal kyphotic curvature at C4 and C5 levels | Loss of lordotic curvature | Loss of lordotic curvature at C4 and C5 levels |
| Loss of cervical lordosis                     | Focal kyphotic curvature at C4 and C5 levels | Focal kyphotic curvature      | Usually absent with only posterior epidural soft-tissue component as predominant feature if present kyphosis may be present below C7 vertebral level |
| Cervical spine kyphotic deformity             | Unstable cervical spine at C4 and C5 vertebra |                      |
| Cervical spine instability or other associated risk factors |                      |                              |

horn cells) and increased signal in the spinal cord reflecting gliosis (i.e., best seen on axial T2W MRI). The “snake eyes sign” on MRI is a poor prognostic finding.¹⁰ Dynamic flexion cervical MRI (i.e., without and with contrast) should show a pathological soft-tissue “lesion” in posterior epidural space (i.e., an engorged epidural venous plexus) resulting in dorsal

![Figure 4](image1.png)

**Figure 4:** Pre contrast T1W magnetic resonance (MR) sagittal (a) and axial image (b) in flexion position demonstrating prominent posterior epidural space displacing the posterior dural sac anteriorly and causing compression of the spinal cord. Post contrast T1W MR sagittal (c) and axial image (d) in flexion demonstrating homogeneous enhancement in the posterior epidural space at C3–C6 vertebral levels (Arrows).

![Figure 5](image2.png)

**Figure 5:** Post contrast T1W sagittal magnetic resonance imaging image (a) in flexion position demonstrating the characteristic crescent shaped posterior epidural enhancement at C3–C6 vertebral levels. (b) Represents graphical outline of crescent shaped enhancement.
cord compression/ventral displacement at the lower cervical levels. This lesion shows moderate contrast enhancement and the posterior “crescent” sign. On MR imaging, PHD demonstrates characteristic imaging features as compared to the classical form of the disease [Table 1].

Treatment

The treatment for PHD is cervical decompression with fusion to prevent progression. However, in select cases on patient's refusal for surgery, cervical collar prevents further progression of the symptoms.

CONCLUSION

PHD a rare variant of HD is characterized by proximal upper extremity atrophy. Its unique clinical and MR findings help differentiate it from classical HD.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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