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Case Report

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

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ABSTRACT

Background: Proximal "Hirayama" disease (PHD) is characterized by proximal upper extremity atrophy. It is a rare variant of Hirayama disease (HD) which involves the proximal upper limb. Recognition of PHD's unique magnetic resonance (MR) findings is critical as the treatment options differ versus classical HD.

Case Description: A 17-year-old male presented with gradual progressive upper extremity weakness and atrophy. On MR, PHD was demonstrated by C4-C5 kyphosis with a posterior epidural soft-tissue mass compressing the C4-C5 cord resulting in gliosis. As the patient declined surgery, he was followed for 1 year with a cervical collar during which time his deficit stabilized.

Conclusion: PHD, characterized by proximal upper extremity weakness and atrophy, has characteristic MR findings of kyphosis associated with cord compression and ischemia/gliosis. Select patients as the one we described who decline surgery may stabilize radiographically and clinically with the protracted utilization of a cervical collar.

Keywords: Epidural soft tissue, Flexion magnetic resonance imaging, Hirayama disease, Magnetic resonance imaging, Proximal type Hirayama disease

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/gliosis along with kyphosis.[1] 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.

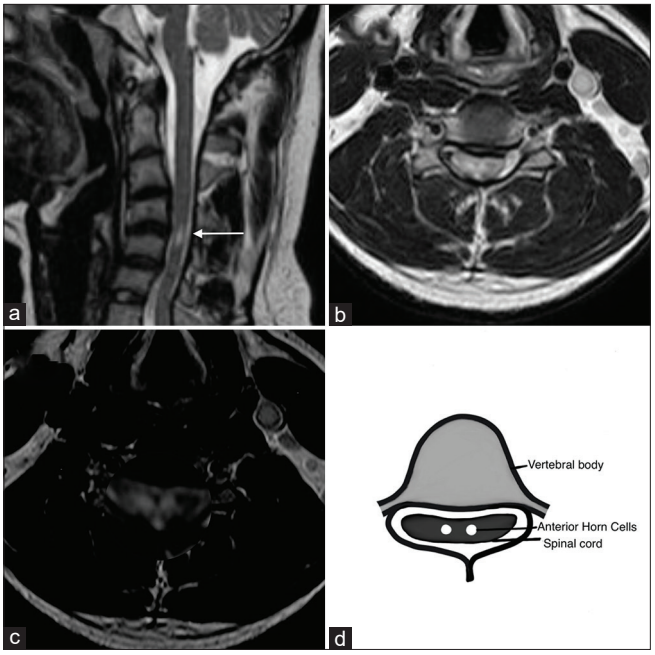

Figure 2: (a) Sagittal T2W magnetic resonance imaging (MRI) in neutral position image demonstrating loss of normal lordotic curvature with focal kyphotic angulation of the cervical spine with increased signal intensity in the cervical spinal cord at C4-C5 intervertebral level. Degeneration of intervertebral 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.

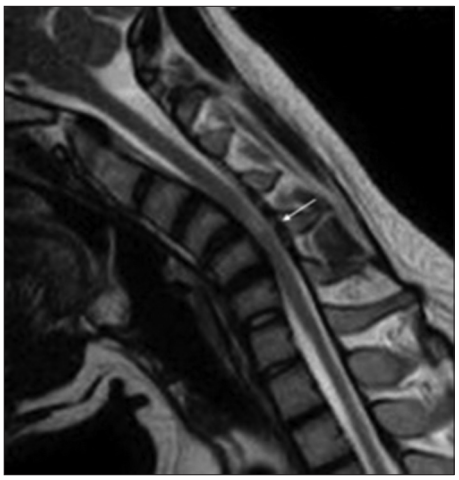

Figure 3: Sagittal T2W magnetic resonance imaging in flexion position demonstrating prominent posterior epidural soft-tissue component (arrow) displacing the posterior dural sac anteriorly and causing compression of the spinal cord with increased signal intensity(myelomalacia/subacute infarct) at C4 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	Hirayama disease type				
	Proximal type				Classical distal or acral type
	Present case report	Paeng *et al.*[7]	Yokote *et al.*[11]	Tsuzuki *et al.*[9]	
Level of cervical cord hyperintensity with cord flattening	C4–C5 level	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	C6 and may extend caudally to dorsal spine level usually long segment and greater thickness. Rarely may extend from C3 level however maximum diameter below C5 level
Presence of predisposing factors: Loss of cervical lordosis Cervical spine kyphotic deformity Cervical spine instability or other associated risk factors	Loss of lordotic curvature Focal kyphotic curvature at C4 and C5 levels Unstable cervical spine at C4 and C5 vertebra	Loss of lordotic curvature	Focal kyphotic curvature at C4 and C5 levels	Loss of lordotic curvature 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

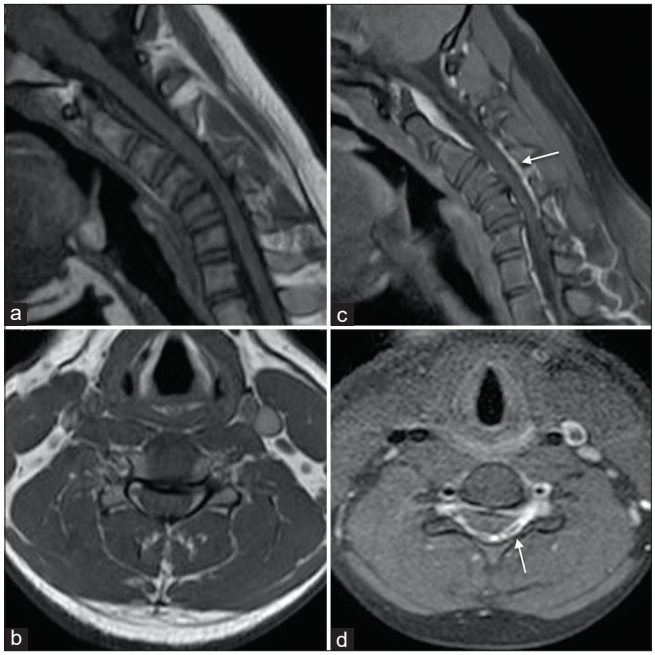

Figure 4: Pre contrast T1W magnetic resonance (MR) sagittal (a) and axial image (b) in flexion position demonstrating prominent posterior epidural space displacing the posterior dural sac anteriorly and causing compression of the spinal cord. Post contrast T1W MR sagittal (c) and axial image (d) in flexion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

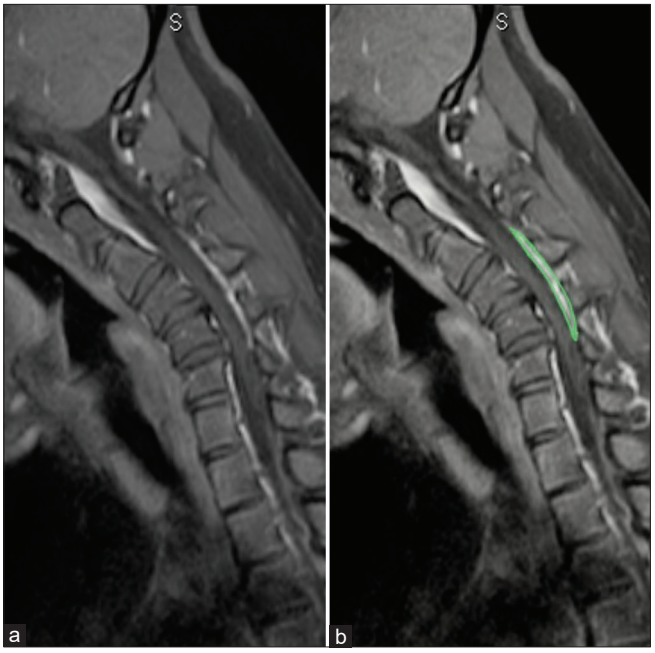

Figure 5: Post contrast TIW sagittal magnetic resonance imaging image (a) in flexion position demonstrating the characteristic crescent shaped posterior epidural enhancement at C3–C6 vertebral levels. (b) Represents graphical outline of crescent shaped enhancement.

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

cord compression/ventral displacement at the lower cervical levels.[5] This lesion shows moderate contrast enhancement and the posterior "crescent" sign.[8] 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.[3] 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.

REFERENCES

1. Hirayama K, Tokumaru Y. Cervical dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. Neurology 2000;54:1922-26.
2. Hirayama K. Juvenile muscular atrophy of unilateral upper extremity (Hirayama disease) Half-century progress and establishment since its discovery. Brain Nerve 2008;60:17-29.
3. Imamura H, Matsumoto S, Hayase M, Oda Y, Kikuchi H, Takano M. A case of Hirayama's disease successfully treated by anterior cervical decompression and fusion. No To Shinkei 2001;53:1033-8.
4. Kim J, Kim Y, Kim S, Oh K. Hirayama disease with proximal involvement. J Korean Med Sci 2016;31:1664-7.
5. Lai V, Wong YC, Poon WL, Yuen MK, Fu YP, Wong OW. Forward shifting of posterior dural sac during flexion cervical magnetic resonance imaging in Hirayama disease: An initial study on normal subjects compared to patients with Hirayama disease. Eur J Radiol 2011;80:724-8.
6. Lehman VT, Luetmer PH, Sorenson EJ, Carter RE, Gupta V, Fletcher GP, *et al*. Cervical spine MR imaging findings of patients with Hirayama disease in North America: A multisite study. Am J Neuroradiol 2013;34:451-6.
7. Paeng SH, kim YJ, Oh SI, Bae JS. Predominant proximal upper extremity involvement in Hirayama disease. Neurol Asia 2015;20:301-3.
8. Prior DE, Ghosh PS. Neuroimage: The crescent moon sign of Hirayama disease. Acta Neurol Belg 2021. Doi: 10.1007/s13760-020-01565-2. Epub Ahead of Print.
9. Tsuzuki U, Ando T, Sugiura M, Kawakami O. A case of proximal-type Hirayama disease associated with neck axial rotation. Rinsho Shinkeigaku 2021;61:120-6.
10. Xu H, Shao M, Zhang F, Nie C, Wang H, Zhu W, *et al*. Snake-eyes appearance on MRI occurs during the late stage of Hirayama disease and indicates poor prognosis. Biomed Res Int 2019;2019:9830243.
11. Yokote A, Fukuhara K, Tsugawa J, Tsuboi Y. Juvenile muscular atrophy of the proximal upper extremity as so-called proximal-type Hirayama disease: Case report and review of the literature. Case Rep Neurol 2019;11:106-11.

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