

Magnetic resonance imaging in Hirayama Disease

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ABSTRACT

Hirayama disease (HD) is a rare type of cervical myelopathy related to flexion of the neck characterized by progressive muscular weakness and atrophy of the distal upper limbs most frequently seen in young males. HD is thought to be secondary to an abnormal anterior displacement of the posterior dura with secondary compression of the lower cervical spinal cord and chronic injury to the anterior gray matter horns. We present two patients with HD and discuss its pathophysiology and imaging characteristics.

CASE REPORT

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Case 1

A 21-year-old man presented with 1- year history of gradual loss of muscle bulk and weakness in his right hand. Recently, he had noted increased weakness of the right hand extending to the forearm. He denied any other neurological symptoms. Physical examination showed significant muscle loss of his right upper limb involving all muscle groups except for the deltoid and brachioradialis muscles. Strength was reduced in the finger extensors, abductor digit mini, flexor pollicis longus, abductor pollicis brevis, pronator teres and ulnar innervated muscles. Strength in the left upper and lower limbs was normal. Sensory examination was normal. He had a nerve conduction study and EMG with normal sensory responses and decreased median motor responses with denervation in median, radial and ulnar muscles, all findings right sided. There was no evidence of conduction block. Neutral position cervical sagittal MR images revealed asymmetric spinal cord flattening and atrophy at C6-7 (Figure 1). In flexion, there was anterior displacement of the posterior dura with flow voids in the epidural space that corresponded to venous engorgement, as well secondary cord compression caused by this abnormal displacement. (Figure 2).

Case 2

A 33-year-old man presented with a longer than 10-year history of weakness and atrophy of the right upper limb. He denied sensory symptoms or weakness in other limbs. Neurological examination showed marked atrophy and fasciculations in his median and ulnar innervated muscles with weakness of the upper limb involving the finger extensors, finger flexors, ulnar innervated muscles and flexor pollicis longus. Vibration and position sense was normal. EMG showed no conduction block. MR study showed that on neutral neck position the spinal cord was atrophied at C6-7 with asymmetrical (right > left) flattening of its anterior surface (Figure 3) associated with degenerative changes at these levels. On flexion images there was posterior epidural space enlargement and cord compression. (Figure 4).

DISCUSSION

Hirayama disease, also called nonprogressive juvenile spinal muscular atrophy of the distal upper limbs or brachial monomelic amyotrophy, was described by Hirayama in 1959 [1] with approximately 200 cases reported in the literature. This disease is considered a pure motor focal amyotrophy caused by dynamic compression of the spinal cord leading to

atrophy and weakness in the distribution of C7, C8, and T1 spinal segmental innervated muscles.

HD has characteristic clinical features that include progressive muscular weakness of one or both hands and forearms without compromise of brachioradialis muscles and for this reason it is also called oblique amyotrophy [2]. Its progressive course lasts 3-5 years ending in a second stationary stage. HD usually occurs in young males between 14 - 38 years old with a mean age of onset of 16 years and a male: female ratio of 7:1 [2]. Clinical involvement in almost all patients is unilateral, predominantly in the right upper limb, although bilateral compromise has also been reported [3].

There is debate about the mechanism and origin of this disease. Normally, the spinal dura mater is a loose sheath around the cord attached to the foramen magnum, C2-C3, and posteriorly to the longitudinal ligament by fibrous bands. The dural sac is separated from the bony vertebral canal by the epidural space which contains a plexus of veins and loose areolar tissue [4]. Normally, the dural sheath is larger than what is needed for its contents. Its size is greater in the cervical and lumbar regions allowing it to have some loose folds. During neck extension and during flexion it stretches due to the increased length of the spinal cervical canal [5]. In normal flexion there is no significant displacement of the posterior dural wall or compression of the spinal cord due to its posterior attachments and relatively increased length. In HD, during neck flexion there is abnormal anterior displacement of the posterior dural wall with secondary compression of the cord. Kikuchi et al postulated that the mechanism underlying this finding is an abnormal tightening of the dural sheath secondary to an imbalance caused by relative shortening of the dura compared to the length of the spinal canal. [6] This relative shortening is explained by a disproportional growth between the vertebral column (larger) and its contents (smaller) during growth spurts, a fact that would explain the relatively young age of presentation in most patients and the male preponderance as during adolescence they undergo a faster increase in height than females. [6]. Other mechanisms proposed to explain the abnormal posterior dural wall displacement include the association of HD to thickening and alteration in the number of elastic fibers in the dural sheath [7] [8]. This may lead to an inelastic dura that is relatively short resulting in a similar dynamic mechanism as described above. Others propose that there may be a lack of posterior cervical epidural ligaments that results in loss of anchoring between the posterior dura and the ligamentum flavum allowing for abnormal anterior displacement of the dura. [9]

Regardless of the underlying cause, the anterior shift of the posterior dural wall causes spinal cord compression and possible chronic ischemia of the anterior spinal gray matter due to repeated compression of the anterior spinal artery resulting in a myelopathy [10]. There are few case reports that suggest that focal spinal cord degeneration may be also related to atopic myelitis with increased levels of immunoglobulin E and superoxide dismutase 1 mutations in familiar cases. [11-12]

Conventional radiological studies in HD show abnormal alignment of the cervical spine or may be unremarkable [5]. CT myelography in neutral position demonstrates asymmetric flattening of the anterior spinal cord with a normal oval shaped dural sac. In flexion, CT myelography shows anterior displacement of the posterior dural sac with diminished anteroposterior diameter and compression of the cord [13].

MR studies in neutral position show asymmetric flattening of the anterior aspect of cord, especially at C6 with varying degrees of cord atrophy at C6, C7 and T1 as well as high T2 signal intensity in the anterior cord due to myelomalacia. It has been suggested that loss of attachment between the posterior dura and subjacent lamina of more than one-third of the length of the lamina is a reliable finding for the diagnosis of HD on neutral position axial MR images [14].

Flexion MR shows anterior displacement of the posterior dura with an enlarged epidural space seen as a crescent high T1 and T2 intensity posteriorly on axial images. Inside this enlarged posterior epidural space there are flow voids and prominent enhancement on postcontrast images within these spaces. Flow voids correspond to enlarged posterior veins which some authors believe to be secondary to negative pressure in the posterior spinal canal and diminished jugular venous return, however Patel et al described lack of changes in epidural venous pressure measurement from neutral to flexion positions suggesting that venous engorgement is a passive process without decreased or increased venous pressure [15]. This finding explains why these flow voids are not seen on neutral position. On flexion MR studies, enlargement of the posterior dural space is associated with compressive flattening of the spinal cord and high T2 signal intensity in its anterior aspect. It is important to keep in mind that almost a one half of normal subjects may show posterior epidural space enlargement without cord compression and this is a normal finding that should not be confused with HD [16]. Postcontrast MR images depict nicely the epidural engorged venous plexus but otherwise are not necessary for diagnosis.

Morphologic changes on MR images correlate well with clinical and electromyography data [17]. Medical treatment in HD patients consists of a cervical collar for 3- 4 years to avoid compressive myelopathy, whereas surgery may be indicated for patients with constant or progressive neurological deficits resulting from spinal cord compression [18].

TEACHING POINT

Hirayama disease (HD) should be suspected in young male patients with a chronic history of weakness and atrophy involving the upper extremities. MR imaging is the best way to make the diagnosis but it necessitates the use of both extension/flexion and post contrast studies. HD requires a long period of conservative treatment which impacts lifestyle.

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FIGURES

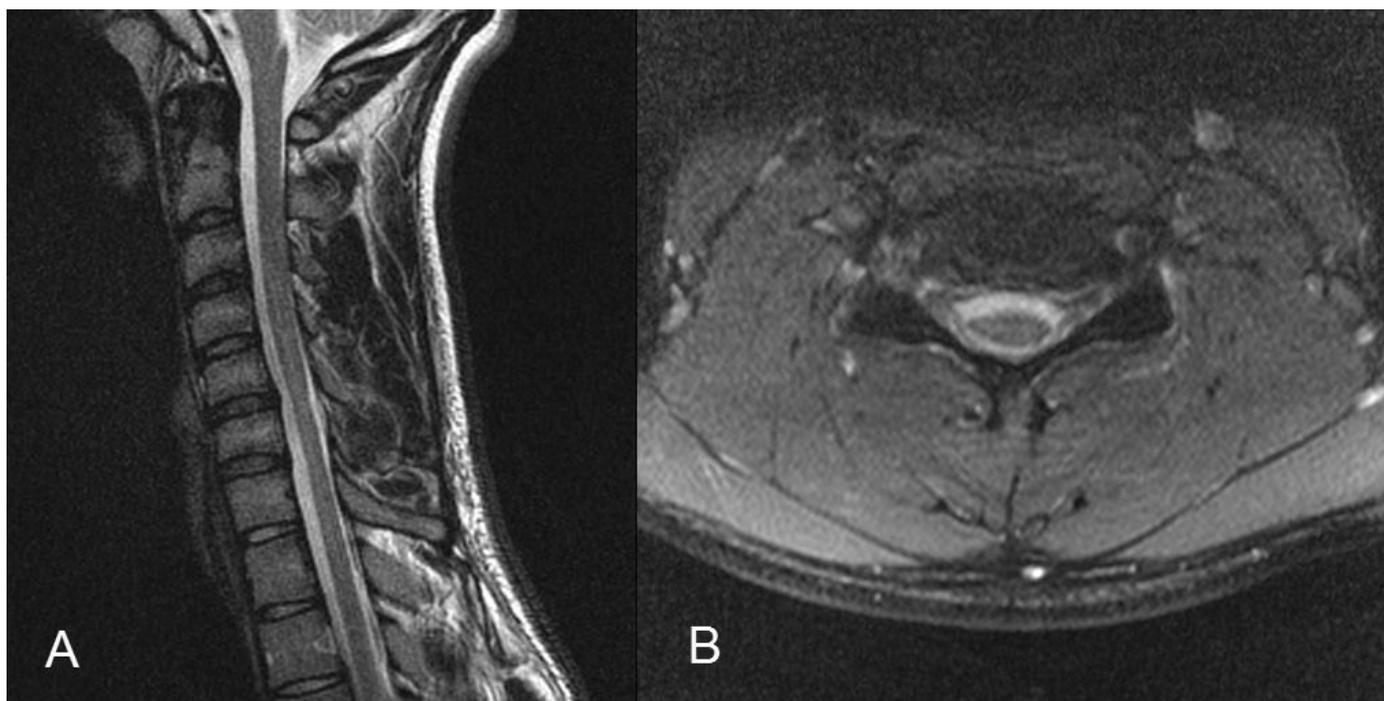


Figure 1. 21 year old male with progressive right upper limb atrophy secondary to Hirayama disease. Neutral neck position MR images. Sagittal T2WI (Parameters- TR: 4000ms, TE: 115ms, Slice thickness: 3 mm, Field strength: 1,5T) (a) shows straightening of cervical curvature with localized spinal cord atrophy at C6 and C7. Axial T2WI obtained at C6 (b) exhibits asymmetric (right > left) anterior spinal cord flattening at C6-7. (Parameters- TR: 4000ms, TE: 115ms, Slice thickness: 3 mm, Field strength: 1,5T)



Figure 2. 21 year old male with progressive right upper limb atrophy and diagnosis of Hirayama disease. Flexion sagittal T2WI (a) (Parameters- TR: 4000ms, TE: 115ms, Slice thickness: 3 mm, Field strength: 1,5T) and postcontrast sagittal with fat suppression T1WI (b) (Parameters- TR: 647ms, TE: 12ms, Slice thickness: 4 mm, Field strength: 1,5T, fat suppression, gadolinium dose 0.1 mmol/kg) and axial T1WI without fat suppression obtained at C6 (c) (Parameters- TR: 647ms, TE: 12ms, Slice thickness: 4 mm, Field strength: 1,5T, gadolinium dose 0.1 mmol/kg) show forward displacement of the posterior dural wall, more prominent at C6, with widening of the posterior epidural space which contains flow voids (a). Note homogeneous enhancement on postcontrast images (b). Secondary cord compression is seen associated to high T2 signal intensity (a) of the spinal cord at same level.

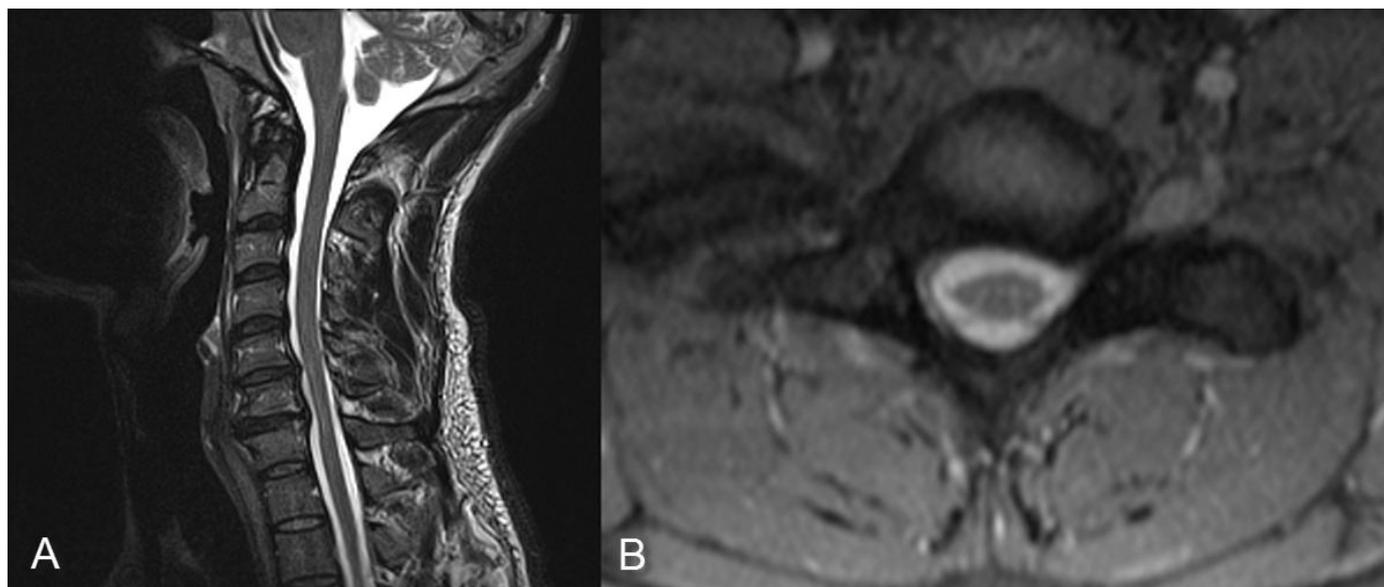


Figure 3. 33 year old male with right upper limb weakness and atrophy secondary to Hirayama disease. Neutral position MR images. Sagittal T2WI (a) shows discrete straightening of cervical curvature with localized spinal cord atrophy at C6-7 and associated mild-to-moderate degenerative changes at these levels. Axial T2WI obtained at C6 (b) exhibits asymmetric anterior spinal cord flattening, particularly right sided. (Parameters- TR: 4000ms, TE: 115ms, Slice thickness: 2 mm, Field strength: 1,5T)

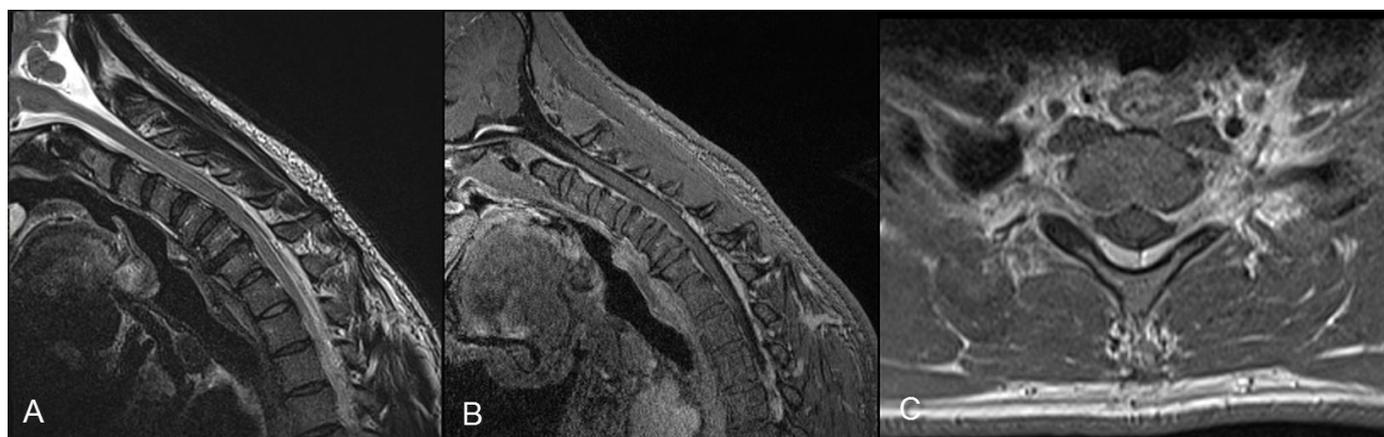


Figure 4. 33 year old male with hirayama disease. Flexion sagittal T2WI (Parameters- TR: 4000ms, TE: 115ms, Slice thickness: 3 mm, Field strength: 1,5T) (a) sagittal postcontrast T1WI with fat suppression (b) (Parameters- TR: 647ms, TE: 12ms, fat suppression, Slice thickness: 1 mm, Field strength: 1,5T, gadolinium dose 0.1 mmol/kg) and axial T1WI obtained at C7 parallel to disc orientation (c) shows forward displacement of the posterior dural wall, more prominent at C7-T1, with widening of the posterior epidural space, presence of flow voids, and homogeneous enhancement (b,c). Secondary cord compression is seen associated to subtle high signal intensity of the spinal cord at this same level. (Parameters- TR: 647ms, TE: 12ms, Slice thickness: 4 mm, Field strength: 1,5T, gadolinium dose 0.1 mmol/kg)

Etiology	Abnormal anterior displacement of the posterior dural wall with secondary compression and myelopathy of the spinal cord
Incidence	Rare
Gender ratio	Male: female ratio of 7:1
Age predilection	Mean age of onset of 16 years
Risk factors	Asian origin Male gender Familiar cases: Allergy? Superoxide dismutase 1 mutations?
Treatment	Cervical collar for 3-4 years. Surgical treatment with progressive neurological deterioration
Prognosis	Variable grades of right upper limb neurological deficit
Findings on imaging	<i>Plain film, CT:</i> Normal, abnormal alignment of the cervical spine <i>CT Myelography:</i> Displacement of the posterior dural sac with diminished anteroposterior diameter and compression of the cord <i>Neutral MR:</i> Asymmetric flattening of the anterior aspect of cord, especially at C6. High T2 signal intensity in the anterior cord due to myelomalacia <i>Flexion MR:</i> Anterior displacement of the posterior dura with an enlarged epidural space that contains flow voids and prominent enhancement on postcontrast images

Table 1. Summary table for hirayama disease

	X ray	MRI T1	MRI T2	MRI DWI
Hirayama disease	Normal. Abnormal alignment of the cervical spine.	Spinal cord atrophy at C6 – C7 levels. Dural anterior displacement of dural wall WITH compression of the spinal cord. Asymmetric flattening of the anterior aspect of the cord.	High signal intensity at the anterior aspect of the cord.	Non restricted
Normal	Normal.	Dural anterior displacement of dural wall WITHOUT compression of the spinal cord in 50% of subjects.	Normal	Non restricted
Degenerative disease	Sclerosis, abnormal alignment of the cervical spine. Osteophytes. Foraminal narrowing.	Disk-osteophyte complexes with/without secondary compression of the cord.		Non restricted
Demyelinating disease	Normal	Low signal intensity focal or diffuse lesion.	High signal intensity focal or diffuse lesion	Restricted/non restricted
Spinal cord infarct	Normal	Hypointensity in vascular territory distribution, usually anterior (anterior spinal artery).	Hyperintensity in vascular territory distribution, usually anterior (anterior spinal artery)	Restricted

Table 2. Differential diagnosis table for hirayama disease

ABBREVIATIONS

HD: Hirayama Disease
MR: Magnetic resonance
MRI: Magnetic resonance imaging

KEYWORDS

spinal cord compression; magnetic resonance imaging; spinal muscular atrophy

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