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Original Article

Imaging features in Hirayama disease

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Purpose: To evaluate the MR findings in clinically suspected cases of Hirayama disease. **Materials and Methods:** The pre and post contrast neutral and flexion position cervical MR images of eight patients of clinically suspected Hirayama disease were evaluated for the following findings: localized lower cervical cord atrophy, asymmetric cord flattening, abnormal cervical curvature, loss of attachment between the posterior dural sac and subjacent lamina, anterior shifting of the posterior wall of the cervical dural canal and enhancing epidural component with flow voids. The distribution of the above features in our patient population was noted and correlated with their clinical presentation and electromyography findings. **Observations:** Although lower cervical cord atrophy was noted in all eight cases of suspected Hirayama disease, the rest of the findings were variably distributed with asymmetric cord flattening, abnormal cervical curvature, anterior shifting of the posterior wall of the cervical dural canal and enhancing epidural component seen in six out of eight (75%) cases. An additional finding of thoracic extension of the enhancing epidural component was also noted in five out of eight cases. **Conclusion:** Dynamic post contrast MRI evaluation of cervicothoracic spine is an accurate method for the diagnosis of Hirayama disease.

Key words: Dynamic magnetic resonance imaging, Hirayama disease, monomelic amyotrophy

Juvenile muscular atrophy of the distal upper limb [Hirayama disease] is a rare disease affecting primarily young men in the second to third decades.[1,2] Hirayama disease, juvenile muscular atrophy of the distal upper extremity, is also known as monomelic amyotrophy,[3] benign focal amyotroph,[4] juvenile muscular atrophy of a unilateral upper extremity[5] and juvenile asymmetric segmental spinal muscular atrophy.[6] Hirayama disease differs from the known types of motor neuron diseases because of its nonprogressive behavior and pathologic findings of focal ischemic changes in the anterior horn of the lower cervical cord.[2,7] Various magnetic resonance imaging (MRI) features have been described in the literature for the diagnosis of this entity. The present study summarizes these different MRI findings and evaluates their distribution among the studied patient population.

Materials and Methods

Eight patients with clinical suspicion of Hirayama disease were evaluated with MR imaging. The pre and post contrast MR imaging was done in neutral and flexion position of cervical spine and imaging findings were correlated with their neurological presentation, disease course and electromyography findings. All the patients evaluated were males between 11-24 years of age with the mean age of 18.5 years. The criteria for patient selection were weakness and wasting predominantly in C7, C8 and T1 myotomes in one upper limb or asymmetrically in both upper limbs, insidious onset in teens or early twenties, initial fast progression for one to three years followed by arrest of disease or relatively benign course, irregular coarse tremors in the fingers of the affected hand[s], mild transient worsening of symptoms on exposure to cold, electromyography evidence of chronic denervation in the clinically or subclinically affected muscles and absence of objective sensory loss.[1] Electrophysiological study included nerve conduction study (NCS) and electromyography (EMG). Motor conduction studies were done in median and ulnar nerves (and radial in few cases) of both upper limbs and peroneal and tibial nerves of one lower limb. Sensory conductions were studied from median, ulnar and sural nerves of corresponding limbs. Needle EMG studies were done in muscles of C7 - T1 myotomes including the cervical paraspinals.

The MR imaging was done on a superconducting 1.5-T system (Signa, GE Medical Systems, Milwaukee, Wisconsin). The neutral-position MR protocol included transverse and sagittal T1-weighted (spin echo, repetition time msec/echo time msec of 500-600/15-20), transverse T2*-weighted (gradient-echo, 400-500/15-20, flip

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angle of 20°-30°) MR imaging and sagittal T2-weighted MR imaging (fast spin echo, 3000-3500/90-140 [effective]). The flexion MR imaging protocol consisted of sagittal T1-weighted (500-600/15-20) and T2-weighted (4,000/85-99, matrix size of 256 × 256) MR imaging and transverse T2*-weighted MR imaging (400-500/15-20, flip angle of 20°-30°, matrix size of 256 × 192) with approximately 30-40° neck flexion by using an indigenous positioning sponge. Post gadolinium transverse and sagittal T1 weighted images in flexion and extension were obtained in all cases. Section thickness was 4 mm with 1-mm gap for both sagittal and transverse MR imaging with all sequences. Lower cervical cord was defined as the cord between C4 and C7. The following features were evaluated: localized lower cervical cord atrophy, asymmetric cord flattening, abnormal cervical curvature, loss of attachment between the posterior dural sac and subjacent lamina, anterior shifting of the posterior wall of the cervical dural canal and enhancing epidural component with flow voids. An additional feature detected was the thoracic extension of the enhancing epidural component. Localized cord atrophy was defined as a decrease in cord size in comparison with the normal cord above and that below the affected level confirmed on axial images.

Asymmetric cord flattening was evaluated on transverse MR images. To avoid confusion with cord compression due to adjacent spurs or herniated disks, cord flattening was defined as flattening without a narrowed or obliterated adjacent subarachnoid space. An elliptic spinal cord was considered normal, a pear-shaped spinal cord was considered asymmetric cord flattening and a triangular spinal cord was considered symmetric cord flattening. Cervical curvature was classified according to the principles suggested by Guigui *et al*.[8] and Batzdorf and Batzdorff.[9] Cervical curvature was measured according to the relationship of the dorsal aspect of the vertebral bodies C3 through C6 to a line drawn from the dorsocaudal aspect of the vertebral body C2 to the dorsocaudal aspect of the vertebral body C7. By definition, normal lordotic cervical curvature is curvature in which no part of the dorsal aspect of the vertebral bodies C3 through C6 crosses the line from C2 through C7. An abnormal [straight or kyphotic] curvature is curvature in which part or all of the dorsal aspects of the vertebral bodies C3 through C6 meet or cross through the line from C2 through C7 [Figure 1]. For evaluating the loss of attachment between the posterior dural sac and subjacent lamina, the lamina was defined as the part of vertebra between junctions of laminae medially and laterally by a tangential line along the medial aspect of the pedicle. This was divided equally into three parts. More than 33.3% loss of attachment between the posterior dural sac and subjacent lamina was considered significant.[10] On flexion studies, anterior displacement of dural sac and appearance of enhancing epidural component posterior to the thecal sac, which disappeared on neutral position, was noted.

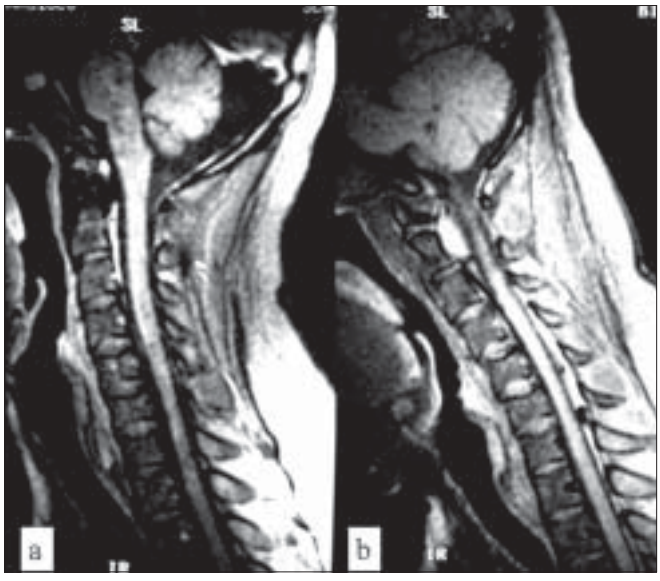

Figure 1: Neutral and flexion position post contrast fat saturated sagittal MRI images of patient no. 5 showing the anterior displacement of the dural sac and enhancing posterior epidural component with flow voids

Results

Clinical features

All the patients were male with mean age of 18.5 years (range 11-24) years at the time of clinical examination. The duration of illness at the time of clinical examination and electrophysiology study was three to 12 months in seven patients and 48 months in the eighth (patient no. 4). Major symptoms included hand wasting (8/8), grip weakness (6/8), tremulousness of fingers (4/8). The patient who presented after four years of illness also had forearm wasting, weakness and dysaesthesia. On clinical examination, all the patients had pure motor, asymmetrical, upper limb, lower motor neuron type of involvement [Table 1]. None of the patients had any neck pain or radicular symptoms.

Table 1: Clinical findings

Clinical findings	No. of patients
Hand wasting [intrinsic muscles]- asymmetrical	6
Forearm wasting	4
Brachioradialis sparing [oblique atrophy]	3
Arm wasting [biceps > triceps]	1*
Hand weakness	8
Forearm weakness [FDP > FDS > WF > PQ]	6
Minipoly myoclonus	5
Brisk deep tendon reflexes	1†
Bilateral involvement	6

FDP - Flexor digitorum profundus FDS - Flexor digitorum superficialis, WF - Wrist flexors PQ - Pronator quadratus *The patient who presented late. No elbow or shoulder weakness in spite of arm wasting, †No other upper motor neuron features. MRI shows dynamic changes

Table 2: Electrophysiological findings

	Electrophysiologic findings (patients number)							
	1	2	3	4	5	6	7	8
Distal latency - Ulnar	N	↑	↑	↑	N	↑	N	N
Median	N	N	N	↑	N	↑	N	N
Radial	N	N	N	N	N	↑	N	N
CMAP - Ulnar	N	↓	↓	↓	↓	↓	N	↓
Median	N	N	N	↓	N	N	N	N
Radial	N	N	N	N	N	↓	N	N
F wave latency-Ulnar	N	↑	↑	±	0	0	N	N
Median	N	N	N	0	0	N	N	N
Radial	N	N	N	N	N	N	N	N
SSEP	-	-	N	-	-	-	-	-
EMG	I, D	I, FCR	I, FCR FCU	I	I	I	I	I
Bilateral involvement	+	+	-	+	-	-	+	+

CMAP - Compound muscle action potential, SSEP - Somatosensory evoked potential, EMG - Electromyography, I - Intrinsic hand muscles [first dorsal interosseus, abductor pollicis brevis mostly tested], D - Deltoid, FCR - Flexor carpi radialis, FCU - Flexor carpi ulnaris, ↑ - Prolonged, ↓ - Reduced, N - Normal, ± - Inconsistent, 0 - Absent

Electrophysiology

Nerve conduction study and EMG were done in all the patients within a week of the clinical examination. In addition, somatosensory evoked potentials (SSEP) were studied in one patient. Ulnar compound muscle action potential (CMAP) was reduced in six patients while median and radial in one each. Distal latencies and F-wave latencies were variably affected. Conduction velocities were normal in all nerves. Sensory conduction study was absolutely normal in all the patients. Median SSEP done in one patient was normal. All the eight patients showed denervation changes (fibrillations, positive sharp waves, large amplitude potentials but no polyphasics or fasciculations) mainly in C8-T1 muscles. Cervical paraspinal EMG was done in six patients and all were normal [Table 2].

Imaging findings [Table 3]

All eight cases of suspected Hirayama disease showed lower cervical cord atrophy with asymmetry noted in six of eight cases [Figure 2]. These six cases were also showing other findings like abnormal cervical curvature, anterior shifting of the posterior wall of the cervical dural canal and enhancing epidural component. The thickness of the enhancing epidural component was, however, variable ranging from 1.5 to 6 mm. No relation was found between the duration of illness and thickness of the epidural component. An additional finding of thoracic extension of the enhancing epidural component was also noted in five cases. Abnormally prominent posterior epidural flow voids were noted in four cases. Loss of attachment between the posterior dural sac and subjacent lamina was noted in four of these cases [Figure 3].

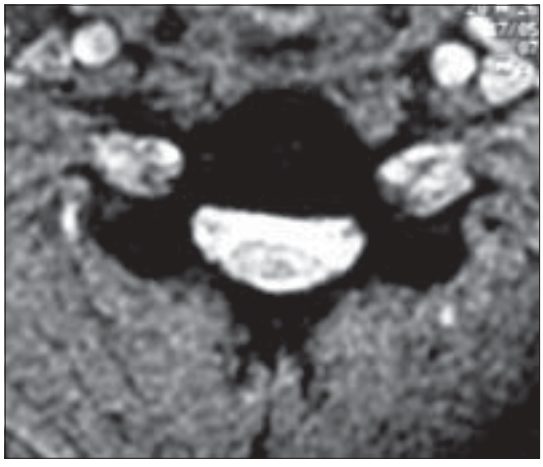

Figure 2: Axial neutral position gradient echo MR image from the cervical spine of patient no.4 showing asymmetric cord flattening

Table 3: magnetic resonance imaging features

Magnetic resonance imaging features	No. of patients
Localized lower cervical cord atrophy	8
Asymmetric cord flattening	6
Abnormal cervical curvature	6
Loss of attachment between the posterior dural sac and subjacent lamina	4
Anterior shifting of the posterior wall of the cervical dural canal	6
Enhancing epidural component in lower cervical region	6
Thoracic extension of the enhancing epidural component	5
Prominent epidural flow voids	4
Parenchymal signal changes in lower cervical cord	3

Follow-up

Follow-up was available in six patients. One patient (patient no. 5) was initially evaluated in June 1996 when clinical and electrophysiology features were suggestive of Hirayama's disease but MRI (no dynamic study done) was negative. Repeat examination after eight years (September 2004) showed mild progression (increased wasting and reflex loss). Repeat MRI done was suggestive of Hirayama's disease in the dynamic study. The other five patients had last follow-up after four to 11 months of presentation (four using cervical collar, including patient no.3 whose MRI is negative). All the five had static course clinically.

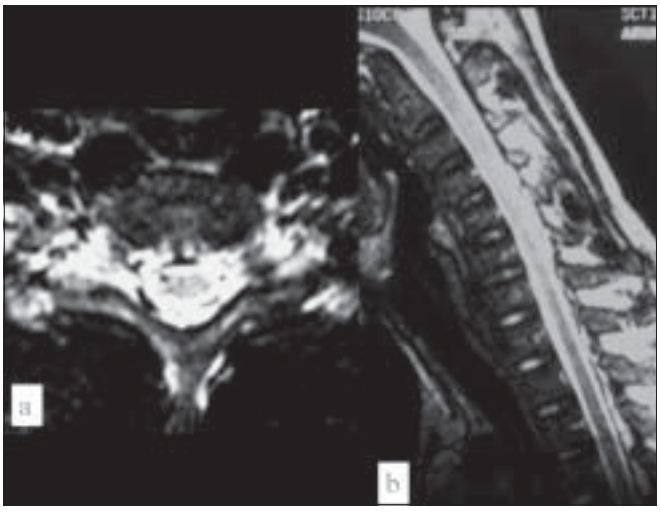

Figure 3: Neutral axial and sagittal flexion FSE T2 MRI images showing loss of attachment between the posterior dural sac and subjacent lamina, signal changes, anterior displacement of the dural sac at the cervicothoracic junction

Discussion

The pathogenesis of this entity is debated. An assumption of imbalanced growth between the patient's vertebral column and spinal canal contents has been suggested.[6,11,12] This imbalanced growth will cause disproportional length between the patient's vertebral column and the spinal canal contents causing a tight dural sac and anterior displacement of posterior dural wall when the neck is flexed. The different growth rates between male and female patients have been proposed to be the factor related to the male preponderance of Hirayama disease by Toma *et al*.[13] The incidence of atopic disorders and levels of serum IgE and mite antigen-specific IgE were also found to be higher in patients with Hirayama disease than in the general population suggesting that atopy may be a contributing factor for Hirayama disease.[14]

In a normal spine, the spinal dura mater is a loose sheath that is anchored in the vertebral canal by the nerve roots and by its attachment to the periosteum in two places: one at the foramen magnum and the dorsal surfaces of C2 and C3 and the other at the coccyx. Normally, the slack of the dura mater can compensate for the increased length in flexion. In patients with Hirayama disease, however, the tight dural sac can result in separation of the posterior dural sac from its subjacent lamina and various degrees of abnormal cervical curvature (straight or kyphotic). On neck flexion, the tight dural sac cannot compensate for the increased length of the posterior wall, which causes anterior shifting of the posterior dural wall and consequent compression of the cord against the posterior margin of adjacent vertebral bodies. This compression may cause microcirculatory disturbances in the territory of the anterior spinal artery in the lower cervical spinal cord.[1,7] The chronic circulatory disturbance resulting from repeated or sustained flexion of the neck may produce gliosis and localized cord atrophy at the lower cervical region.[15] Different pathophysiological mechanisms have been proposed for the venous engorgement seen on flexion studies. Firstly, the negative pressure in the posterior spinal canal resulting from anterior shifting of the dural canal is thought to increase the flow to the posterior internal vertebral venous plexus causing their prominence. Second, the posture of neck flexion decreases the venous drainage of the jugular veins, impeding the venous return of internal vertebral venous plexus causing their prominence and engorgement.[16] Finally, the compressed anterior internal vertebral venous plexus caused by anterior displacement of the dural canal increases the burden of the posterior internal vertebral venous plexus leading to its distension.[17] The extensions of posterior epidural enhancement in the thoracic region found in our cases suggest that jugular vein compression, as a cause of prominence of the epidural venous plexus is less likely.

The various MR imaging features described in Hirayama disease are as follows: localized lower cervical cord atrophy, asymmetric cord flattening, parenchymal signal changes in lower cervical cord, abnormal cervical curvature, loss of attachment between the posterior dural sac and subjacent lamina, anterior shifting of the posterior wall of the cervical dural canal, enhancing epidural component in lower cervical and thoracic region and prominent posterior epidural flow voids suggestive of dilated epidural venous plexus. The findings reported more frequently are asymmetrical/symmetrical atrophy of lower cervical cord, prominence and enhancement of posterior epidural venous plexus on flexion studies and anterior shifting of posterior dural sac on flexion. Loss of attachment between the posterior dural sac and subjacent lamina on neutral position, anterior shifting of the posterior wall of the cervical dural canal, enhancing epidural component in the lower cervical and thoracic region and prominent posterior epidural flow voids suggestive of dilated epidural venous plexus on flexion studies are reported as highly suggestive for the diagnosis of Hirayama disease.[1,6,10,12,15,17]

Early diagnosis and therapeutic intervention in the form of cervical collar therapy to prevent neck flexion may minimize the functional disability of the young patients. This induces a premature arrest of disease progression and is more beneficial in those cases with shorter duration of illness. Since the progressive stage is expected to cease in a few years, application of a cervical collar for three to four years generally has been advocated.[18] In selected patients, encouraging results have also been obtained with surgical intervention, which involves mainly cervical decompression and/or fusion with or without duraplasty.[19]

Conclusion

Dynamic post contrast MRI evaluation of cervicothoracic spine is a helpful method in arriving at the correct diagnosis of Hirayama disease and should be an essential part of the protocol in cases with high suspicion of motor neuron disease.

Acknowledgment

Authors wish to thank the Director, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum, Kerala, India for providing the facilities and for the kind permission to present this report.

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Accepted on 24-10-2007
Source of Support: Sree Chitra Tirunal Institute for Medical Sciences and Technology, Kerala, India. **Conflict of Interest:** None declared.