CONTRAST DYNAMIC MR IMAGING IN HIRAYAMA DISEASE ON A 3-TESLA MRI SCANNER IN A TERTIARY CARE CENTRE IN EASTERN INDIA

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ABSTRACT

BACKGROUND
Hirayama disease, also termed non-progressive juvenile spinal muscular atrophy of the distal upper limbs, is a type of cervical myelopathy related to flexion movements of the neck.

The objective of the study was to evaluate the role of Dynamic MR imaging findings in young patients with clinical suspicion of Hirayama disease.

MATERIALS AND METHODS
Thirteen patients (age range from 16 to 26 years) with clinical suspicion of Hirayama disease underwent thorough clinical evaluation and dynamic cervical MRI in the Department of Radiology at IPGME&R within a time duration of June 2014 to September 2016.

RESULTS
All the thirteen patients showed anterior shifting of posterior dural sac, cord flattening, abnormal curvature, enhancing epidural component. Nine of them showed localised cord atrophy and intramedullary T2 hyperintensities. Ten patients showed prominent flow voids.

CONCLUSION
Hirayama disease, a rare disease affecting young adults almost always in the second to third decades of life, is characterised by insidious onset and slowly progressive course followed by static phase of unilateral or asymmetric atrophy of the hand(s) and forearm(s) with sparing of the brachioradialis, characterised as oblique amyotrophy. Dynamic contrast MRI has accurate and characteristic findings which help in early diagnosis and early institution of therapy.

KEYWORDS
MATERIALS AND METHODS

Thirteen patients (age range from 16 to 26 years) with clinical suspicion of Hirayama disease underwent thorough clinical evaluation and dynamic cervical MRI in the Department of Radiology of SSKM and IPGME&R Hospital within a time duration of June 2014 to September 2016. Inclusion criteria for patients were:

1. Weakness and atrophy of the upper limb muscles in forearm and hand with relative sparing of the brachioradialis and cold paresis.
2. Above mentioned findings may be unilateral or bilateral.
3. Gradual onset of the above mentioned clinical symptoms followed by arrest of symptoms.
4. Absence of sensory or pyramidal tract symptoms.

Dynamic MRI for the cervical spine was done on a 3-Tesla (super conducting) MRI machine, WIPRO-GE Signa HDxt. First in neutral position sagittal T2 weighted images (Repetition time (TR)/Echo time (TE), 3000/105.8) and axial T2 weighted (TR/TE, 37000/102.1) images were performed. Then in flexed position, sagittal T2 weighted (TR/TE, 2980 /105.6) and axial T2 weighted (TR/TE 4420/104.7) images were taken.

Later gadolinium based contrast was given to the patients in a dose of 0.2 mL/kg and further sagittal and axial T1 sequences were taken. Flexion was achieved by placing a foam rest behind the head to an angle of approximately 45° and the chin was resting over the chest. For all sequences the slice thickness was 4 mm.

The diagnostic criteria evaluated were: localised cervical cord atrophy, presence of cord flattening, intramedullary signal hyperintensity on T2 weighted sequences, anterior shifting of the posterior wall of the cervical dural sac, presence of flow voids in the posterior epidural space during flexion and in post-contrast study, enhancement of the epidural space is noted. Most of the findings were appreciated in our study population.

Localised cord atrophy is described as decrease in cord size in comparison of normal cord above and below the affected area in sagittal MR images and is confirmed by transverse images. To avoid confusion with cord compression due to adjacent spurs or herniated disks, cord flattening is described as flattening without obliterated or narrowed adjacent subarachnoid space. An elliptical spinal cord is considered normal, whereas a pear shaped spinal cord is considered asymmetric cord flattening and a triangular cord considered to be symmetric cord flattening.

Cervical curvature is classified according to the principles given by Guigui et al and Batzdorf and Batzdorff. Cervical curvature is 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 body of C2 vertebra to the dorsocaudal aspect of the body of C7 vertebra. By definition, normal lordotic cervical 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.

For evaluation of the loss of attachment between the posterior dural sac and adjacent 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 the adjacent lamina is considered significant.

On flexion studies, anterior displacement of dural sac and appearance of epidural flow voids with enhancing epidural component posterior to the thecal sac was noted, it disappeared on neutral positions.

RESULTS

Clinical features: Clinical criteria was fulfilled by 13 of the patients. All the patients in the study were male with mean age of 19 years.

**Table 1. Clinical Findings**

<table>
<thead>
<tr>
<th>Clinical Findings</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insidious onset</td>
<td>13</td>
</tr>
<tr>
<td>Cold paresis</td>
<td>5</td>
</tr>
<tr>
<td>Oblique amyotrophy</td>
<td>11</td>
</tr>
<tr>
<td>Unilateral</td>
<td>10</td>
</tr>
<tr>
<td>Bilateral</td>
<td>3</td>
</tr>
<tr>
<td>Fine tremulousness</td>
<td>7</td>
</tr>
<tr>
<td>Hyperaesthesia on dorsum of hand</td>
<td>2</td>
</tr>
<tr>
<td>Sensory symptoms</td>
<td>0</td>
</tr>
</tbody>
</table>

**Table 2. MRI Findings in Hirayama Disease in Neutral, Neck Flexed and Post-contrast Study**

<table>
<thead>
<tr>
<th>MRI Findings</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior shifting of posterior dural sac</td>
<td>13</td>
</tr>
<tr>
<td>Epidural flow voids</td>
<td>13</td>
</tr>
<tr>
<td>Cord flattening</td>
<td>13</td>
</tr>
<tr>
<td>Localised cord atrophy</td>
<td>9</td>
</tr>
<tr>
<td>Intramedullary T2 Hyperintensities</td>
<td>9</td>
</tr>
<tr>
<td>Abnormal cord curvature</td>
<td>12</td>
</tr>
<tr>
<td>Enhancing epidural component</td>
<td>13</td>
</tr>
</tbody>
</table>

All the patients showed anterior shifting of posterior dural sac, cord flattening, abnormal curvature, enhancing epidural component. Nine of them showed localised cord atrophy and intramedullary T2 hyperintensities. 10 showed prominent flow voids.
CONCLUSION

Hirayama disease is a benign self-limiting disease. In our study, we found young men mainly affected by it. Mostly the affection was unilateral but in three of the cases we got bilateral asymmetrical presentation clinically which was confirmed on dynamic MRI. The diagnosis of Hirayama is quite clear in flexion MR studies, but real challenge for the radiologist is to identify it in non-flexion studies. So routinely flexion studies should be done in young male with hand wasting and also if motor neuron disease has been excluded. There are limited differentials for Hirayama disease like ulnar neuropathy, motor neuron disease, syringomyelia, traumatic myelopathy, etc. Therefore, it becomes important for early identification and thereby stopping further progression of disease.

The most widely accepted hypothesis is based on supposition of imbalance of growth between vertebral column and the spinal cord. The difference in growth rates in male and female may be the explanation behind the male dominance of Hirayama disease. Usually the exiting nerve roots with its dural covering acts as anchor, to hold the dural sac in place in the vertebral canal. Spinal dura is attached to the vertebral column at foramen magnum and dorsal aspect of C2-3 and other at the coccyx. Remaining dura is loosely suspended inside the vertebral canal. In the neutral position of neck, the dura is slack but with neck flexion there is overall increase in length of vertebral canal. The difference in length of dura in flexion and extension D1 to top of atlas is 1.5 cm at the anterior wall and 5 cm at the posterior wall. Flexion causes stretching in the dura, which remains abutting to the walls of the vertebral canal in normal persons. In patients of Hirayama, the dura is already shorter in length and is tight in neutral position. On flexion this tight dura detaches from its fixed sites and falls forward causing abutment of the spinal cord to the anterior vertebral column. The recurrent compression causes atrophy and gliosis and all the changes noticed in MR has been discussed in detail.

Forward shifting of the posterior dural sac and engorgement of posterior epidural venous plexus are considered the characteristic signs of HD in flexion cervical MRI. Some authors have reported the presence of forward shifting of the dural sac in all their patients. Nevertheless, this sign is not always present in Hirayama patients, a prevalence respectively of 71% and 76% of forward shifting. Lai et al demonstrated the presence of anterior shifting of posterior dural sac in 46% of healthy subjects, of a lesser degree compared to patients group and without evidence of cord compression. Therefore, they suggested that the forward shifting of the posterior dural sac on neck flexion can occur as normal variation and its presence does not always lead to diagnosis of HD. In our study, anterior displacement of posterior cervical dural sac was present in all the patients, in 100% cases.

Localised cervical cord atrophy was reported by Hirayama and Tokumaru in about 50% of cases. Sonwalkar et al and Raval et al demonstrated cord atrophy in 100% of their patients, Hassan et al in 82% of cases. Among our
patients, nine presented with cervical cord atrophy, about 69%.

12 out of 13 (92%) of our patients had loss of normal cervical lordosis. In agreement with our finding were the studies of Hassan et al and Raval et al that depicted loss of cervical curvature in 91% and 100% of their patients.13,18

Nine of our patients (69%) showed T2 hyperintensities of the spinal cord. High signal intensities on T2 weighted images without external cord compression are described in the literature.

Neurophysiological studies such as somatosensory evoked potential (SEP) have contributed in understanding the disease aetiology. A study by Nalini A et al19 shows most frequently the abnormality was absence or reduced amplitude of the cervical N13 potential. It was seen in all Hirayama patients.

Conservative treatment like hard cervical collar which prevents cervical flexion has shown to reduce advancement of the disease. A study by Cortese et al has shown timely diagnosis use of cervical collar lowers the progression of disease. Surgery is indicated in selective cases where condition worsens even after conservative treatment and includes anterior cervical decompression surgery or duroplasty.

Hirayama is a frequently undiagnosed condition with simple and inexpensive treatment. There should be high suspicion in young male patients with upper limb weakness. MR has predictive value for its diagnosis. Early detection not only halts the disease progress but improves the future of the patient.

Thus, we conclude dynamic MRI in flexion position is most important for diagnosis of Hirayama disease.

Acknowledgement

We thank the Department of IPGME&R and SSKM Hospital and our participants for their cooperation.

REFERENCES


