Case Report

Flexion MRI in a case of Hirayama disease

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Abstract

We report the case of an 18-year old male with a history of asymmetric weakness and amyotrophy of distal upper extremities, suggestive of Hirayama disease. Magnetic resonance imaging (MRI) of the cervical spine was obtained both in flexion and neutral position. Flexion MRI showed forward displacement of the dura and subsequent cord compression, associated marked enlargement and postcontrast enhancement of posterior epidural plexus. These findings are pathognomonic of the disorder. On neutral MRI abnormalities may be subtle: in our case, they included loss of physiological lordosis, asymmetric atrophy and increased T2 signal intensity of the lower anterior cervical cord. The ability to identify abnormalities on neutral MRI however is even more important in that it allows the radiologist to include a flexion sequence in the MRI examination, if not specifically requested by the referring physician, and in cases in which the suspicion of the disorder has not been raised.

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Case report

Hirayama disorder (HD) is a rare condition characterized by distal asymmetric weakness and wasting of upper extremities mainly affecting the C5-T1 or C5-C7 segmental myotomes [1,2]. The disease most often occurs in young adults, with a male predominance [3]. It is mostly sporadic. However, a handful of familial cases have been reported in the literature [4–9]. Clinical progression is typically self-limiting.

We present the case of an 18-year-old male admitted with a history of progressive weakness and amyotrophy of distal upper limbs, along with abnormal coloration and hyperhidrosis. Findings predominated to the right. The patient reported episodes of painless burns while cooking. He also reported to have suffered a neck injury from a fall 2 years earlier, of no major clinical significance. No imaging was performed at that time.

Interestingly, the patient mentioned that also his father had a history of amyotrophy of the superior upper limbs, with onset at age 30. He was investigated at another institution and no definite diagnosis was made. Unfortunately, we did not have access to his clinical records.

Our patient’s neurological examination revealed brisk reflexes, erythrocyanosis, and thermalgesic hypoesthesia of the hands. It confirmed distal amyotrophy, predominating to
the right, with relative sparing of the brachioradialis – a picture referred to as oblique atrophy. Cerebral magnetic resonance imaging (MRI) and lumbar puncture were unremarkable. No oligoclonal bands were found and testing for anti-Q4 was negative, therefore a primary demyelinating etiology was excluded. HD was suspected and an MRI of the cervical spinal cord was requested to confirm the diagnosis.

Images were acquired on a 1.5 Tesla scanner (Achieva, Philips Medical Systems, Best, The Netherlands). The MRI protocol included imaging in neutral position followed by imaging in hyperflexion. As in Hassan et al [10], hyperflexion of the neck was achieved by asking the subject to first extend the head as much as possible and then to touch the chin to the chest. The subjects’ shoulders were placed as far caudal as possible. Hyperflexion was maintained by supporting the neck and shoulders with MR-compatible foam pads.

Neutral MRI of the cervical spine showed loss of cervical lordosis, focal atrophy of the lower cervical cord and associated increased T2 signal intensity of anterior horns, asymmetric to the right. There was no evidence of loss of dural attachment.

Flexion MRI demonstrated anterior shifting of posterior dura, with subsequent cervical cord compression. The posterior epidural space was markedly enlarged, with prominent flow voids and contrast enhancement on postgadolinium fat-suppressed T1-weighted images (WI).

The maximum forward shifting of posterior dura, measured on flexion MR postgadolinium fat-suppressed T1 WI in the mid-sagittal plane, as in Boruah et al [11], was 6.5 mm.

MRI findings are illustrated in Figure 1.

Discussion

On neutral MRI, there was loss of physiological cervical lordosis, which is a common finding in patients with HD, although nonspecific [11–13]. Cervical curvature was assessed as illustrated by Chen et al [13,14], by drawing a line from the inferior aspect of the C2 vertebral body to the inferior aspect of the C7 vertebral body and investigating the relationship of the dorsal aspect of the C3-C6 vertebral bodies to this line. The authors had referred to previous works [15,16], according to which an abnormal (straight or kyphotic) curvature is one in which the dorsal aspect of all or part of the C3 to C6 vertebral bodies meet or cross the line from C2 to C7, whereas in a physiological cervical lordosis curvature no part of the dorsal aspect of the C3 to C6 vertebral bodies crosses the line from C2 to C7.

Signal abnormalities and cord atrophy were demonstrated at the C5-C6 level, in agreement with previous literature reporting that HD more commonly affects the C5–C7 myotomes in Western countries, whereas the C7-T1 segment is predominantly affected in Asian countries [2].

The predominant involvement of the right hemicord is also in line with the existing evidence: the disorder is known to more frequently affect the right upper limb than the left, with contralateral upper limb involvement subsequently observed in 50% of cases with progression of the disease [17,18]. The mechanism underlying the predominant involvement of the right hemicord, however, remains unclear. Shinomiya et al [19,20] proposed a “posterior epidural ligament factor” theory: according to the authors, asymmetric cord compression might be related to an unequal right-to-left distribution of posterior epidural ligaments between the posterior dura and the ligamentum flavum.

On neutral MRI abnormalities might be subtle. Findings on flexion MRI, conversely, are highly specific. The MRI features of HD were first described by Pradhan and Gupta in 1997 [12]. Forward shifting of posterior dura has also been observed in healthy subjects on flexion MRI, however displacement ranged from 1.0 to 4.2 mm as compared to 6.1–7.8 mm in HD patients [21].

Weakness of distal upper limbs in HD has been related to segmental injury of the lower cervical cord anterior horn cells. Because of the benign course of the disease, biopsy is typically avoided. Pathology findings could be obtained only several years after HD was first described (in 1959 [1]), from a patient who died of lung cancer at the age of 38, 23 years after the onset of HD [22]. The authors report that changes predominated at the C7-C8 level. Both anterior horns had shrunk to less than half of the normal anteroposterior diameter; both large and small nerve cells had decreased in number, and the remaining nerve cells showed degenerative changes such as lipofuscin accumulation, chromatolysis and shrinkage. The posterior horns and the white matter were unaffected [22].

The observed pathology differences differed from those of any previously reported disorder affecting the spinal cord, however, the authors outline how they closely resembled the “téphromalacie antérieure” of Marie and Foix, which is characterized by infarcts of the lower cervical anterior horns resulting from syphilitic arteritis or arteriosclerosis [23]. Unlike HD, however, both syphilitic arteritis and arteriosclerosis occur more often in elderly patients. Moreover, in their axial sections of the cord, the necrotic changes were not limited to the anterior horns but involved also the anterior portion of the posterior funiculi.

Selective injury of lower cervical anterior horn cells in HD has been related to microcirculatory disturbances induced by repeated or sustained flexion of the neck and is in keeping with their extremely high vulnerability to ischemia [24,25].

Kikuchi et al suggested that a disproportion in length between the vertebral column and the spinal canal content, resulting in a “tight dural sac,” could represent a predisposing factor [26]. The dura mater is a dense laminated membrane made of collagen and elastic fibers oriented concentrically around the spinal cord [27]. It is firmly attached at the foramen magnum cranially and to the coccygeal vertebrae caudally. Laterally, it extends over and blends with the nerve roots epineurium [28]. Within the spinal canal, the dura is only loosely attached to the posterior longitudinal ligaments and is therefore free to move considerably during spinal movements. Most notably, the cervical spine increases in length approximately 3 cm during neck flexion. Consequently the dura, attached above and below, migrates within the spinal canal, compensating for the increased length of the cervical canal in flexion [28,29].

A short dura, conversely, cannot compensate for the increased length of the cervical canal during flexion and is therefore displaced anteriorly, compressing the spinal cord [26].
According to Toma et al, the imbalance in growth between spinal canal and dural sac could be accentuated during the juvenile growth spurt, explaining the onset of the disorder in adolescence. Moreover, the different growth rates between male and female patients could explain the male preponderance of HD [30].

HD differs from motor neuron disorders: prognosis in this condition is favorable; prompt recognition allows early intervention, which has been shown to stop disease progression.

HD has characteristic imaging features on flexion MRI. The ability to identify abnormalities on neutral MRI however is even more important in that it allows the radiologist to include a flexion sequence in the MRI examination, thereby increasing the detection rate of the disorder. Radiologists should maintain a high level of clinical suspicion for HD in young male patients referred for MRI for asymmetric weakness and atrophy in the lower cervical and T1 distributions. On neutral MRI abnormalities might be subtle. A thorough search for loss of physiological lordosis, loss of dural attachment, asymmetric

Fig. 1 – MRI findings. Neutral sagittal T2-WI (a) and axial T2-GRE WI (d) demonstrate focal atrophy and increased signal intensity of the spinal cord at the C5-C6 level, less than two vertebral bodies in height (arrow). Signal abnormalities are restricted to the anterior horns of the cervical cord and are asymmetric to the right (arrow). In (a), a line hypothetically drawn from the inferior aspect of the vertebral body of C2 to the inferior aspect of C7 would be indistinguishable from the dorsal aspects of the vertebral bodies C3 to C6, indicating loss of physiological cervical lordosis according to Chen et al [13]. Flexion sagittal (b) and axial GRE (e) T2-WI show anterior shifting of posterior dura, and asymmetrical cervical cord flattening (arrow). The posterior epidural space is markedly enlarged, with prominent flow voids (arrow). There is complete obliteration of posterior subarachnoid space from the C3-C4 level to C6-C7. Postgadolinium fat-suppressed T1-WI (c) show vivid enhancement of posterior epidural venous plexus (arrow).
atrophy and increased T2 signal intensity of the lower anterior cervical cord is mandatory.

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