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CASE REPORT

Magnetic Resonance Imaging Findings of Hirayama's Disease

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ABSTRACT

Hirayama's disease is a rare form of cervical myelopathy resulting in juvenile muscular atrophy of the upper limb. We describe the magnetic resonance imaging findings of Hirayama's disease in a 20-year-old male, who demonstrates abnormal detachment and forward displacement of the posterior cervical dura during neck flexion, with the lower cervical cord abutting on the posterior border of the lower cervical vertebral bodies. This compression causes ischaemic changes in the lower cervical cord and hence the subsequent denervation and atrophy of the distal upper limb muscles. A high index of clinical suspicion and magnetic resonance imaging of the cervical spine with neck flexion are the key to diagnosis.

Key Words: *Cervical vertebrae; Magnetic resonance imaging; Muscular atrophy, spinal; Spinal cord diseases*

中文摘要

平山病的磁共振成像表現

鄧業勤、朱志揚、梁錦榮、岑承輝、盧成瑋、鄭志成、邱麗珊

平山病是一種罕有的頸髓病，病者一般會在青少年時期出現上肢肌萎縮。本文報告一名20歲平山病男性青年患者的磁共振成像。當他頸部向前屈曲時，頸椎後硬膜出現異常的分離及向前移位，而下頸髓則靠近下頸椎椎體的後緣。這壓迫現象使下頸髓出現缺血性改變，繼而引致上肢肌神經性萎縮。診斷此症的關鍵是要對它的臨床症狀有高度警覺，並留意病人頸椎前屈時之磁共振成像表現。

INTRODUCTION

Hirayama's disease is a rare form of cervical myelopathy resulting in juvenile muscular atrophy. It is a unilateral or less commonly asymmetric bilateral disease, involving predominantly the distal muscles of the upper limbs due to compression and ischaemic injury to the cervical cord from detachment of the posterior dura. We report a case of Hirayama's disease in a 20-year-old male, highlighting the classical presenting features and typical imaging findings on magnetic resonance imaging (MRI).

CASE REPORT

A 20-year-old male was referred by a general practitioner to a neurologist at Pamela Youde Nethersole Eastern Hospital for weakness and atrophy of the right upper limb muscles for eight months, which was slowly progressive, in October 2010. The patient denied the presence of any sensory disturbance. There was no history of recent injury, nor was there any family history of neuromuscular disease.

Physical examination showed wasting of the

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intrinsic muscles of the right hand. There was no fasciculation observed. The brachioradialis muscle was spared. Sensation and tendon reflexes were intact. Electromyography showed chronic denervation changes over right hand muscles, while the proximal muscles were spared. In view of the clinical suspicion of Hirayama's disease, the neurologist requested an MRI of the cervical spine in both neutral and fully flexed positions.

MRI was performed using a 1.5 T scanner (Magnetom Symphony Maestro Class, Siemens, Germany). There was loss of normal cervical lordosis. Mild localised cervical cord atrophy and asymmetrical cord flattening were noted at C5/6 levels in the neutral position. There was no abnormal intramedullary T2 hyperintense signal seen in the cervical cord. No intramedullary or extramedullary mass lesion causing cord compression was seen. With the neck fully flexed, the cervical cord became more flattened compared to that seen in the neutral position. The lower cervical cord was seen abutting on the posterior border of the C5 and C6 vertebral bodies. The posterior dura was detached and displaced forwards at C3 to T2 levels. Flow voids were seen within the posterior epidural space. Post-gadolinium images showed intense enhancement posterior to the detached dura due to enhancement of the engorged posterior epidural venous plexus (Figure). Findings were typical of Hirayama's disease.

DISCUSSION

Hirayama's disease was first described in Japan in 1959.[1] Since then, further cases have been reported in other Asian countries and the rest of the world.[2,3]

The classical presentation is development of weakness and atrophy of the hand in young individuals in their second to third decades, with insidious onset and slow progression. Since the brachioradialis muscle is typically spared, the pattern of forearm involvement is referred to as an oblique amyotrophy.[4] The right upper limb is more frequently involved regardless of handedness, although there are also rare cases of bilateral symmetrical involvement.[5] In some cases, there is an associated finding of cold paresis,[6] but there has been no reported sensory or pyramidal tract involvement. There is a male predominance and while most reported cases are sporadic, there are also documented cases of familial involvement.[3]

The pathogenic mechanism is unknown, but the most accepted explanation is that there is an imbalance in growth between the vertebral column and spinal canal contents, causing neck flexion–induced myelopathy. The spinal dura mater is attached to the periosteum at two ends: proximally at the foramen magnum and dorsal surfaces of C2 and C3, and distally at the coccyx.[7] The rest of the dura mater is loosely suspended inside the vertebral column. Normally, the dura mater is slack when the neck is in the neutral position, and is stretched when the neck is flexed because there is an increase in the overall length of the vertebral canal, but the posterior dura mater still remains abutting the posterior wall of the spinal canal. In patients with Hirayama's disease, due to the presumed imbalance in growth between the vertebral column and the dura, the dura mater is already tighter in the neutral position. During neck flexion, the tightened posterior dura mater detaches from the posterior lamina and compresses the spinal cord against the anterior vertebral column. This compression causes ischaemic changes in the territory of the anterior spinal artery, particularly at C4-C6 levels, and hence the subsequent denervation of the distal upper limb muscles.

However, a more recent European study comparing patients with Hirayama's disease and normal individuals has shown results that argue against a neck flexion–induced myelopathy, and instead support the view that Hirayama's disease is an intrinsic motor neuron disease.[3]

What most researchers agree is that Hirayama's disease has a self-limiting course and that it reaches a stable stage after a period of initial deterioration over a few years. Current treatment involves the use of a neck collar to reduce repeated trauma to the cervical cord from neck flexion and to prevent further progression of the disease.[8] Other treatment options include surgery, namely anterior decompression and fusion, as well as posterior fusion.[9]

It is thought that Hirayama's disease is underdiagnosed. In a young patient with clinical evidence of hand and / or forearm muscle atrophy and asymmetric thinning of the cervical spinal cord, a diagnosis of Hirayama's disease should always be considered.[4] Conventional radiographic studies of the cervical spine usually show no specific abnormalities except straight alignment or scoliosis.[6] Non-flexion MRI of the cervical spine may only show subtle asymmetric cord atrophy in the lower cervical region and this can be overlooked. Chen et al[10] investigated findings seen in neutral position

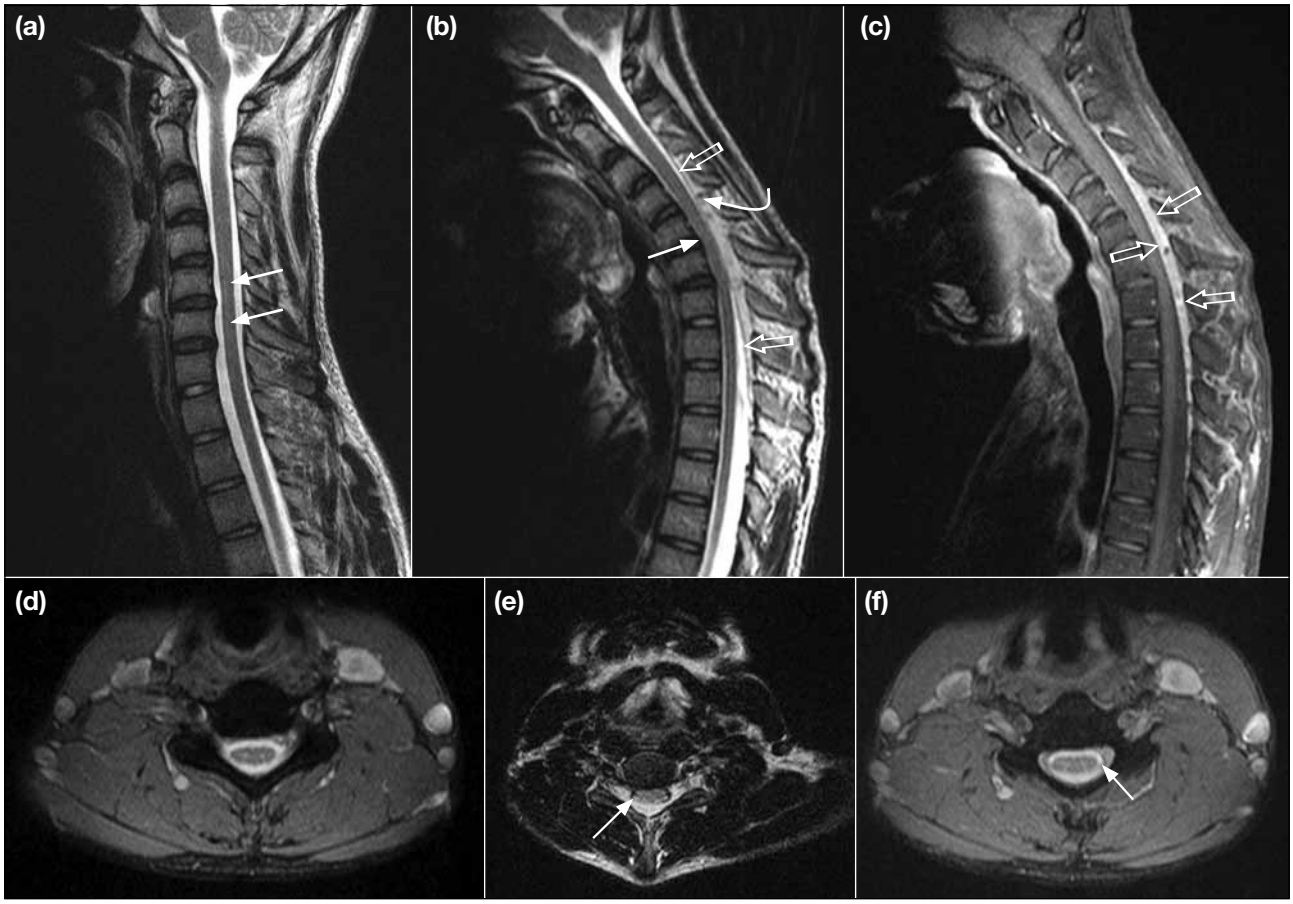

Figure. (a) A T2-weighted sagittal image of the cervical spine in neutral position shows loss of normal cervical lordosis. There is no intramedullary or extramedullary lesion causing cord compression. Mild cord atrophy at C5/6 levels is noted (arrows). (b) In the fully flexed position, the lower cervical cord is abutting on the posterior border of the C5 and C6 vertebral bodies (long straight arrow). The posterior dura is detached (open arrows) and displaced forwards at the C3 to T2 level. Flow voids are seen within the posterior epidural space (curved arrow). (c) A post-gadolinium image shows intense enhancement posterior to the detached dura due to enhancement of the engorged posterior epidural venous plexus (open arrows). (d) A T2*-weighted axial image of cervical spine (C6 level) in neutral position. (e) A T2-weighted axial image at the same level in full flexion position, showing anterior displacement and flattening of the lower cervical cord (arrow). (f) A T2*-weighted axial image of cervical spine at C6 level in neutral position shows loss of attachment between the posterior dural sac and subjacent lamina (arrow).

cervical MRI of patients with Hirayama's disease, and highlighted the value of loss of attachment between the posterior dural sac and subjacent lamina in suggesting the diagnosis (Figure f). This finding, together with a high index of clinical suspicion and MRI of the cervical spine with neck flexion, may be the key to correct diagnosis.

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