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CASE REPORT

Concomitant Unilateral Hirayama Disease and Contralateral Disc Protrusion of the Cervical Spine

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ABSTRACT

Hirayama disease is a type of cervical myelopathy affecting young men and causing wasting of the small muscles of the hand. The underlying pathophysiology is thought to be due to disproportionate growth of the vertebral column and the spinal canal resulting in a tight dural sac. The presentation of Hirayama disease is usually that of muscular atrophy with no sensory symptoms. Hirayama disease is not progressive. Magnetic resonance imaging with flexion views is required to make the diagnosis, and demonstrates the distended epidural venous plexus that results from the disproportionate growth and compresses on the spinal canal. The distended venous plexus can be unilateral or bilateral, but is usually unilateral. The underlying cause is unknown, and it is not known why Hirayama disease is usually unilateral. This report presents a patient with a magnetic resonance diagnosis of Hirayama disease with atypical clinical features. Magnetic resonance imaging with flexion views confirmed unilateral Hirayama disease and concomitant contralateral disc protrusion. We propose that these two findings may be inter-related.

Key Words: *Amyotrophy, monomelic; Intervertebral disc displacement; Magnetic resonance imaging; Muscular atrophy; Spinal cord diseases*

中文摘要

單側平山病併發對側頸椎椎間盤突出症

陳煥章、曾佩琪、袁銘強

平山病是一種頸部脊髓病變，會造成手部小肌肉萎縮；多好發於年輕男性。平山病的發病機制認為與脊柱和椎管之間的發育不平衡並壓迫硬脊膜有關。平山病不會惡化，典型的表現為肌肉萎縮，一般沒有疼痛等的感覺障礙表現。診斷用屈曲位磁共振成像（MRI），表現為因發育不均衡而導致脊硬膜壁被拉直而壓迫脊椎。擴張的靜脈叢多為單側，雖然也可累及雙側。平山病的致病機制仍不甚明確，亦不清楚為何單側受累多見。本文報告一名呈現非典型臨床症狀的平山病患者的MRI診斷。屈曲位MRI證實為單側平山病，並伴隨對側頸椎椎間盤突出。這兩個診斷表現可能互相關聯。

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INTRODUCTION

Hirayama disease is a type of cervical myelopathy related to flexion movements of the neck. It is described more among Asian ethnic groups and commonly affects young men. Hirayama disease is known to be non-progressive. Patients typically present with muscle wasting of the small muscles of the hand without sensory involvement.

This report is of a patient with atypical clinical features and disease course. Magnetic resonance imaging (MRI) with flexion views confirmed unilateral Hirayama disease and concomitant contralateral disc protrusion. It is not known why the disease is usually unilateral. We propose that these two findings may be inter-related.

CASE REPORT

The patient was a 33-year-old woman, who drank alcohol and smoked. She first presented in 2003 with wasting of the small muscles of the right hand and mild right upper limb numbness. Physical examination and nerve conduction study suggested C8-T1 radiculopathy. The muscle wasting progressed more proximally to involve the muscles of the forearm. MRI only showed mild central disc protrusion at the C6/7 level without significant compression. MRI also ruled out any brachial plexus lesions. A muscle biopsy was negative.

The cause of the condition was not known at the time. The disease became static after around 2 years. The patient was then lost to follow-up.

The patient came to medical attention again in 2009. The weakness had progressed to involve the arm. Physical examination showed right-sided limb weakness with muscle power grade 4/5 in the C5-7 myotomes, 3/5 in the C8 myotome, and 2/5 in the T1 myotome. Nerve conduction studies and electromyogram showed chronic neurogenic changes in the right C7-T1 myotomes and subacute neurogenic changes in the right C5-6 myotomes.

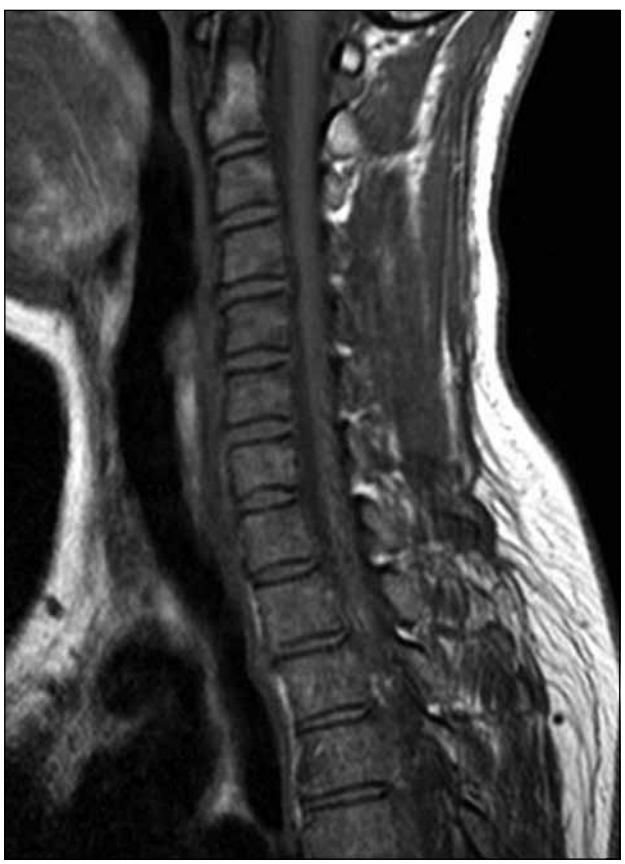

Figure 1. A sagittal T1-weighted magnetic resonance image in the neutral position.

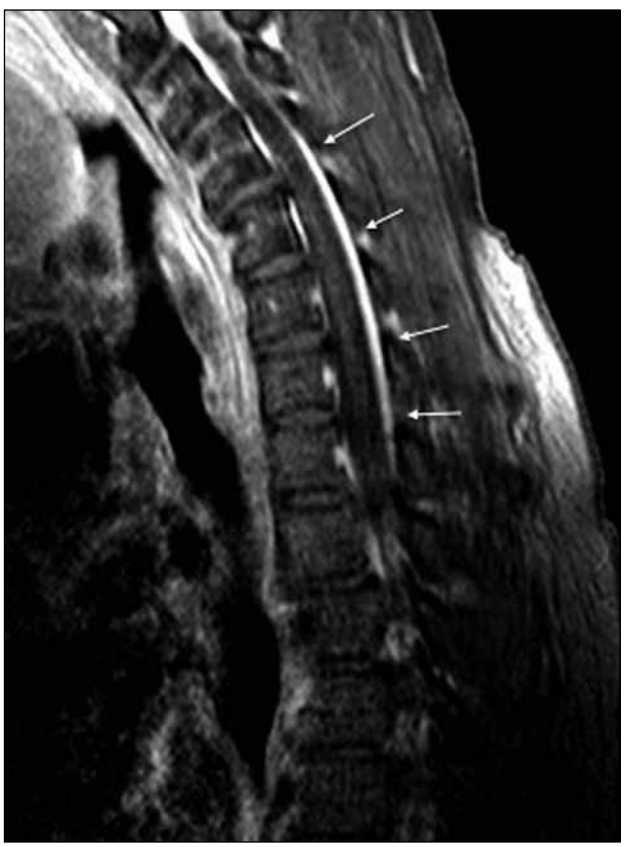

Figure 2. A sagittal T1-weighted magnetic resonance image with contrast in the flexion position shows peridural enhancement (arrows).

MRI with flexion views showed abnormal increased contrast enhancement of the posterior right epidural space from C4 to C8. There was forward shift of the spinal cord, slightly towards the left side. A broad-based left paracentral disc protrusion was seen at the C5/6 level with narrowing of the thecal sac. Milder right paracentral disc protrusion was also seen at the C4-5 level and central disc protrusion at the C6-7 level. The imaging features were compatible with unilateral right-sided Hirayama disease with concomitant disc protrusion, with the most significant narrowing on the left side (Figures 1 to 5).

DISCUSSION

Hirayama disease is a type of cervical myelopathy related to flexion movements of the neck. Hirayama disease was first described by Hirayama and Tokumaru[1] in 1959 as a sporadic juvenile muscular atrophy of the upper limb predominantly affecting young men. The condition is characterised by an insidious unilateral or asymmetric bilateral muscular atrophy and weakness of the hand and forearm without sensory or pyramidal signs.[1] Hirayama disease is a benign disorder with a stable stage after a progressive course.

The proposed pathophysiology of Hirayama disease is disproportional growth between the vertebral column and the spinal canal resulting in a tight dural sac. On flexion of the spine, which normally increases the length of the cervical canal, the tight dural sac cannot increase in length and must displace anteriorly as a compensatory mechanism. This anterior movement of the dural sac compresses the spinal cord.[2] The compression is most severe at the most kyphotic level usually at the lower

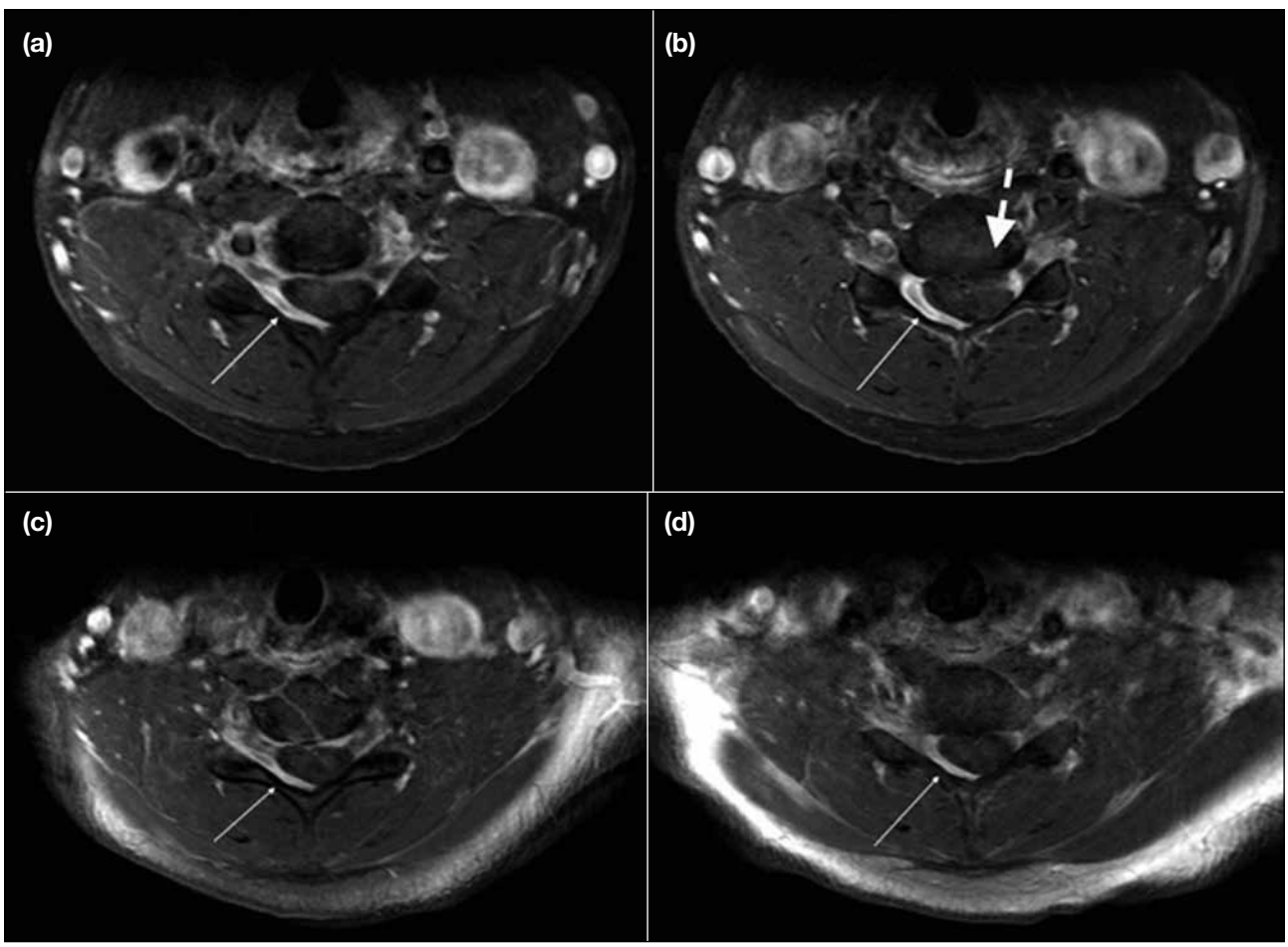

Figure 3. Serial axial T1-weighted magnetic resonance images with contrast show epidural enhancement at the right posterior aspect of the spinal canal (arrows) — the cord is shifted to the left; disc protrusion at the left paracentral aspect (dotted arrow), at which level the cord is most compressed.

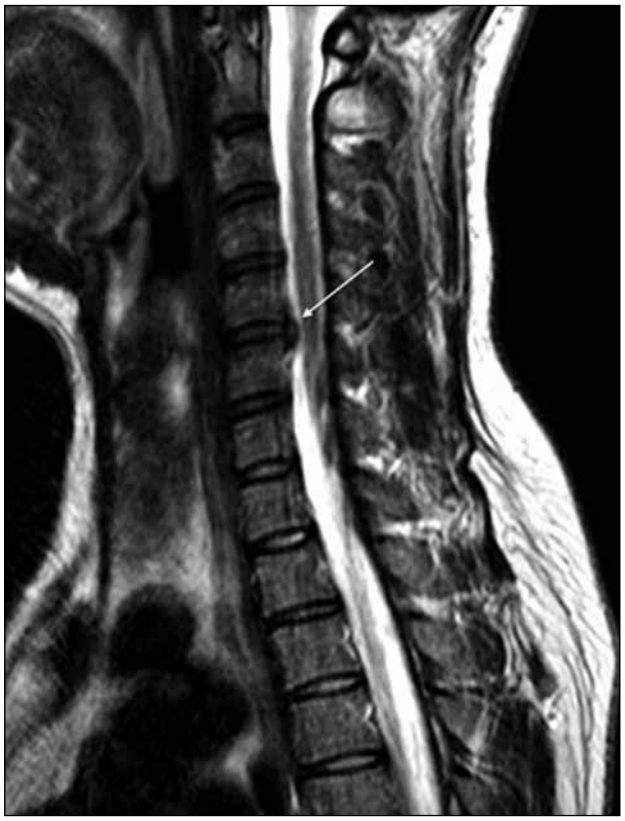

Figure 4. A right parasagittal T2-weighted magnetic resonance image shows a protruded disc at the left paracentral aspect (arrow).

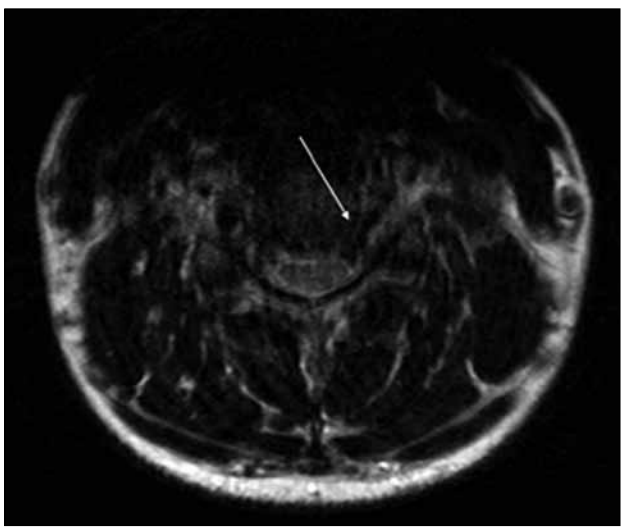

Figure 5. An axial T2-weighted magnetic resonance image shows left paracentral and foraminal disc protrusion (arrow). The cord shows no abnormal signal and no significant atrophy.

cervical spine. The repeated compressive effects cause ischaemia to the anterior horn cells at that level, whereas the white matter tracts are spared.[3]

MRI study in neck flexion shows the forward displacement of the posterior wall and a well-enhanced crescent-shaped mass in the posterior epidural space of the lower cervical canal. This mass is thought to represent congestion of the posterior internal vertebral venous plexus and vanishes once the neck returns to a neutral position.[2] This finding, however, needs to be interpreted with caution as it has been shown to be present in healthy individuals.[4]

Our patient with Hirayama disease was not diagnosed when she first presented because Hirayama disease was not suspected at the time and MRI in flexion posture was not performed to confirm the diagnosis. Her condition rapidly progressed in the first 2 years, but was then relatively stable, which is in keeping with the clinical course reported from previous studies.[1,5] However she presented again after around 5 to 6 years with slight worsening of symptoms with muscle wasting progressing more proximally and disabling pain of the right upper limb, although there was no sensory loss. This is contrary to the typical description of Hirayama disease, which is expected to be stable with purely motor symptoms.

On MRI, epidural enhancement likely representing an engorged venous plexus was seen on the right side, corresponding to the side of the patient's symptoms. Previous reports of the disease were commonly unilateral, sometimes bilateral asymmetrical and, rarely, bilateral symmetrical.[6] The reason for its one-sidedness is not clear. In this patient, there were concomitant disc protrusions. The one causing the most severe narrowing of the thecal sac was on the contralateral side of the distended venous plexus. We postulate that the presence of the protruding disc on the contralateral side prevented the development of the distended venous plexus due to 'pressure effects'. To our knowledge, the concomitant presence of cervical disc protrusion and Hirayama disease has seldom been described. It would be interesting to know whether the two conditions are related in the evolution of the disease and further studies are likely to be required.

CONCLUSION

Hirayama disease is a rare cause of cervical myelopathy presenting with non-progressive unilateral upper limb muscle weakness and atrophy commonly affecting men. Presented here is a patient with several atypical features, including female sex, sensory and motor neurological deficits, and apparent progression of untreated disease.

MRI confirmed unilateral Hirayama disease with the presence of a distended epidural venous plexus on flexion as well as a cervical disc protrusion on the contralateral side. The concomitant occurrence of both conditions has not been described in the literature. We propose a possible explanation for the phenomenon of unilateral Hirayama disease in this patient.

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