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Case Report

Venous hypertensive myelopathy associated with cervical spondylosis

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Abstract

BACKGROUND CONTEXT: Venous hypertensive myelopathy (VHM) results from spinal vascular malformations of arteriovenous shunting that increases spinal venous pressure, leading to congestive edema and neurologic dysfunction. There has been no report of VHM associated with cervical spondylotic myelopathy (CSM).
PURPOSE: The aim of this study was to report an extremely rare case of VHM likely due to CSM.
STUDY DESIGN: This study is a case report and review of the literature.
PATIENT SAMPLE: The patient was a 51-year-old man with CSM exhibiting relatively rapid neurologic deterioration with an abnormal expansion of a centromedullary hyperintense lesion on T2-weighted magnetic resonance imaging (MRI) in the absence of traumatic injury.
METHODS: Neurologic examination and radiologic imaging were taken by various means.
RESULTS: The patient developed a cervical radiculopathy, followed by gait disturbance and motor weakness. The MRI of the cervical spine demonstrated spinal canal stenosis due to disc bulging and flavum hypertrophy at the C5/C6 and C6/C7 levels as well as hyperintense area over the C5–C7 levels on T2-weighted images. Although decompression surgery was planned, an acute inflammatory process such as transverse myelitis or demyelinating disease other than cord compression was also considered, and the patient received intravenous steroids. His walking improved for several days. However, his symptoms then became significantly worse, and he had difficulty walking. Subsequent MRI demonstrated marked progression of the T2 hyperintense lesion over the C4–T1 vertebral levels. Flow voids were also noted on the dorsal surface of the upper cervical cord on T2-weighted MRI. His lab work, medical history, and the local enhancement on contrast-enhanced MRI indicated low probability of spinal inflammatory diseases. Therefore, the decision was made to perform anterior cervical discectomy and fusion surgery on two levels. Following surgery, his symptoms improved promptly.
CONCLUSIONS: Our case indicates that VHM could be caused by spondylotic cord compression in the absence of spinal vascular malformations. The diagnostic features for VHM are progressive deterioration of myelopathy, easing/worsening of symptoms associated with postural changes, and centromedullary hyperintensity over multiple segments and the flow voids on dorsal surface of the spinal cord on T2-weighted MRI. © 2016 Elsevier Inc. All rights reserved.

Keywords: Ascending myelopathy; Diagnosis; Magnetic resonance image; Spinal inflammatory diseases; Spondylosis; Venous hypertensive myelopathy

Introduction

Cervical spondylotic myelopathy (CSM) is a common cause of compressive spinal cord dysfunction. Degenerative changes including disc bulging, osteophytes, and hypertrophy of posterior spinal ligament, reduction in canal diameter, and compression of the spinal cord, which can cause chronic neural injury and ischemia. The typical course of CSM is a relatively slow, progressive, and stepwise deterioration in

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neurologic function with stable plateau periods [1,2]. Rapid progress of neurologic deterioration of patients with CSM is fairly rare, and most cases are associated with minor trauma or segmental instability [3]. Even in such cases, signal changes are not often observed on MRI. Here, we report an extremely rare case of a patient with CSM exhibiting relatively rapid neurologic deterioration with an abnormal expansion of the high signal intensity area on T2-weighted MRI in the absence of traumatic injury.

Case presentation

A 51-year-old man, with no significant medical history including infections, presented with a tingling sensation in his left thumb and index finger with spontaneous onset. Within a week, his symptoms progressed, and he began to have difficulty with fine motor movements and weakness of his left hand. At 3 weeks after onset, he developed weakness of his left upper and lower extremity and gait disturbance with increased tone and stiffness. He got bilateral arm and leg numbness when fully supine position after several minutes. He slept on his right side because sleeping on his left side also caused numbness in the left arm and leg. He was examined by a neurosurgeon, and imaging studies were ordered. Plain radiographs demonstrated moderate degenerative spondylotic changes without evidence of instability. Magnetic resonance imaging demonstrated spinal canal stenosis at the C5/C6 and C6/C7 intervertebral levels as well as a relatively large area of high intensity area on T2-weighted images (Fig. 1). There were no signal changes in the spinal cord on T1-weighted MRI. Although the decompression surgery was planned, an acute inflammatory process, such as a transverse myelitis or demyelinating disease other than cord compression, was also considered given MRI findings. His reflexes were increased in both upper extremities, and Hoffman sign as well as Babinski were positive on the left side. He received intravenous steroids, and his walking improved for 1–2 days. However, his symptoms then worsened, and he had significant difficulty with walking. He then consulted with a neurologist, and repeat MRI of the cervical spine demonstrated marked progression of T2-weighted high signal area over the C4–T1 vertebral levels (Fig. 2). T2 high-intensity area was also observed in the right side of spinal cord in the axial section at the C5 vertebral level (Fig. 2B). However, the patient did not exhibit symptoms on his right side. Contrast-enhanced magnetic resonance (MR) images demonstrated the cord enhancement at C5/C6 level (Fig. 3, Top left), and only the left hemicord was enhanced in the axial plane, although there was no significant laterality in canal narrowing at this level (Fig. 3, Bottom left). To rule out the myelitis, he was tested for Lyme and neuromyelitis optica, and additional imaging of the whole spine and brain was performed. However, no abnormalities were noted in his subsequent work up, and he was then transported to our institution. Differential diagnoses included compressive myelopathy, transverse myelitis, demyelinating disease, and, less likely, tumor. After being

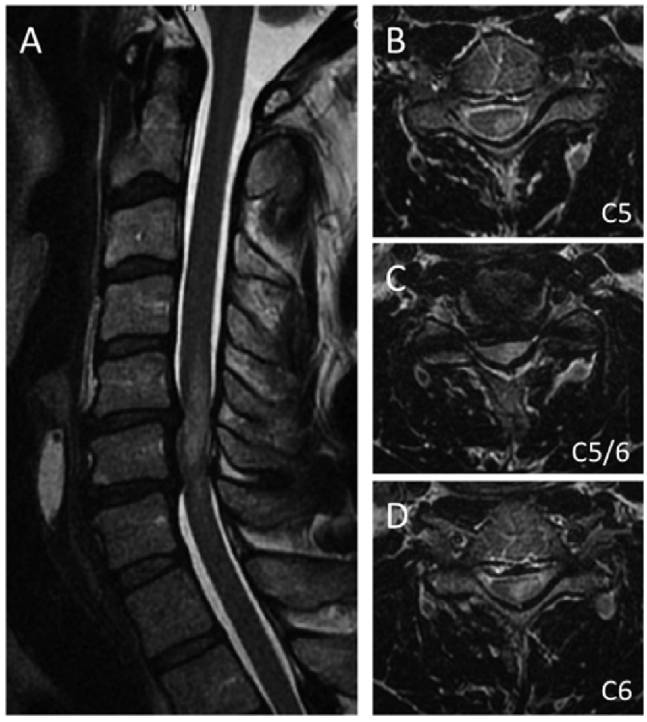

Fig. 1. Cervical magnetic resonance (MR) images before rapid deterioration. (A) Sagittal images demonstrate both anterior and posterior indentations at C5/C6 and C6/C7 levels as well as the T2-weighted high-signal intensity area. (B–D) Axial images show circumferential effacement of subarachnoid space at C5/C6 and T2-weighted high-signal intensity area at the left side of cord.

evaluated again by the neurology, neurosurgery, and orthopedic spine services, it was determined that the presence of his cervical disc herniations at C5/C6 and C6/C7 had some harmful impact on the cord edema, resulting in progressive neurologic deterioration. In addition, flow voids were noted on the dorsal surface of the upper cervical cord on T2-weighted MRI of whole spine (Fig. 3, Right), which is very specific for spinal venous hypertensive myelopathy (VHM) [4]. He agreed to undergo the operative decompression despite a higher risk of further neurologic deterioration in patients with pre-existing cord injury. A two-level anterior cervical discectomy and fusion (ACDF) with anterior plate was performed. Both the surgical and postsurgical courses were uneventful. After operation, his gait disturbance was significantly improved although his partial loss of dexterity in the left hand remained.

Discussions

We report an extremely rare case of ascending myelopathy presenting with rapid neurologic deterioration as well as abnormal expansion of high signal intensity area on T2-weighted MRI. In this case, coexisting transverse myelitis or multiple sclerosis other than compressive myelopathy was suggested by the enlargement of the T2 high-intensity area. However, the localized enhancement on contrast-enhanced

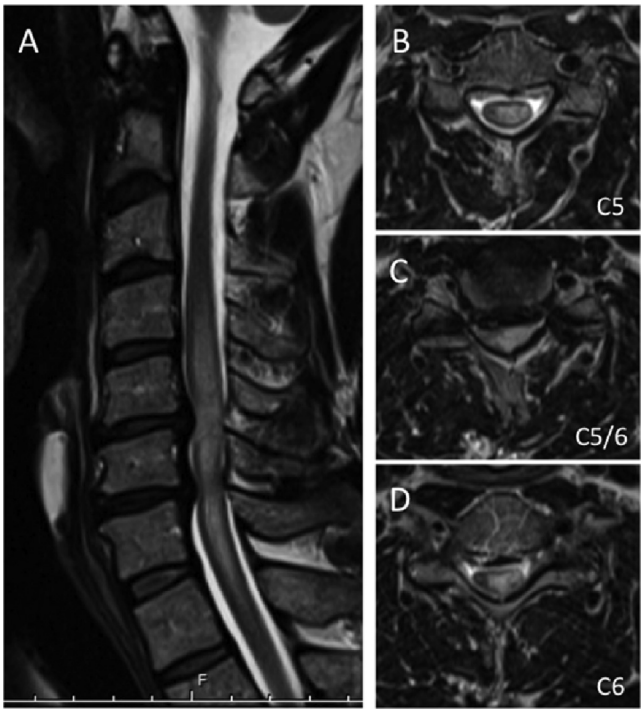

Fig. 2. Cervical magnetic resonance (MR) images after rapid deterioration. (A) Sagittal images demonstrate marked expansion of T2-weighted high-signal intensity area although the extent of cord compression did not change. (B–D) Axial images demonstrate T2-weighted high signal intensity area, which is mostly located in the gray matter.

MRI, as well as negative lab work and patient's medical history, indicated low probability of systemic transverse myelitis (Fig. 3, Top left, Bottom left). Also, high intensity area on T2-weighted MRI was observed mainly in gray matter (Figs. 1 and 2), which expressed skepticism for an inflammatory demyelinating disease as areas of high T2 signal are usually observed in the white matter of both brain and spinal cord in multiple sclerosis patients [5]. Therefore, we considered that the cord compression had some degree of harmful impact on the spinal cord, such as blood supply impairment, and decompression surgery of two-level ACDF was performed. After operation, gait disturbance and muscle weakness promptly improved, and MRI taken 8 weeks postsurgery exhibited small local T2 high-intensity area, strongly suggesting that the physical compression of the spinal cord was the probable cause for the initial neurologic deterioration and the abnormal expansion of T2 high-intensity area.

In addition to the T2 expansion, we also confirmed the flow voids on the dorsal surface of the upper cervical cord on T2-weighted MRI after neurologic deterioration (Fig. 3, Right). It is notable that these were quite similar to the characteristic MRI appearance frequently observed in spinal dural arteriovenous fistulas (AVFs) [4,6]. Arteriovenous fistula represents arteriovenous shunts that prevents efficient draining of the spinal cord, resulting in enlargement and engorgement of the cord venous system. This congestive spinal edema can progress to cord ischemia and lead to acute or subacute deterioration of neurologic function, namely VHM. One of the characteristic MR images of VHM is reversible diffuse T2 hyperintense lesion extending usually more than four segments as well as flow voids on the dorsal surface of the cord on T2-weighted images [4,6,7]. As these MR findings reflect systemic spinal cord edema and abnormal spinal venous flow due to the impairment of venous drainage, we hypothesized that cervical canal stenosis and cord compression in our case impaired spinal venous system, resulting in progressive myelopathy.

In addition, the course of neurologic deterioration and MRI findings of our patients were similar to those of patients with subacute posttraumatic ascending myelopathy (SPAM). This disorder is very rare, but well known to be a disease exhibiting neurologic deterioration as well as marked enlargement of intramedullary high intensity signal area on T2-weight MRI after spinal cord injury. Patients with SPAM usually suffer significant neurologic deterioration after a few days of clinical stability of after spinal cord injury with MR findings of extensive high-T2 signal, spanning more than three segments from the initial injured segment. Although the pathophysiology behind SPAM remains is still unknown, several recent case reports indicate the involvement of spinal

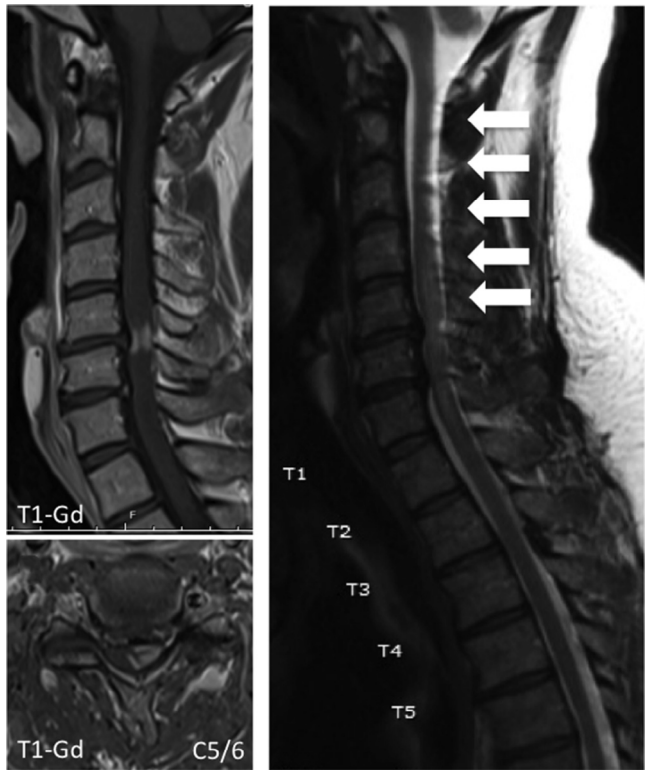

Fig. 3. Cervical magnetic resonance (MR) images postcontrast after rapid deterioration did not demonstrate the expansion of myelitis. (Top left, Bottom left) Sagittal and axial postcontrast images demonstrate the localized enhancement at the left side of spinal cord at the C5/C6 level. (Right) Sagittal image from the whole spine MR demonstrates, in addition to the T2-weighted high-intensity signal area in the spinal cord, that flow voids are present in the dorsal surface of the cord at the C2–C5 levels (arrows).

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venous drainage impairment and hypertension [8–10]. The following points support venous hypertension as a possible cause of SPAM: neurologic level of deterioration dose not correspond with the T2 high area; neurologic deficit as well as the MRI changes ease overtime; postural changes are often associated with the easing or worsening of symptoms; T2 flow voids on the dorsal surface of the cord. In our case, all of these characteristics were observed, strongly supporting that the pathologic mechanism in our case was closely associated with the impairment of spinal venous drainage.

Generally, VHM is considered to be inseparable from spinal vascular malformations including spinal dural AVF and spinal arteriovenous malformations. However, we present a rare case of cervical spinal spondylosis-related VHM. In fact, Krishnan et al, reported that non-compressive C5/C6 disc herniation brought about VHM and its MRI was very similar to AVF, and that the myelopathy recovered completely by C5/C6 ACDF [11]. Also, Auler et al reported a case of traumatic mediastinal hematoma caused by compression of the brachiocephalic vein, resulting in VHM, which resolved after the resolution of the mediastinal hematoma [12]. It is also probable that the pathophysiology of SPAM overlaps with that of spinal VHM, as Tan et al suggested that subacute delayed ascending myelopathy is a more appropriate terminology as it is not just a posttraumatic disorder [10]. These studies suggest that VHM can be developed by various causes even in the absence of vascular anomalies. We suggest that some cases reported as rapid deterioration of CSM with an unknown cause might actually be secondary to VHM [13,14].

Reversibility of both imaging abnormality and neurologic deterioration after resolution of venous hypertension is a clinical characteristic of VHM. However, persistent increase of the intraspinal pressure would lead to breakdown of the venous system and intramedullary hemorrhage, resulting in the irreversible spinal lesion and severe permanent paralysis [9]. Therefore, prompt diagnosis and intervention to reduce intraspinal pressure before irreversible damage develops are crucial.

In conclusion, our case indicates that the VHM could be caused by spondylotic cord compression in the absence of spinal vascular malformations. The diagnostic features on MRI for VHM are spinal cord swelling, abnormal expansion of intramedullary high intensity area, and flow voids on the dorsal surface of the spinal cord on T2-weighted MRI. Also, in the case of progressive deterioration of myelopathy with discrepancy between the T2 hyperintensity area and the neurologic deficit level as well as in the presence of easing/worsening myelopathy associated with postural changes, the physician should include VHM in the differential diagnosis.

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