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Spinal Dural Arteriovenous Malformation Confused with Progressive Myelopathy: A Walking Disorder that Alternates in Symptoms of Wax and Wane

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Abstract

Spinal dural arteriovenous fistula (SDAVF) is a rare vascular malformation of the spine. Those who have spinal dural arteriovenous malformation (SDAVM) in their thoracic or lumbar spine may suffer from severe neurological problems. A 66-year-old man, who felt weakness in his legs when hiking, checked into an emergency room and did not find any neurological problems according to MR imaging. Later on, however, we were able to detect SDAVF affecting both the thoracic and lumbar spine, using spine MRI with enhancement. For treatment of his arteriovenous malformation, total laminectomy was performed from T12 to L2 as well as feeding artery ligation with clipping. Eventually, the patient's condition improved and he was able to resume hiking.

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Introduction

A spinal dural arteriovenous malformation (SDAVM) tends to confuse the clinician because it involves a fluctuation of various symptoms including progressive gait disturbance, motor weakness, sensory loss, and bladder, bowel, and sexual dysfunction [1,3]. Therefore, it is easy to overlook the diagnosis and treatment of SDAVF, resulting in considerable morbidity as well as the progression of neurological problems [2,3]. However, even if SDAVM is confused with cervical spondylosis or myelopathy and treated as such, we can still expect a good improvement of the symptoms [3].

Case Representations

Clinical history and neurological examination:

A 66-year-old Korean male has suffered from recurrent gait disturbance since October 2012. His past medical history was significant for hypertension and benign prostate hyperplasia. He stopped smoking 2 years ago, although he used to be a heavy smoker, going through as many as 30~40 packs in a year. In addition, he drank alcohol socially. Also, he was a farmer until he retired a few years ago, and now he enjoyed hiking in his spare time. He first felt bilateral lower extremity weakness in October 2012, and two months later, he felt a progression of this weakness as well as urinary retention while he was shoveling snow in his garden. His symptoms continued to worsen and it became difficult for him to even walk five or six steps, which motivated him to visit the emergency room. The patient's physical examination was absolutely normal with a blood pressure of 120/80 mm Hg and a regular heart rate. His neurologic examination showed normal results for mental status, cranial nerve function, as well as for upper extremity motor and sensory examinations. However, his legs exhibited a mild weakness in his proximal muscles, although not for his distal muscles. The Medical Research Council (MRC) strength scale revealed IV/IV for hip flexion and extension. Also, the sensory examinations of the lower extremities showed hyperesthesia below T4 level on the dermatome and demonstrated hypoesthesia below L2 level. Furthermore, all sensory modalities such as pain, temperature, light touch and vibration were decreased on lower extremities. There was about 30 percent of sensation on the right lower leg, compared to 50 percent on the left lower leg. Deep tendon reflexes were pathologically increased at the knee but not at the Achilles tendon; Babinski's sign was absent in both legs. Rectal tone was mildly decreased.

Laboratory and Neuroradiologic finding

His routine complete blood count and chemistry panel were normal. Also, he did not show abnormal thyroid function, Vitamin B12 levels, folate levels, methylmalonic acid levels, homocysteine levels or tumor marker levels for PSA, AFP, CEA and CA 19-9. The patient was negative for autoimmune disease work-up, including anti-dsDNA antibody levels, anti-cardiolipin antibody levels, anti-sm antibody levels, anti-SS-A/B antibody levels, HLA B27 levels for ankylosing spondylitis, HLA B51 levels for Behçet's Disease, hepatitis B and C, as well as HIV. The chest X-ray in the ER showed non-specific features (Figure1) and his evoked potential study revealed abnormalities between the thoracic cord and the cerebral cortex but the nerve conduction study showed no defects. Testing of the cerebrospinal fluid, including the oligoclonal IgG band, gave normal results. Upon admission to the hospital, he regularly climbed the stairs for exercise, which surprisingly worsened the strength of his lower extremities from IV/IV to II/II on the MRC strength scale. At times the patient would be reduced to a state in which he could not walk at all. In these instances, the patient was able to resume walking after approximately 15 minutes of rest. Unenhanced magnetic resonance imaging (MRI) of the thoraco-lumbar spine depicted edematous change of the thoracic spinal cord (Figure 2a). Also, enhanced MRI of the spine showed a diffusely abnormal hyperintense T1 weighted signal, extending from the T6 vertebral body level down to the L2 vertebral body level (Figure 2b). Furthermore, the T2 weighted images (T2WI) showed high signal intensity on the same lesion (Figure 2c).

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Brain MR images showed multiple small high signal intensity foci in cerebral white matter on T2WI and FLAIR (Fluid-attenuated inversion recovery) and revealed no definite enhancement on brain parenchyma from T1WI (Figure (3a & 3b).

Final diagnosis and management

The patient's MR imaging results showed blood vessels with multiple signal voids in the spinal canal from T1 vertebral level to the lumbar spine level (Figure 4a and Figure 4b).

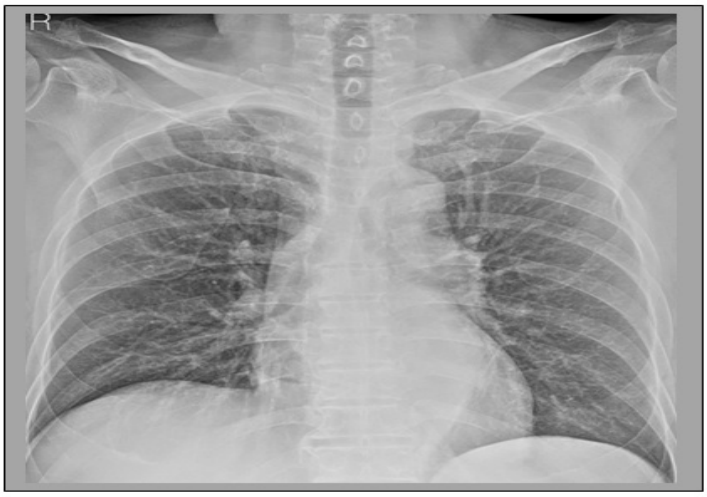


Figure 1: The patient's chest X-ray showed a non-specific feature.

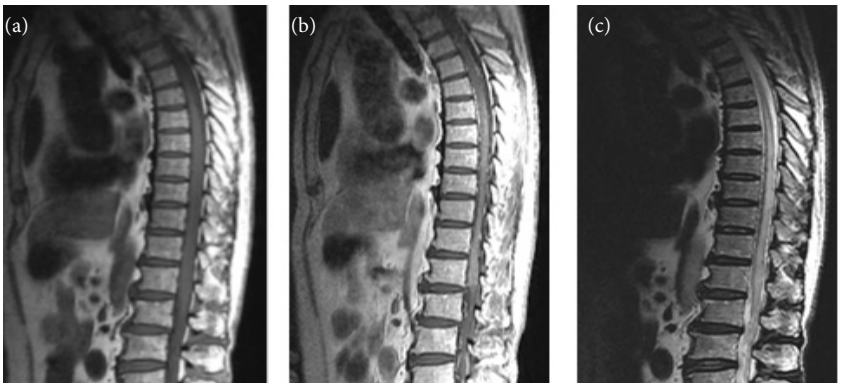


Figure 2: (a) Unenhanced magnetic resonance imaging (MRI) of the thoraco-lumbar spine depicted edematous change in the thoracic spinal cord. (b) Enhanced MRI of the spine showed a diffusely abnormal hyperintense T1 weighted signal extending from the T6 vertebral body level down to the L2 vertebral body level. (c) The T2 weighted images of his vertebrae showed high signal intensity at the same lesion.

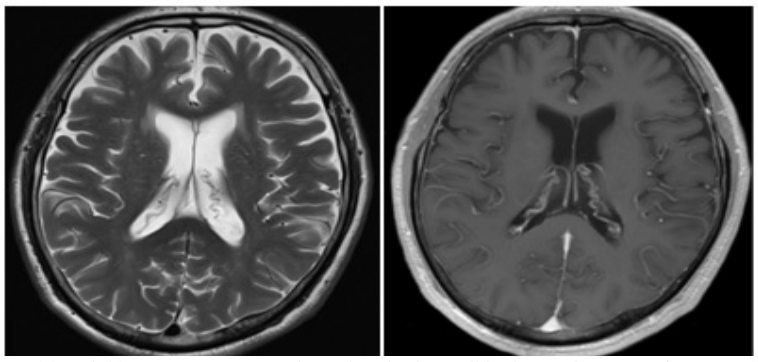


Figure 3(a) and (b): Brain MR images showed multiple small high signal intensity foci in cerebral white matter on T2WI and revealed no definite enhancement of the brain parenchyma from T1WI.

Also, the T2 weighted images of the spine MR depicted high signal intensity from T6 vertebral level to L2 vertebral level. Furthermore, a spinal arteriogram revealed spinal dural AVF with a feeding artery arising from the L1 segmental artery (Figure (5a & 5b)).

For treatment of his spinal arteriovenous malformation, a laminectomy was performed from T12 to L2 vertebrae, with clipping and ligation of the feeding artery (Figure 5c). First, the fistula site was confirmed to be at T12 level since the feeding artery originated from the left segmental artery, passed through the left neural foramen at L1-2 level, and moved into T12 intradural region. Then a temporary clip was placed on the feeding artery for 10 minutes, and there were no abnormal changes in SEP and MEP during this time. After conducting indocyanine green (ICG) video angiography to confirm the absence of blood flow to the engorged vein, the clip was removed and bipolar coagulation was performed on the feeding artery. Finally, a hemoclip was applied and ligation was performed. As a result of the procedure, the patient's condition improved and he was able to resume hiking.

Discussion

Spetzler et al. suggested three categories of classification for spinal vascular lesions: neoplasms, aneurysms, and arteriovenous lesions [6]. Spinal Dural Arteriovenous Fistulas (SDAVFs), a type of arteriovenous lesions, represent a surgically treatable form of nontraumatic myelopathy with an annual incident rate of approximately 10 cases per 1 million people [7]. Most patients diagnosed with SDAVF are male (~80%) and between the ages of 60 and 70 years (~66%) [7]. 1% of patients also present with subarachnoid hemorrhage [7]. In addition, a study of 80 patients revealed the median time to diagnosis to be 15 months [7].

SDAVFs are easily overlooked because they have overlapping symptoms with those of more common causes of myelopathy such as cervical spondylosis [3]. SDAVFs are predominantly found at the lower thoracic and upper lumbar levels with presenting symptoms secondary to myelopathy along with back, radicular and nonspecific pain [7]. The arteriovenous shunt is generally located inside the dura

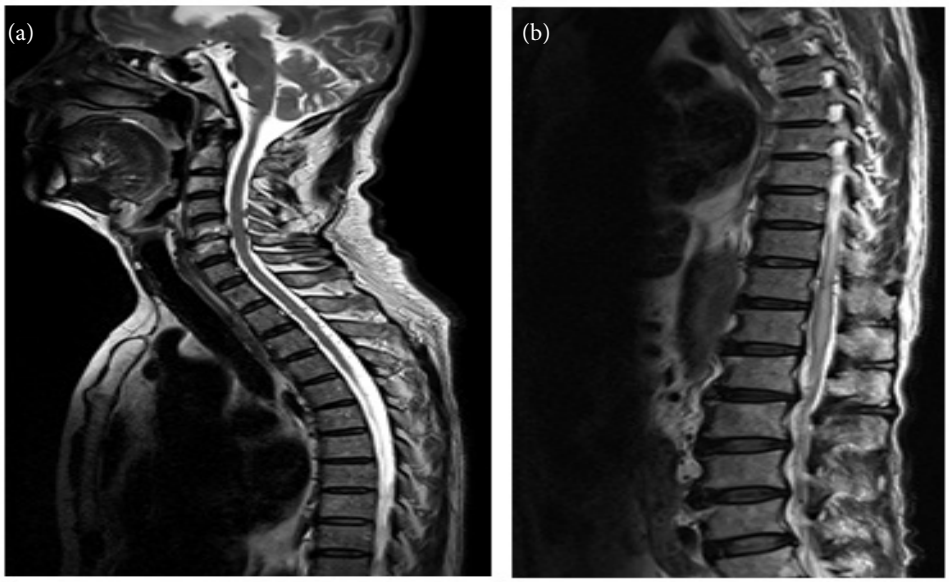
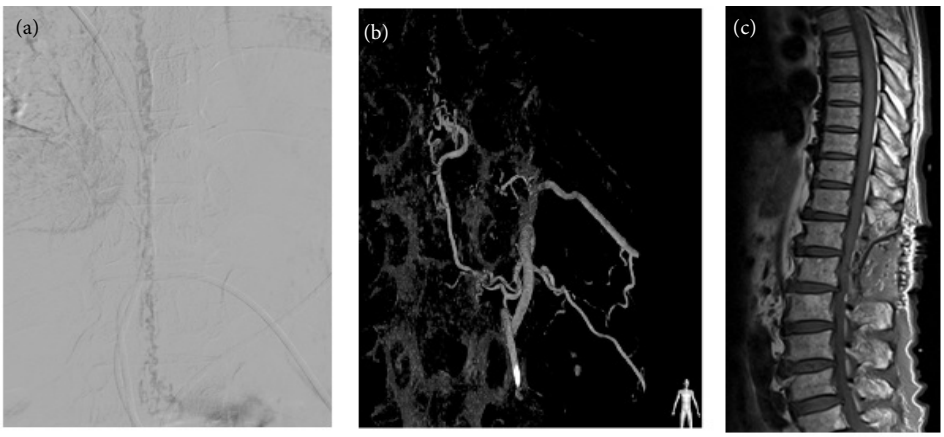


Figure 4: (a & b) Whole spine MR images indicated suspected multiple vascular flow voids in the spinal canal from T1 vertebra level to the lumbar vertebral level.



Figures 5: (a & b) Spinal arteriography showed spinal dural AVF with a feeding artery from the L1 segmental artery. (c) Laminectomy was performed from T12 to L2 vertebrae with clipping and ligation of the feeding artery.

mater close to the spinal nerve root where the arterial blood from a radiculo-meningeal artery enters a radicular vein [1]. Spinal venous pressure is increased, resulting in venous congestion and drainage of normal spinal veins, which ultimately reveals the clinical findings of progressive myelopathy [1]. Some characteristic findings of MR imaging would be the combination of cord edema, perimedullary dilated vessels, and cord enhancement [1].

Therapeutic approaches may include the occlusion of the shunting zone, either by super-selective embolization with a liquid embolic material or by a neurosurgical approach [1]. The problematic arteriovenous malformations can be stopped through ligation or closure of the fistula [1]. It is important for us to understand that SDAVFs are a treatable cause for progressive para- or tetraplegia even if the exact etiology remains unknown [1].

Competing Interests

The authors declare that they have no competing interests.

References

1. Krings T, Geibprasert S (2009) Spinal Dural Arteriovenous Fistulas. Am J Neuroradiol 30: 639-648.
2. Krings T, Lasjaunias PL, Geibprasert S, Hans FJ, Thron AK, et al. (2009) Classification of Spinal Vascular Malformations. The Neuroradiology Journal 22: 97-106.
3. Spain RI, Stucrert E, Sharan A, Sridmore CT (2009) Spinal Dural Arteriovenous fistula: An Overlooked Cause of Progressive Myelopathy. Hospital Physician.
4. Koch C (2006) Spinal dural arteriovenous fistula. Curr Opin Neurol 19: 69-75.
5. Apostolova M, Nasser S, Kodsi S (2012) A rare case of spinal dural arteriovenous fistula. Neurol Int 19: 87-89.
6. Rosenblum B, Oldfield EH, Doppman JL, Chiro GD (1987) Spinal arteriovenous malformations: a comparison of dural arteriovenous firstulas and intradural AVM's in 81 patients. J Neurosurg 67: 795-802.
7. Park KW, Park SI, Im SB, Kim BT (2009) Spinal Dural Arteriovenous Fistula with Supply from the Lateral Sacral Artery-Case Report and Review of Literature. J Korean Neurosurg Soc 45: 115-117.