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Case Report

SPINAL DURAL ARTERIOVENOUS FISTULA: A RARE CAUSE OF PROGRESSIVE PARAPARESIS

Abstract

Spinal dural arteriovenous fistula (dAVF) is a rare and disabling but potentially treatable vascular malformation of the spine. The lesion is more common in men over the age of 50 years and affects the thoracic and lumbar spine. These patients usually present with slow and progressive paraparesis with both upper and lower motor neuron lesion findings, which hinders the clinical differentiation of spinal dAVF from polyneuropathies and other causes of myelopathies. Available treatment includes endovascular embolization and microsurgical techniques. Because a patient's prognosis is strongly correlated with the time of the diagnosis and early treatment, spinal dAVF should always be considered in the differential diagnosis of patients with paraparesis. A 65-year-old male patient was admitted to our clinic with a 3-month history of progressive bilateral lower extremity weakness. Neurological examination revealed hypoesthesia in the L4 dermatome bilaterally, and the strength in the bilateral hip flexors and knee extensor muscles was 4/5. On contrast magnetic resonance imaging, vascular structures surrounding the dural sac were prominent in the lumbar and thoracic regions. In angiography a spinal dAVF was detected. Following the worsening of the patient's paraparesis, endovascular embolization was applied to the fistula. Patient's clinic completely restored after the procedure.

In this report, it has been aimed to present a male patient who admitted with progressive paraparesis and diagnosed as subsequent lumbar dAVF in the light of updated literature.

Key Words: Arteriovenous Fistula, Spinal Dural; Paraparesis; Aged; Rehabilitation.

Olgu Sunumu

SPİNAL DURAL ARTERİOVENÖZ FİSTÜL: NADİR BİR PROGRESİF PARAPAREZİ NEDENİ

Öz

Spinal dural arteriovenöz fistüller (sdAVF) omurganın nadir görülen, sekel bırakabilen ancak tedavi edilebilir vasküler malformasyonlarındandır.Genellikle 50 yaş üstü erkek hastalarda lomber ve torakal omurgada gözlenir. Hastalar genellikle yavaş ve progresif seyirli üst ve alt motor nöron lezyonları ile seyreden paraparezi kliniği ile başvururlar ki bu, spinal dAVF'nin diğer polinöropati ve miyelopati nedenlerinden ayırtedilmesini zorlaştırır. Mikrocerrahi teknikleri ve endovasküler embolizasyon mevcut tedavi secenekleridir. Hastanın prognozu tanı zamanı ve erken tedavi ile yakın ilişkili olduğundan spinal dAVF paraparezili hastaların ayırıcı tanısında mutlaka göz önünde bulundurulmalıdır. 65 yaşında erkek hasta kliniğimize 3 aydır devam eden alt ekstremitelerinde güçsüzlük şikayeti ile başvurdu. Nörolojik muayenesinde her iki L4 dermatomunda hipoestezi vardı ve her iki kalça fleksiyon ve diz ekstansiyon kas güçleri 4/5 idi. Kontrastlı manyetik rezonans görüntülemede lomber ve torasik omurgada dural sakı çevreleyen vasküler yapılarda belirginleşme ve anjiografide spinal dAVF saptandı. Hastanın paraparezisinin ağırlaşmasını takiben fistüle endovasküler embolizasyon uygulandı. İşlem sonrasında hastanın kliniği tamamen düzeldi.

Bu yazıda alt ekstremitelerinde progresif güçsüzlük ile başvuran ve spinal dAVF tanısı alan erkek bir hastanın güncel literatür ışığında sunulması amaçlanmıştır.

Anahtar Sözcükler: Spinal Dural Arteriyovenöz Fistül; Paraparezi; Yaşlı; Rehabilitasyon.

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Introduction

Spinal dural arteriovenous fistula (dAVF) is a rare and disabling but potentially treatable vascular malformation of the spine. The resultant venous hypertension decreases spinal cord perfusion, leading to ischemia and edema, which results in slowly progressive myelopathy, sensory disturbances, and bowel and bladder dysfunction. The lesion is more common in men over 50 years of age. It usually affects the thoracic and lumbar spine (1).

Few cases of this vascular malformation have been reported in the literature (2,3). Clinical symptoms and imaging findings are non-specific and can be easily confused with other causes of myelopathy. However, early diagnosis and treatment of patients with dAVF is crucial for prognosis (4).

Here, we describe the case of a male patient who presented with gradually progressive bilateral lower extremity weakness with subsequent lumbar dAVF diagnosis and a literature review.

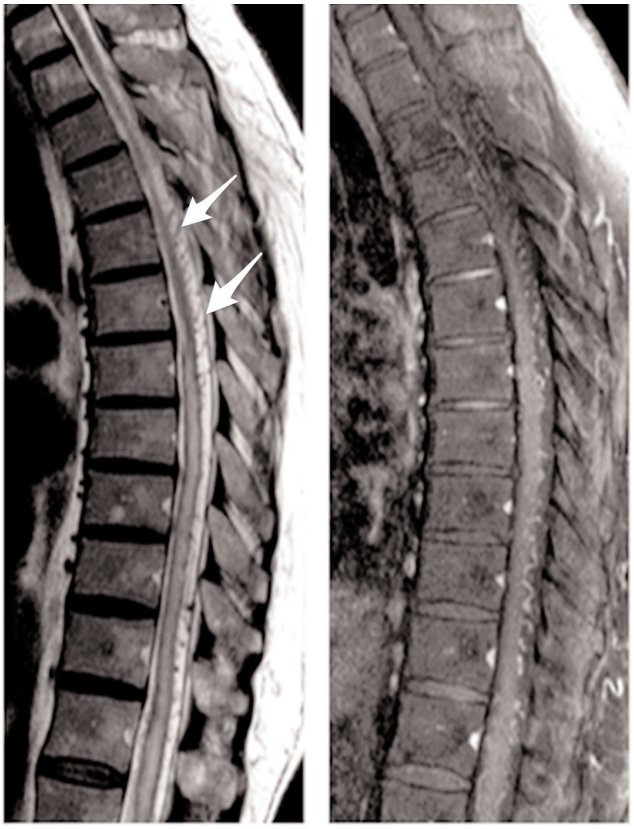
Case Report

A 65-year-old male patient was admitted to our clinic with a 3-month history of progressive bilateral lower extremity weakness with periods of recovery. There was no history of trauma, weight lifting, lower back pain, or neuropathic pain. The patient did not complain of bladder and bowel problems or sexual dysfunction.

On admission, neurological examination revealed hypoesthesia in the L4 dermatome bilaterally, and the strength in the bilateral hip flexors and knee extensor muscles was 4/5. There was no spasticity. Hyperactive deep tendon reflexes were noted bilaterally in the lower extremities. Babinski's response was also positive. Evaluation on admission showed the following findings: Functional Ambulation Scale (FAS), category 5 (independent ambulation) and Functional Independence Measure (FIM) score, 125 (18–126).

Antinuclear antibodies (ANA), anti-phospholipid antibodies, anti-double-stranded DNA antibodies, human immunodeficiency virus, and markers for hepatitis infection were negative on laboratory tests. Erythrocyte sedimentation rate, thyroid stimulating hormone and parathyroid hormone were in normal limits.

Cranial and spinal magnetic resonance imaging (MRI) were planned because of the inconsistent course of paresis and reflex abnormalities. Spinal MRI revealed multiple disc protrusions and an extruded hernia at the level of the T8–9 vertebrae. The diameter of the spinal canal was within normal limits. On contrast imaging, vascular structures surrounding the dural sac were prominent in the lumbar and thoracic regions. Spinal cord edema was observed (Figure 1). Cranial MRI was normal. The patient was evaluated at the Interventional Radiology Clinic and angiography was performed with the suspicion of arteriovenous malformation. A spinal dAVF was detected. It originated from the bilateral lumbar arteries and was supplied with a large number of small arteries at the level L1–2. The spinal dAVF was drained to the inferior vena cava by the spinal perimedullary veins.



A B

Figure 1— A) T2 weighted sagittal MRI shows intramedullary increased signal in the distal spinal cord without associated expansion as well as signal void serpingious structures (arrows) around the spinal cord representing dilated vessels. **B)** Postcontrast T1 weighted sagittal image with fatsat shows contrast enhancement of the dilated vessels.

Patient was scheduled for endovascular embolization at the Interventional Radiology Clinic. After the patient's muscle weakness worsened to 1/5 bilaterally, with complete loss of

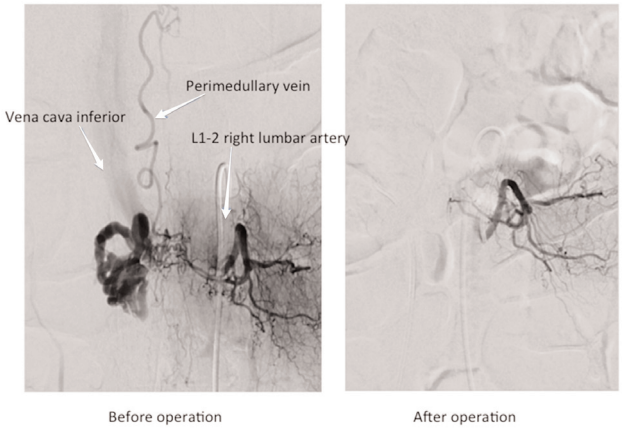


Figure 2— The spinal dural arteriovenous fistula in angiography images; feeding from right lumbar artery at L1-L2 level, draining to perimedullary vein and vena cava inferior.

sensation in the lower extremities, an emergent angiography was performed. Preoperative FAS was 0 (non-functional ambulation) and FIM was 63. Under general anesthesia, the iliac veins and inferior vena cava were accessed by entering from the right common femoral vein. The fistula was also viewed with arterial catheterization. After passing the level of the fistula transvenously, the perimedullary vein was catheterized. Starting at this level, the venous system and fistula were closed with a fluid embolization agent. Control angiography did not reveal any finding of dAVF (Figure 2). No complications were observed after angiography. According to the clinical condition of the patient, a rehabilitation program was planned, which included passive and active-assistive range of movement (ROM) exercises, neuromuscular electrical stimulation, progressive resistance exercises, balance and coordination exercises, lower limb robotic rehabilitation, and progressive ambulation training.

Before embolization, the patient was paraplegic; however, he showed a significant improvement in the first month of rehabilitation. At discharge, the patient's lower extremity muscle strength returned to normal. He had minimal hypoesthesia in the L5/S1 dermatomes and did not have any gait disability. FAS was 5 (independent ambulation) and FIM was 123. The patient was scheduled for follow up after 2 months.

Discussion

Spinal dAVF is a cause of vascular-related spinal cord injury (5). The incidence is estimated to be approximately 5–10 cases per million. The lesion is more common in men over the age of 50 years and affects the thoracic and lumbar spine (1). These patients usually present with slow and progressive myelopathy. Trauma, infection, surgery, and syringomyelia are all considered as alternative diagnoses because the exact etiology is often unclear. The pathological lesion is a shunt between the radicular artery and vein, which causes venous hypertension in the spinal cord (1). Vascular steal and spinal cord compression have also been suggested as the mechanism for damage. However, the current theory is that shunting of arterialized blood causes increased venous pressure in the coronal venous plexus and leads to congestion, edema, and eventually ischemic injury in the affected region of the cord (6).

Although it is a treatable cause of myelopathy, spinal dAVF is rarely considered and is usually diagnosed late. This is because it is so infrequently encountered in daily practice and the clinical presentation is non-specific (1). Gait disability is usually the first symptom. Progression to full-blown myelopathy or paraplegia is slow. Intermittent paresthesia and symptoms of sensory loss, suggestive of peripheral nerve lesions, may be observed (4). Bladder dysfunction, intestinal dysfunction, and impotence may also be observed (6). Our patient had a history of difficulty in walking and motor symptoms; however, he did not have any bladder or intestinal dysfunction until he was completely paraplegic.

Physical examination may reveal upper motor neuron lesion findings, such as increased muscle tone and increased deep tendon reflexes, as well as early lower motor neuron lesion findings, and this can complicate the diagnosis. The average time between the onset of the symptoms and the diagnosis ranges from 12 to 44 months (4). Our patient was symptomatic for 3 months, and, although he described paraparetic episodes, the physical findings were ambiguous. Unnecessary interventions, surgeries, and misdiagnosed cases have been reported in the literature (4,7).

In the differential diagnosis, the causes of non-traumatic progressive myelopathy, which are extensive, should be considered (8). Patients with a more rapid progression should primarily be investigated for compressive lesions, such as metastatic neoplasms and spinal dural abscesses, which require immediate intervention. Also, cervical spondylosis, which is the most common cause of quadriparesis, and lumbar disc steno-

sis should be considered (9). Aortic dissection, postoperative ischemia, vascular embolism, or systemic hypotension can cause similar clinical manifestations by creating spinal cord ischemia (5). Inflammatory myelopathies can have acute, subacute, or chronic onset. They can be isolated or be a component of multiple sclerosis, and, rarely, may be associated with chronic infections [e.g., acquired immunodeficiency syndrome (AIDS), syphilis, etc.], and rheumatic or connective tissue diseases, such as systemic lupus erythematosus (SLE) and anti-phospholipid syndrome (10). The other causes of myelopathy, such as anterior horn motor neuron disease, paraneoplastic syndromes, radiation or electrical injury, and nutritional etiologies, especially in patients with gastrointestinal disease and gastric bypass operations, should also be considered (8).

Family history, duration of symptoms, patient age, comorbid diseases, systemic symptoms, and thorough questioning for peripheral nervous system symptoms are helpful in the diagnosis. Mass lesions, discopathy, and inflammatory myopathies can be excluded through imaging modalities and cerebrospinal fluid analysis (8). Early symptoms, such as paresthesia and lower motor neuron findings, can also suggest polyradiculopathies. Upper extremity symptoms are rarely seen in spinal dAVF, and the absence of glove-like sensory loss, asymmetry, and bladder dysfunction may help to exclude polyneuropathies (4).

A spinal dAVF is seen as a hyperintense lesion on T2-weighted MRI images, and often a corresponding hypointense signal can be found on T1-weighted images. These findings are secondary to the cord ischemia and edema. Additional MRI findings include prominent intradural veins, spinal cord enhancement and enlargement, and scalloping and irregular cord surface. However, these findings are non-specific and must be confirmed by the gold standard imaging method of angiography (11). In our patient, on spinal MRI, there were no degenerative findings, discopathy, or stenosis to explain the clinical symptoms. Due to the edema and the expansion of the vascular structures at the thoracic and lumbar regions of the cord, an angiography was performed to investigate vascular lesions.

Endovascular embolization and microsurgical techniques are available as treatments for spinal dAVF. Our patient was treated with endovascular embolization. Hessler et al. stated that there is no consensus as yet for the optimal treatment of dAVF, and surgery can be performed on patients with failed embolization (2). The level of healing is thought to be related to the degree of spinal lesion caused by spinal venous congestion (7). Prognosis and success of treatment have been reported to be better in younger patients with fewer symptoms. Pre-operative severe neurological clinical signs are one of the major factors that worsen the prognosis (12,13).

Early diagnosis is strongly correlated with a better prognosis. Better treatment outcomes have been shown in early-diagnosed patients who were treated with either embolization or surgery (13). Micturition, pain, and muscle spasms often have a worse response to treatment compared to gait disability (4,13). Our patient was symptomatic for 3 months when he underwent embolization. Due to the early intervention, he showed rapid improvement after the treatment and did not have gait or any serious disability on discharge. In most of the case series in the literature, data on the time between the onset of rehabilitation and fistula treatment are limited (14). However, in most cases, it has been reported that if the intervention is delayed, even prolonged rehabilitation does not change the grave prognosis (15).

In conclusion, spinal dAVF is an important and treatable condition with slow clinical progression and non-specific symptoms. Because a patient's prognosis is strongly correlated with the time of diagnosis, spinal dAVF should be considered in the differential diagnosis of patients who present with paresis and plegia in daily practice. Embolization and surgery are available treatment options and should be supported with appropriate rehabilitation programs.

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