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CASE REPORT

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A Rare Presentation In The Differential Diagnosis of Paraparesis: Spinal Dural Arteriovenous Fistula - A Case Report

Paraparezi Ayırıcı Tanısında Nadir Görülen bir Tablo: Spinal Dural Arteriovenöz Fistül - Olgu Sunumu

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ABSTRACT

Spinal dural arteriovenous fistulas (SDAVFs) are acquired vascular malformations characterized by spinal cord dysfunction. Patients present with symptoms such as paraparesis, sensory deficits and dysfunction of the bowel and bladder. The gold standard in diagnosis is spinal angiography. However, delays in diagnosis and treatment are common. In this article, we aim to present a case of SDAVF presenting with paraparesis and urinary incontinence. A 40-year-old male patient presented to our clinic with weakness in the legs and difficulty urinating following an upper respiratory tract infection. Neurological examination revealed paraparesis consistent with a 1st motor neuron lesion and sensory deficit was present starting from level T9. Spinal cord MRI showed hyperintense signal changes on T2-weighted sequences enhancing from the T5 to L1 vertebral levels. Spinal MR angiography performed with the diagnosis of SDAVF revealed a vascular lesion resembling SDAVF extending between T4 and L2 vertebrae and the patient underwent endovascular embolization therapy. SDAVFs most commonly occur in the thoracolumbar region and predominantly affect middle-aged male patients. The absence of a typical clinical presentation complicates early diagnosis at initial presentation. Delayed diagnosis can lead to further neurological deficits and more severe outcomes. Therefore, in cases presenting with paraparesis and extensive hyperintense lesions on MRI, SDAVF should always be considered in the differential diagnosis.

Keywords: Spinal dural arteriovenous fistula, paraparesis, transverse myelitis.

ÖZ

Spinal dural arteriovenöz fistül (SDAVF)'ler spinal kord disfonksiyonu ile karakterize edinsel bir vasküler malformasyondur. Hastalar paraparezi, duyu kusurları, bağırsak ve mesane disfonksiyonu bulguları ile başvururlar. Tanıda altın standart spinal anjiografidir. Bununla birlikte tanı ve tedavide gecikmeler sık görülmektedir. Bu yazıda paraparezi ve idrar inkontinansı ile başvuran bir SDAVF olgusu sunmayı amaçladık. 40 yaşında erkek hasta üst solunum yolu enfeksiyonu sonrası başlayan bacaklarda güçsüzlük, idrar yapma güçlüğü şikayeti ile kliniğimize başvurdu. Nörolojik muayenede 1. motor nöron tipinde paraparezi ve T9 seviyesinden itibaren seviye veren his kusuru mevcuttu. Spinal kord MR incelemesinde T5 ile L1 vertebra seviyesi boyunca kontrastlanma gösteren T2A sekanslarda hiperintens patolojik sinyal artışı izlendi. SDAVF şüphesi ile yapılan spinal MR anjiyografide T4 ile L2 vertebralar arasında uzanan SDAVF ile uyumlu vasküler yapı görüldü ve endovasküler embolizasyon tedavisi uygulandı. SDAVF'ler en sık torakolomber bölgede yerleşirler ve ağırlıklı olarak orta yaşlı erkek hastaları etkilemektedir. Tipik bir klinik prezetasyonun olmaması, ilk başvuruda tanı koymayı güçleştirir. Tanıda gecikme daha fazla nörolojik defisite neden olabilir ve daha fazla sekel ile sonuçlanabilir. Bu nedenle paraparezi kliniği ile başvuran ve MRG'da uzun segment hiperintens lezyon görülen olgularda ayırıcı tanılar arasında mutlaka SDAVF'ler yer almalıdır.

Anahtar Kelimeler: Spinal dural arteriovenöz fistül, paraparezi, transvers miyelit.

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CASE PRESENTATION

Spinal dural arteriovenous fistula (SDAVF) is an acquired vascular malformation that is the cause of spinal cord dysfunction. Although it is the most frequent cause of vascular malformations located in the medulla spinalis, it constitutes only 5% of all neurovascular pathologies .[1,2] Due to its rarity and early non-specific clinical findings, patients may be misdiagnosed with intramedullary tumors, inflammatory lesions, polyneuropathy, and degenerative disc disease.[3-5]

The main problem in these patients is that the disease is not part of the clinical and radiological differential diagnosis, and therefore diagnosis and treatment are delayed.[6] Progressive spinal symptoms have been observed in patients with delayed treatment, leading to high morbidity.[7] Spinal angiography is the gold standard for the diagnosis of SDAVF.[8] Treatment options include endovascular embolization and surgery.[6]

In this article, we review a case of SDAVF who presented to our clinic with progressive paraparesis and urinary incontinence. The primary diagnosis was transverse myelitis.

CASE REPORT

A 40 years old male patient was admitted to our clinic with bilateral lower extremity hypoesthesia, paraparesis, and dysuria that began 10 days ago following an upper respiratory tract infection. It was found from his history that he had been under urological follow-up for approximately 6 years due to dysuria, and had been under medical follow-up with a prediagnosis of urethral stricture.

Neurological examination revealed 3/5 paraparesis in right lower extremity and 4/5 paraparesis in left lower extremity, sensory deficit in the form of hypoesthesia leveling from T9, decreased vibration sensation in bilateral lower extremities, lower extremity deep tendon reflex vividness, bilateral extensor sole reflex, and clonus on the right.

There were no abnormalities in routine blood tests or infection parameters. MR examination of the spinal cord demonstrated hypointense pathologic signal enhancement on T1 sequences and hyperintense pathologic signal enhancement on T2 sequences beginning at the level of the T5 vertebra and extending continuously to the level of the L1 vertebra. The contrast-enhanced series showed heterogeneous areas of enhancement. (Figure 1)

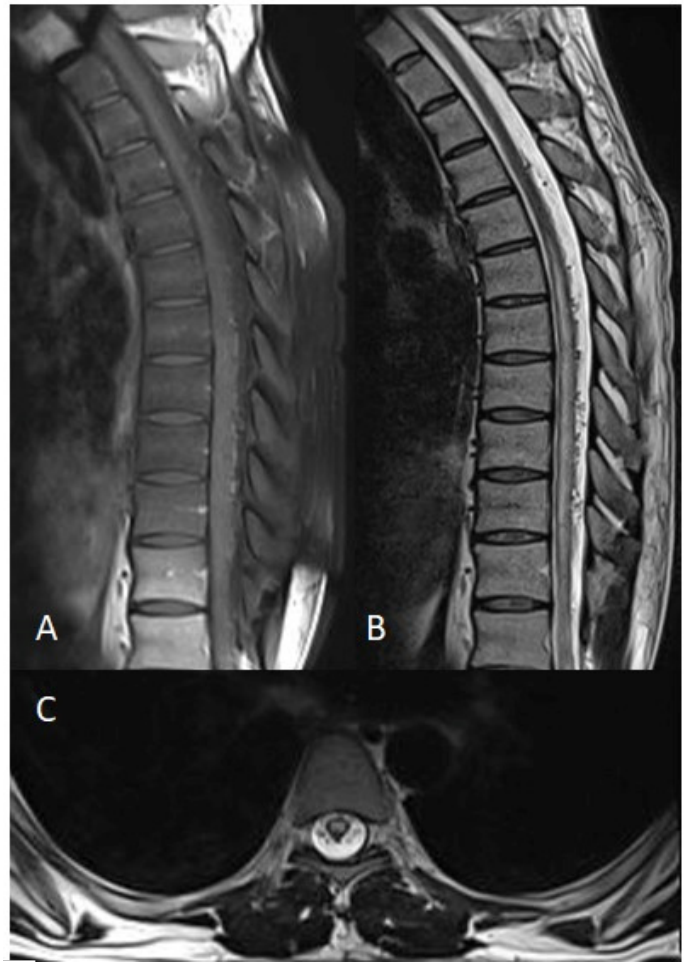

Figure 1. In spinal MRI examination, (A) heterogeneous enhancement in sagittal contrast-anhanced series, sagittal (B) and axial (C) hyperintense pathologic signal enhancement on T2 sequences of the spinal cord at the T5-L1 level and expansion in the spinal cord.

Lumbar puncture performed due to prediagnosis of transverse myelitis revealed an opening pressure of 130 mmH_2O, CSF protein 95 mg/dl, CSF glucose 62 mg/dl (concurrent blood glucose 107 mg/dl), and no cells were observed in the CSF. Anti-NMO(AQP4-IgG) antibody was negative. Intravenous pulse steroid treatment was planned. It was observed that the patient's cough was worsening at night due to an upper respiratory tract infection. The next day, as the neurological examination showed a deterioration, our diagnosis was reviewed in consultation with the neuroradiology department. Spinal MR angiography scheduled for suspected SDAVF.

Spinal MR angiography demonstrated a vascular structure consistent with SDAVF extending between the T4 and L2 vertebrae. The patient, who was referred to Interventional Radiology, was scheduled for endovascular embolisation treatment.

Spinal angiography revealed an Adamkievicz's artery originating from the right T8 root and an opacified dural AVF originating from the right T6 pedicle. Spinal angiogram, including both the upper and lower segments of the lesion after embolization, showed no angiographic evidence of residual AVF. (Figure 2)

MAIN POINTS

- Spinal dural AVF is one of the diagnoses that both clinicians and neuroradiologists should consider in patients with progressive paraparesis who have long-segment hyperintense lesions on spinal MR imaging.
- Spinal angiography is the gold standard for diagnosis.
- Although treatment options vary from patient to patient, the most effective methods are microsurgical occlusion and endovascular embolization.

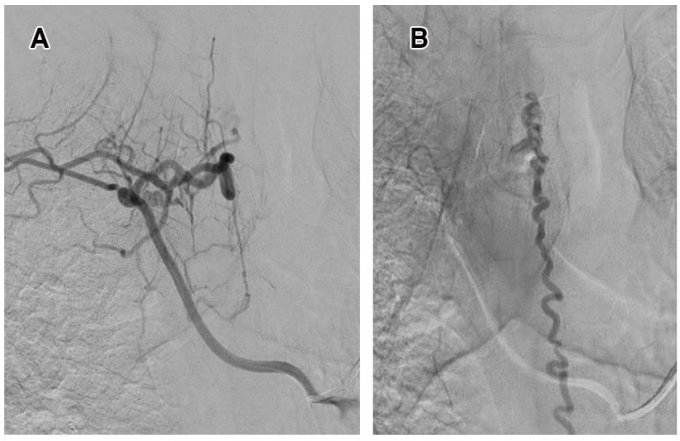

Figure 2. A spinal dural arteriovenous fistula on angiography. (A) Adamkievicz's artery originating from the right T8 root and an opacified dural AVF originating from the right T6 pedicle, (B) after embolization, showed no angiographic evidence of residual AVF.

Neurological examination performed 3 days after surgery showed improvement in the form of 4/5 paraparesis in the lower extremity. At the follow-up examination after 6 months, a 50% regression of the previous findings of hypoesthesia in the bilateral lower extremities was noted. However, intermittent urinary incontinence was persistent. Our patient continues to follow up as an outpatient. For this case report, signed informed consent was obtained from the patient.

DISCUSSION

Spinal dural arteriovenous fistulas are quite rare, accounting for 70% of all spinal vascular malformations.[9] Although their etiology is still unclear, they are thought to be acquired lesions.[10] Anatomically, it is led by an abnormal connection between the radiculomeningeal arteries and the radicular veins in the spinal column. Direct arterial flow to the venous system increases pressure within the dorsal coronal venous plexus. The increased pressure causes the venous plexus to stasis and become more tortuous. The result is intramedullary oedema in the spinal cord[11], known as congestive or venous hypertensive myelopathy.[11]

SDAVFs are most frequently located in the thoracolumbar region and are most common in middle-aged male patients (mean age 55-60 years) [6]. Although our case is similar to the general picture in terms of localisation and gender, it occurs at a younger age.

The absence of a typical clinical presentation makes it difficult to diagnose at first admission.[12] The majority of patients admit with difficulty walking, paraparesis, paresthesia and symmetric or asymmetric sensory deficits.[13] Involvement of the bowel and bladder and erectile dysfunction are most likely to occur in the late stages of the disease.[10,14] In the present case, the urinary symptoms has persisted for 6 years. The patient has followed up with a diagnosis of urethral stricture.

Spinal dural AVF is one of the diagnoses that both clinicians and neuroradiologists should consider in patients with progressive paraparesis who have long-segment hyperintense lesions on spinal MR imaging, started intravenous pulse steroid treatment, and failed to respond to treatment. Andrew et al. reported that 16 out of 46 patients were initially misdiagnosed and that there was a significant delay in diagnosis from the onset of symptoms.[15]

Although the mechanism is not fully understood, severe neurological deficits can develop following steroid treatment and lumbar puncture if misdiagnosed.[16,17] There have been reports in the literature of acute clinical deterioration following lumbar puncture and intravenous pulse steroid treatment. Silvia et al. reported that a patient with SDAVF was diagnosed with transverse myelitis and deteriorated rapidly after the use of intravenous pulse steroids.[18]

In cases reported in the literature, patients' symptoms were noted to increase with physical activity and increased intra-abdominal pressure.[4] This is explained by the fact that the intraoperative vascular pressure value is 74% of the systemic arterial pressure.[5] In our case, the increase in complaints after a severe cough is of a similar nature.

Although spinal angiography is the gold standard for diagnosis, MRI should be the first imaging modality of preference in undiagnosed cases. Expansion of the medulla spinalis due to venous stasis on spinal MRI, and flow cavities in the subarachnoid space corresponding to areas of gadolinium uptake on T2-weighted MRI are specific findings for the diagnosis of SDAVF.[19]

The goal of SDAVF treatment is to prevent the progression of the condition. Although treatment options vary from patient to patient, the most effective methods are microsurgical occlusion and endovascular embolization. The goal is to prevent abnormal blood flow by suspending the fistula itself or the drainage vein proximally. This is why endovascular embolization is the first preferred method. It is less invasive, allows rehabilitation to be initiated earlier and does not completely exclude the option of surgery.[20]

There are studies in the literature on the comparison of success rates between endovascular and surgical treatment. Hessler et al. reported that endovascular embolectomy was less successful than surgical treatment in 156 patients with SDAVF over the past 30 years.[21] Similarly, Fillis et al. compared the success rates of treatment modalities in SDAVF patients. Failure to treat was observed in 5 of 11 patients (45.5%) who underwent endovascular treatment, compared with 3 of 70 patients (4.3%) who were treated using surgical techniques.[22] On the other hand, as a result of recent developments in endovascular treatment, the success rate of embolisation has reached approximately 77%.[23] In our case, endovascular embolisation was considered appropriate and achieved with effective treatment.

The prognosis after treatment varies depending on how long the symptoms lasted and the clinical situation before treatment. After the closure of the fistula, about two thirds of the patients have an improvement of the motor symptoms and about one third have an improvement of the sensory symptoms. Although recovery continues for up to a year after treatment, sphincter-related problems and erectile dysfunction tend to be more permanent.[24,25] A retrospective review of patients treated with a diagnosis of SDAVF reported that 80% of patients had improvement in at least one symptom, and motor symptoms had the best prognosis.[26]

CONCLUSION

In conclusion, because SDAVF does not have a typical clinical presentation and is not the first diagnosis that comes to mind on imaging findings, it may be diagnosed at a later stage. A delay in diagnosis can lead to a more severe neurological deficit and result in a greater number of sequelae.[27] Therefore, SDAVFs should be considered in the differential diagnosis of patients with paraparesis and long-segment hyperintense lesions on MRI.

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