



Case Report

Spinal dural arteriovenous fistula: A rare cause of progressive myelopathy and bladder and bowel dysfunction

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Received: October 01, 2018 **Accepted:** February 11, 2019 **Published online:** May 18, 2020

ABSTRACT

Spinal dural arteriovenous fistula (SDAVF) is a rarely seen vascular lesion in the spinal cord and is often overlooked. If left untreated in the early stages, it is associated with severe morbidity and may lead to progressive myelopathy and bladder and bowel dysfunction. A 55-year-old male patient was admitted with complaints of lower extremity weakness, gait disorder, urinary retention, and stool retention. Based on physical examination and magnetic resonance imaging findings, a preliminary diagnosis of SDAVF was made. The diagnosis was confirmed by spinal angiography showing SDAVF on the left T6. Microsurgery was planned, once endovascular embolization failed. Although symptoms of progressive myelopathy and bladder and bowel dysfunction are rarely seen, SDAVF diagnosis should not be overlooked, and it should be kept in mind that early diagnosis and treatment prevent severe morbidities.

Keywords: Angiography, fistula, magnetic resonance imaging, myelopathy, rehabilitation, spinal cord.

Spinal dural arteriovenous fistula (SDAVF) is a vascular lesion caused by the interactions between the arterial and venous vascular systems of the spinal cord.^[1,2] It may induce abnormal flow of the blood from the arterial system to the venous system, venous hypertension, venous occlusion, intramedullary edema, and progressive myelopathy.^[3-5] The incidence of SDAVF is 5 to 10 in a million, and the majority of patients are male patients between their 60s and 70s.^[4,5]

Since SDAVF is a rare entity with an unknown etiology, its diagnosis can be easily overlooked.^[1,2] To prevent permanent neurological disorders such as gait disorder, urinary retention, or stool retention, early diagnosis and treatment of SDAVF are critical.^[2] Although the diagnosis of SDAVF is possible with the help of the suspicion of the clinician and attention of the radiologist during magnetic resonance imaging (MRI), the gold standard for the diagnosis is selective spinal angiography.^[2-5] Angiography is used for localization of the vascular lesion and treatment. Microsurgery and endovascular embolization are effective methods for the treatment of SDAVF.^[1,2]

In this article, we report a case of SDAVF presenting with progressive myelopathy and bladder and bowel dysfunction in the light of literature data.

CASE REPORT

A 55-year-old male patient was admitted with complaints of lower extremity weakness, gait and balance disorder, urinary retention, and stool retention for six months with an increasing severity. The patient was previously applied to a variety of hospitals and specialties for his symptoms; however, the definitive diagnosis was unable to be made. On physical examination, the muscle strength was 5/5 in the upper extremities, 3/5 in the left lower extremity and 4/5 in the right lower extremity based on the Global Oxford Scale. There was no spasticity. The deep tendon reflexes of the bilateral patella and Achilles tendons were hyperactive. The Babinski reflex was not evoked in either feet. The Hoffman reflex was negative. Based on the sensory examination, left hypoesthesia was observed starting from the T8 level. The functional

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Cite this article as:

Sucuoğlu H, Aktürk A. Spinal dural arteriovenous fistula: A rare cause of progressive myelopathy and bladder and bowel dysfunction. Turk J Phys Med Rehab 2020;66(2):219-222.

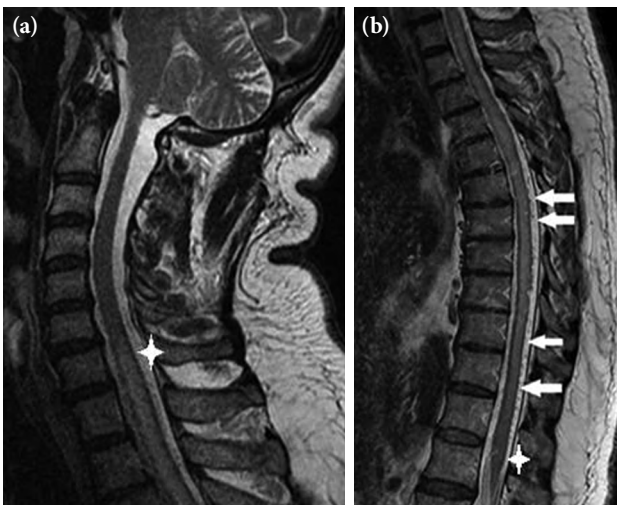


Figure 1. (a) T2-weighted cervical and (b) thoracic spine sagittal magnetic resonance imaging showing intramedullary increased signal from level of C6-7 disc (stars) to conus medullaris with associated expansion as well as signal void serpiginous structures (arrows) around spinal cord representing dilated vessels.

status of the patient was assessed using the modified Aminoff and Logue Scale of Disability (ALS), which grades gait, urinary incontinence, and fecal continence/constipation.^[6] The total score of the ALS was 7/11, indicating moderate disability. The modified ALS sub-scores were as follows: gait-requires one cane or some support for walking “3”; urination-occasional incontinence or retention “2”; defecation-occasional incontinence or persistent constipation “2”. The results of the examinations of the patient which were carried out at external centers and biochemistry, complete blood count, sedimentation, and CRP values were all normal. The lower extremity needle and conduction electromyography and cranial MRI findings were also normal. Based on these findings, a whole spinal column MRI was performed, and the MRI of the structures lying in the central part from the C6-7 disc level to the conus medullaris and filling almost all parts of the medulla spinalis showed hypointensity on T1A, accelerated hyperintense myelopathic signal increase on T2A, and numerous serpiginous dorsal epidural flow voids in the cerebrospinal fluid adjacent to the spinal cord (Figure 1). This situation was evaluated as a SDAVF and, based on the angiography performed for accurate diagnosis and treatment purposes, SDAVF between the radicular artery and the perimedullary veins was found on the T6 level on the left (Figure 2a).

A written informed consent was obtained from the patient. Endovascular embolization was planned by the interventional radiology department; however, it failed.

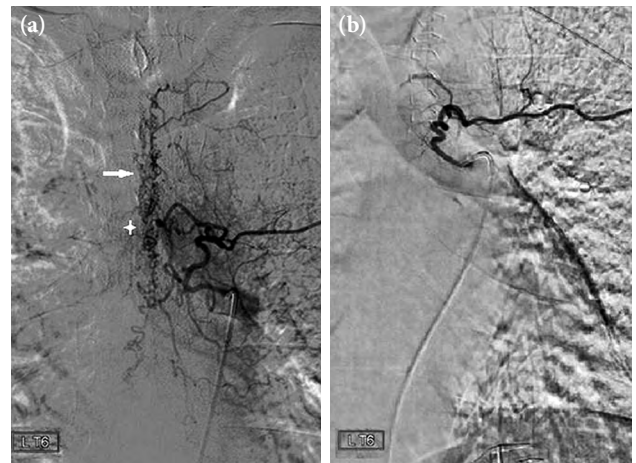


Figure 2. A spinal dural arteriovenous fistula on angiography (a) Before operation: During contrast injection at the left T6 level, abnormal filling of dilated epidural veins (arrow) and spinal dural arteriovenous fistula-dependent glomerular network-like image (star) between radial artery and perimedullary vein are seen. (b) After operation: Normal radial artery filling without abnormal venous enlargement following closure of fistula after surgical coagulation of perimedullary vein at the left T6 level.

Therefore, the patient was referred for microsurgery. During the surgical treatment, the fistula was closed and control angiography showed normal radial artery filling (Figure 2b). An outpatient rehabilitation program including passive and active assisted range of movement exercises, muscle strengthening exercises, neuromuscular electrical stimulation, and balance and coordination exercises was initiated from the first week after the operation. Additionally, the patient was consulted with the relevant clinics for bladder and bowel dysfunction.

DISCUSSION

Spinal dural arteriovenous fistula is a rarely seen spinal vascular malformation leading to severe morbidity, if left undiagnosed and untreated in the early stages.^[2,7] Blood reaches the spinal cord via spinal arteries (one at the anterior and two at the posterior) lying through the vertebral column, nerve roots, and radicular arteries before reaching the spinal cord.^[2,8] Anterior and posterior spinal veins exhibit a similar distribution to that of spinal arteries in the venous system and reach radicular veins, followed by the coronal venous plexus and the vena cava system.^[2,8] Moreover, SDAVF occurs as a result of a direct connection between the radicular artery and the coronal venous plexus. Branches of the radicular artery merge with the venous system like a glomerular net at

the fistula located in the dural envelope of the nerve root.^[3] As a result of the arterialization of the venous system by this way, venous hypertension, venous occlusion, ischemia, and edema develop, leading to progressive myelopathy.^[3,4,7] In our case presenting with edema and venous congestion, there was a distinctive myelopathy reaching from the C6 level to the conus medullaris and spinal cord extension.

Although SDAVF has no clear structural and hemodynamic classification, a series of classifications has been made to reflect its complex vascular anatomy.^[9] Currently, based on the most commonly used classification, spinal vascular malformations are divided into four main categories: Type I, dural arteriovenous fistula (DAVF); Type II, intramedullary glomus-type arteriovenous malformation (AVM); Type III, child or combined AVM, and Type IV, intradural perimedullary DAVF.^[9,10] About 60 to 80% of SDAVFs are located in the thoracic region, and most of these are unilaterally detected at the level of T7-12.^[11,12] This lesion is less frequently seen in the lumbar region and rarely seen at the cervical level.^[7,12] Bilateral or multiple SDAVF phenomena are extremely rare.^[11,12] In our case, it was a unilateral, type I SDAVF at the left T6 level.

The majority of SDAVF patients are males over the age of 50.^[7,11] The initial manifestations of this pathology are non-specific clinical symptoms, and median time to diagnosis ranges from 12 to 44 months.^[13] When myelopathy progresses, patients may present with complaints such as weakness in their lower extremity, gait disorder, sensory loss, and intestinal and urinary bladder dysfunction (incontinence or retention).^[2,3,7,12] The differential diagnosis of SDAVF includes polyneuropathies, demyelinating diseases, spinal neoplasms, and other spinal pathologies.^[14] Although gait disorder and bladder and bowel dysfunction symptoms were distinctive in our 55-year-old patient, the definitive diagnosis was unable to be made at external centers previously, and time from the onset of complaints to the diagnosis was six months. The reason for this delay may be the rareness of SDAVF and, the disease is frequently overlooked for this reason.

Although SDAVF can be detected by spinal MRI, which is the first step of examination for investigating progressive myelopathy, the gold standard for diagnosis is spinal angiography.^[4,5] The distinctive characteristics of SDAVF on MRI are as follows: based on progressive myelopathy, multiple observations of hyperintensity on the T2 sections lying through the

vertebra segment to the conus medullaris, hypo-intense signals on the T1 sections, spinal cord extension based on edema, multiple intradural flow voids based on dilate intradural venous extensions and irregular cord surface images.^[2,3,10] Similar MRI findings were seen in our case. In addition, as in our case, the localization and level of the fistula and detailed characterization of the veins joining the fistula may be detected by spinal angiography.^[11]

Although treatment is individually planned for each SDAVF patient, two effective treatment methods are microsurgery occlusion and endovascular embolization.^[1,2,10] The main goal of the treatment is to prevent abnormal blood flow by suspending the fistula itself or drainage vein from the proximal. Therefore, endovascular embolization is the first preferred method, as it is less invasive, allows starting rehabilitation earlier, and does not completely eliminate the option of having surgery.^[2] Although the success rates of endovascular and surgical treatments are similar, Hessler et al.^[11] reported that endovascular embolization was less successful compared to surgical treatment in 156 SDAVF patients for the previous 30 years. However, as a result of the recent developments in endovascular techniques, the success rate of embolization has reached about 77%.^[12,15]

The prognostic factors of SDAVF vary in the literature. Young age, mild clinical course, and early diagnosis and treatment can be deemed as favorable prognostic factors.^[2,16-18] Contradictory results have been reported for the relationships between sex, fistula level, and short symptom time and prognosis.^[12,18]

After the interventional treatment of SDAVF, early rehabilitation may positively contribute to the prognosis.^[16] According to the clinical condition of the patient, passive and active assisted range of movement exercises, muscle strengthening exercises (isometric or isotonic), progressive resistance exercises, neuromuscular electrical stimulation, balance and coordination exercises, lower limb robotic rehabilitation, and progressive ambulation training can be added to the rehabilitation program.^[16]

In a retrospective study, Ofran et al.^[18] analyzed the rehabilitation results of eight SDAVF patients and reported that five patients were dependent on wheelchairs, despite a mean duration of two months of rehabilitation with a three-hour multidisciplinary approach including physiotherapy, occupational therapy, rehabilitation nursing, social workers, and psychological support. The importance of early diagnosis and early intervention rehabilitation

indicated that success was related to functional ambulation potential. Additionally, most SDAVF patients reported that the rehabilitation process could be maintained in the outpatient setting; however, in patients with severe motor and functional disability, inpatient rehabilitation treatment could be required. In our case, outpatient rehabilitation started immediately after the intervention.

Clinical recovery continues during one-year follow-up after SDAVF treatment.^[12,19] Approximately 97% of patients show recovery (74 to 82%) or stabilization (14 to 22%) in terms of their motor findings.^[12,19] The recovery rate for sphincter dysfunctions is, however, lower and reported to be between 5 and 58%.^[12,19,20]

In conclusion, SDAVF is an important spinal vascular pathology which should not be overlooked in the differential diagnosis of patients with progressive myelopathy, and bladder and bowel dysfunction. The etiopathogenesis of this disease has remained uncertain so far. Therefore, for a definitive diagnosis, it is required to recognize about MRI findings and conduct spinal angiography. Even though there are no precise guidelines for the treatment, endovascular embolization and surgery have been proven to be effective. Since it is a rare disease and easily overlooked, the suspicion of clinicians and attention of radiologists are of utmost importance to establish a definitive diagnosis. Early diagnosis and treatment are the most important prognostic factors for these patients in reducing morbidity.

Acknowledgements

We thank the, Private Nisa Hospital Physical Medicine and Rehabilitation Clinic, Radiology Clinic, for their contributions. We would also thank MD. Abdulkadir Asan, radiologist, who provided valuable contributions to the evaluation of the patient and to the collection of data in this study.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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