

Unveiling The Hidden: Disc Herniation Concealing A Spinal Dural Arteriovenous Fistula-A Case Report

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Abstract:

Spinal dural arteriovenous fistula (SDAVF) represents a relatively rare yet clinically significant vascular anomaly affecting the spinal cord. Despite its infrequency, SDAVF poses a considerable threat to neurological function if left untreated. This review provides an overview of SDAVF, including its epidemiology, pathophysiology, clinical manifestations, diagnostic modalities, and treatment options. SDAVF typically presents with insidious onset and progressive neurological deficits, often mimicking other spinal pathologies, thereby posing diagnostic challenges. Advanced imaging techniques such as magnetic resonance imaging (MRI) and digital subtraction angiography (DSA) play pivotal roles in accurate diagnosis and localization of SDAVF. Treatment strategies encompass a spectrum of interventions ranging from endovascular embolization to surgical resection, with the goal of halting disease progression and preserving neurological function. Despite advancements in diagnostic and therapeutic modalities, SDAVF remains a diagnostic dilemma and therapeutic challenge due to its variable presentation and complex vascular anatomy. This review highlights the importance of early recognition, multidisciplinary management, and ongoing research efforts aimed at optimizing outcomes for patients with SDAVF.

Keywords: Paraplegia, fistula, perimedullary, angioMRI, angiography

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I. Introduction:

Spinal arteriovenous malformations (SAVMs) encompass a spectrum of rare but potentially debilitating vascular anomalies affecting the spinal cord. Within this spectrum, spinal dural arteriovenous fistula (SDAVF) stands as the most prevalent entity, though still relatively uncommon in the broader context of neurological disorders. Despite its prevalence, SDAVF remains frequently underrecognized and diagnosed belatedly, leading to significant neurological morbidity if left untreated. This review aims to delve into the intricate landscape of SDAVF, shedding light on its diverse clinical presentations, the intricacies of diagnostic approaches, and the evolving landscape of therapeutic interventions. By navigating through these complexities, we aim to provide a comprehensive understanding of SDAVF, exploring both the diagnostic challenges and therapeutic advancements that characterize the management of this complex vascular pathology. Furthermore, we endeavor to elucidate recent strides in unraveling the pathophysiological underpinnings of SDAVF and the innovative imaging modalities that have revolutionized its detection and treatment strategies. Through this exploration, we seek to underscore the imperative for heightened clinical vigilance, early diagnosis, and tailored management approaches in tackling this formidable neurological condition.

II. Case Report :

A 38-year-old man repeatedly sought medical attention for back pain, managed with analgesics. He had no significant medical history and worked as a mason but was placed on leave due to inability to resume work by his orthopedist, who diagnosed him with lumbar disc herniation through a CT scan. As weakness in both lower limbs and urinary urgency emerged, a spinal MRI was performed.

On physical examination, the patient exhibited urinary retention and paraplegia. According to the American Spinal Cord Injury Association (ASIA) assessment, the patient was classified as grade B with a sensory level of T10. An MRI of the thoracic spine with and without contrast was ordered, revealing suspicion of a vertebral AVM at T9-T12, as shown on T2-weighted imaging (Figure 1). Differential diagnoses included trauma, tumor, and other vascular anomalies.



Figure 01 : Preoperative MRI with and without contrast agent,T2-weighted

The patient underwent preoperative spinal angiography via digital subtraction to characterize the lesion. The nidus was found centered around the right pedicle of T11, measuring approximately 1 cm. Intraoperatively, the lesion extended from T8 to T12 (Figure 2).

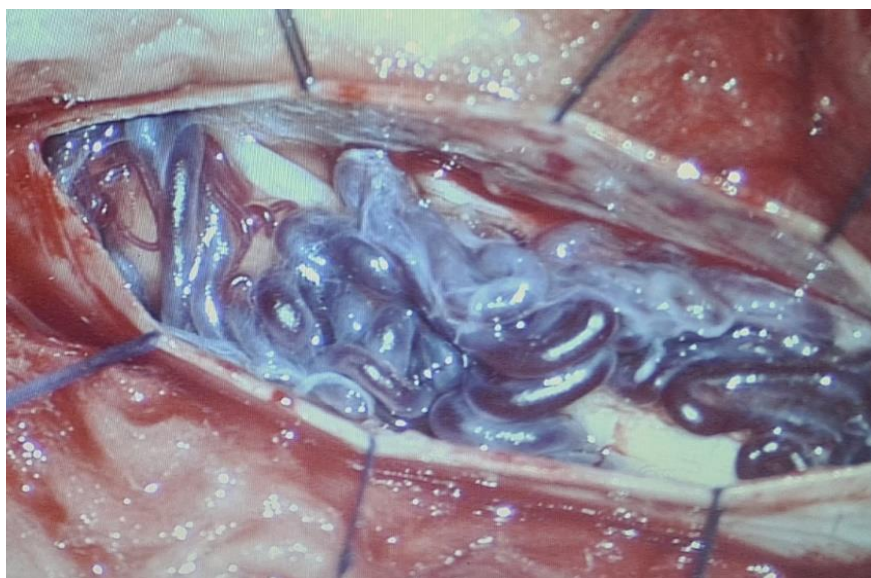


Figure02 :Intraoperative image showing the lesion

The patient was taken to the operating room for surgical intervention. Right laminoplasty and durotomy were performed at T7-T11, revealing a feeding artery piercing the dura mater near the lower border of the right T10 pedicle surrounded by numerous arterialized veins. Several straight clips were placed. Postoperatively, the patient was brought to the angiography suite to confirm AVM obliteration and preservation of the Adamkiewicz artery. A postoperative MRI was also obtained to confirm AVM obliteration.

Postoperatively, the patient remained ASIA grade B with a sensory level of T10. He was referred for functional rehabilitation and followed up in clinic with stable neurological status and no signs or symptoms of AVM recurrence.

III. Discussion :

Spinal dural arteriovenous fistula (SDAVF) is a relatively rare entity but represents the most common spinal vascular malformation. It typically occurs in males after 50 years of age. Clinical signs include motor, sensory, and genitourinary symptoms. Symptom onset is usually progressive, rarely abrupt. Diagnosis is often delayed, leading to significant morbidity in the absence of treatment. Diagnosis relies on spinal MRI and angiography.(05)

We present the case of a 34-year-old man with progressive bilateral lower limb weakness secondary to a vertebral AVM. Based on imaging characteristics, intraoperative findings, and patient demographics, it was likely a Type I AVM. Although rare, our patient's presentation did not fit the typical age range for this type of AVM. It's also noteworthy that while these AVMs are typically embolized to alleviate symptoms, in this case, surgical resection was deemed the most appropriate procedure. The patient tolerated the intervention well, and his back pain improved. Although his ASIA level did not improve, he reported slight contracture of the left big toe.(03)

Early suspicion, evaluation, diagnosis, and management could have led to improved neurological outcomes in this case. Similar to other compressive spinal cord pathologies like tumors, progressive deficits can become permanent. Any further delay in treating this patient could have resulted in worsening deficits and potentially death due to AVM expansion and possible widespread rupture. Furthermore, while the risk of recurrence in adult AVM patients is low, long-term follow-up to monitor for recurrence is important. This is especially true in patients under 18, as previous reports/case series cite recurrence rates between 5.5% and 17.5%, with recurrence occurring one to five years after complete resection.(07)

A key factor to note regarding this lesion is its proximity to the Adamkiewicz artery, which typically runs from the left side of the aorta from T8 to L2. Injury to this artery can cause severe spinal cord ischemia similar to anterior spinal artery syndrome, resulting in neurological deficits including urinary/fecal incontinence, and weakness and paralysis of the lower limbs. Although our patient already had complete motor loss in his bilateral lower limbs, he had the potential to recover bowel and bladder functions; thus, preserving this artery was vital. In fact, the location of this artery relative to our patient's AVM contributed to the necessity of open surgery rather than embolization.(10)

Recent individualization of the pathology, spinal arteriovenous malformations (AVMs) are characterized by an arteriovenous shunt with a true nidus and are associated with an increased risk of hemorrhage and morbidity. Classified into 4 grades, dural arteriovenous fistula is the most frequent and represents 60 to 80% of spinal AVMs. Revealed by progressive symptoms, such as gait and/or sphincter disturbances (myelopathy due to venous hypertension) or atypical sciatic type (radicular syndrome). Without treatment, the progression is slow towards definitive paraplegia. Their frequency is much rarer than that of cerebral vascular malformations with an estimated incidence between 2 and 11% of all cerebrospinal arteriovenous malformations(02). MRI consistently shows a T2 centromedullary hypersignal as well as pathological perimedullary vessels. Spinal angiography remains the key examination for diagnosis. It allows demonstrating the arteriovenous shunt as well as its arterial feeders and venous return, allowing treatment planning. Imaging also plays an important role in post-therapeutic follow-up. MRI highlights the progressive disappearance of initial anomalies, a sign of treatment efficacy. Arteriovenous fistulas with perimedullary venous drainage are a pathology that remains poorly understood. Knowledge of all these imaging elements is fundamental for early diagnosis and management, the only guarantors of good evolution after treatment.

The mechanism of development of vertebral AVMs is not completely understood; however, the majority occur at birth rather than later in life. Due to the diversion of arteriolar blood to the venous system without capillary access or resistance, more than 70% of arterial pressure is transmitted to the venous system. Venous hypertension can precipitate numerous neurological deficits secondary to mass effect and disruption of normal spinal blood flow, as well as an increased risk of hemorrhage.(12)

Classification Type I: Spinal dural arteriovenous fistula Dural arteriovenous fistulas (DAVF) are the most common type of spinal vascular malformation, accounting for up to 85% of lesions. Spinal DAVF usually affects males, with a male-to-female ratio of 5/1. (01)Additionally, this type of SAVM presents with leg weakness, back pain, or slow-progressing radicular pain in patients between 50 and 60 years old. As their name suggests, DAVFs are fistulas existing on the dural surface. These fistulas flow intradurally through the nidus, or AV communication, retrogradely through the spinal vein and to the coronal venous plexus. Therefore, the likely cause of the clinical picture is venous plexus engorgement. Type II: Intramedullary arteriovenous malformation Intramedullary AVMs are a congenital malformation that most often occurs in the thoracolumbar region, specifically at T4 and L3 levels. Clinically, Type II AVMs are frequently present in young adults with an average age in their twenties. Patients may present with myelopathy secondary to mass effect, ischemia, or hemorrhage. A previous study with pooled analysis of multiple studies on Type II AVM hemorrhages determined an annual hemorrhage rate of 4%, which increased to 10% for AVMs that had previously bled. The increased likelihood of hemorrhage with intramedullary AVMs contributes to its higher mortality rate. Type III:

Extradural-intradural arteriovenous malformations Intradural-extradural spinal arteriovenous malformations, also called juvenile vascular malformations, are a rare, complex, and locally aggressive lesion that can involve bones, muscles, dura mater, spinal cord, and nerve roots. This lesion typically presents in younger patients, leading to an average age of 15.0 years (standard deviation 10.5) in a study of 51 patients. Although Type III AVMs may present similarly to Type II, with acute hemorrhages and myelopathy, they can be differentiated by the presence of signs of local tissue involvement. An example is the presence of vertebral instability due to vertebral body involvement and degeneration after AVM hemorrhage. The likelihood that Type III AVMs have multiple feeding vessels jeopardizes the patient's surgical candidacy; therefore, the primary treatment method is embolization for symptom relief and protection against hemorrhages. Type IV: Intradural perimedullary arteriovenous fistula Intradural perimedullary arteriovenous fistulas are a rare fistula caused by a shunt between a radicular artery and intradural veins leading to engorgement of these veins. Although these lesions are rare and the actual incidence rate is difficult to define, previous studies ranged between 4% and 17% prevalence in a large series of spinal AVMs. Clinically, these lesions manifest as progressive myelopathy, acute neurological deficits, or subarachnoid hemorrhage. Due to the possibility of subarachnoid hemorrhage, embolization is typically used to eliminate the fistula.(13)

Digital subtraction angiography of the spine remains the examination of choice but requires meticulous technique. It is essential to remember that the arterial supply site can be anywhere from upper thoracic to sacral areas, with little relation to clinical level or visible nidus.(14)

It is important to note that recent studies have demonstrated operator-dependent and preventable missed diagnoses in patients. These factors included documented but unidentified lesions, lack of documented regions of interest, inadequate injection leading to poor visualization, and involvement of vessels outside the spine. Considering the reluctance due to the invasiveness of this examination, a recent study by Chen and Gailloud demonstrated that this label is a cause of historical data in which multiple intraoperative factors led to a higher complication rate. In reality, the study concluded a low risk of neurological and systemic complications associated with DSA.

MRI is less sensitive than angiography; however, MRI is the modality of choice for the initial visualization of spinal AVMs. Its use to evaluate the spinal cord and surrounding structures can help refine the differential diagnosis. Important signs that can help guide surgeons towards AVMs include the presence of cord edema with increased T2 signal due to cytotoxic edema or myelomalacia due to high-speed flow. Additionally, a dilated intervertebral vein can also be visualized on T2-weighted imaging and should suggest the diagnosis of AVM.

Magnetic resonance angiography (MRA) is often a complement to MRI used to identify the number of possible arterial feeders supplying the malformation. Therefore, this modality plays a facilitating role in subsequent imaging of spinal AVMs via DSA. Additionally, the use of MRA can lead to a significant reduction in radiation levels and contrast volume in spinal angiography.(15)

IV. Conclusion:

In conclusion, spinal dural arteriovenous fistula (SDAVF) represents a complex and often underdiagnosed vascular anomaly of the spinal cord. Despite advancements in diagnostic imaging and therapeutic interventions, SDAVF remains a diagnostic challenge and therapeutic conundrum due to its variable clinical presentation and intricate vascular anatomy. Early recognition and prompt intervention are paramount to prevent irreversible neurological deficits and optimize patient outcomes. Multidisciplinary collaboration between neurologists, neurosurgeons, neuroradiologists, and interventional radiologists is essential for accurate diagnosis and individualized treatment planning. Moreover, ongoing research efforts aimed at elucidating the pathophysiological mechanisms underlying SDAVF and refining treatment strategies are crucial for further improving patient outcomes. By fostering a deeper understanding of SDAVF and implementing comprehensive management approaches, we can strive towards better outcomes and enhanced quality of life for individuals affected by this challenging neurological condition.

References:

- [1] Lad Sp, Santarelli Jg, Patil Cg, Steinberg Gk, Boakye M. National Trends In Spinal Arteriovenous Malformations. *Neurosurg Focus.* 2009;26:1–5. [Pubmed]
- [2] Anson Ja, Spetzler Rf. Classification Of Spinal Arteriovenous Malformations And Implications For Treatment. *Bni Q.* 1992;8:2–8.
- [3] Hassler W, Thron A, Grote Eh. Hemodynamics Of Spinal Dulararteriovenous Fistulas. *J Neurosurg.* 1989;70:360–370. [Pubmed]
- [4] Abecassis Ij, Osburn Jw, Kim L. Classification And Pathophysiology Of Spinal Vascular Malformations. *Handbelin Neurol.* 2017;143:135–143. [Pubmed]
- [5] Flores Bc, Klinger Dr, White Ja, Batjer Hh. Spinal Vascular Malformations: Treatment Strategies And Outcome. *Neurosurg Rev.* 2017;40:15–28. [Pubmed]
- [6] Krings T, Geibprasert S. Spinal Dulararteriovenous Fistulas. *Am J Neuroradiol.* 2009;30:639–648. [Pmc Free Article]

- [7] Oldfield E. Surgical Treatment Of Spinal Duralarteriovenous Fistulas. *Semcerebrovasc Dis Stroke*. 2002;2:209–226.
- [8] Yasargil Mg, Symon L, Teddy Pg. Advances And Technical Standards In Neurosurgery. Vol. 11. Arteriovenous Malformations Of The Spinal Cord; Pp. 61–102. [Pubmed]
- [9] Gross Ba, Du R. Spinal Glomus (Type Ii) Arteriovenous Malformations: A Pooled Analysis Of Hemorrhage Risk And Results Of Intervention. *Neurosurgery*. 2013;72:25–32. [Pubmed]
- [10] Spetzler Rf, Zabramski Jm, Flom Ra. Management Of Juvenile Spinal Avm's By Embolization And Operative Excision. Case Report. *J Neurosurg*. 1989;70:628–632. [Pubmed]
- [11] Gross Ba, Du R. Spinal Juvenile (Type Iii) Extradural-Intraduralarteriovenous Malformations. *J Neurosurg Spine*. 2014;20:452–458. [Pubmed]
- [12] Xu Ds, Sun H, Spetzler Rf. Spinal Arteriovenous Malformations: Surgical Management. *Handbclin Neurol*. 2017;143:153–160. [Pubmed]
- [13] Nagashima C, Miyoshi A, Nagashima R, Ogawa M, Enomoto K, Watabe T. Spinal Giant Intraduralperimedullaryarteriovenous Fistula: Clinical And Neuroradiological Study In One Case With Review Of Literature. *Surg Neurol*. 1996;45:524–531. [Pubmed]
- [14] Mourier Kl, Gobin Yp, George B, Lot G, Merland Jj. Intraduralperimedullaryarteriovenous Fistulae. *Neurosurgery*. 1993;32:885–891. [Pubmed]
- [15] Chen J, Gailloud P. Safety Of Spinal Angiography: Complication Rate Analysis In 302 Diagnostic Angiograms. *Neurology*. 2011;77:1235–1240. [Pubmed]
- [16] Barreras P, Heck D, Greenberg B, Wolinsky Jp, Pardo Ca, Gailloud P. Analysis Of 30 Spinal Angiograms Falsely Reported As Normal In 18 Patients With Subsequently Documented Spinal Vascular Malformations. *Am J Neuroradiol*. 2017;38:1814–1819. [Pmc Free Article]
- [17] Jeng Y, Chen Dy-T, Hsu H-L, Huang Y-L, Chen C-J, Tseng Y-C. Spinal Duralarteriovenous Fistula: Imaging Features And Its Mimics. *Korean J Radiol*. 2015;16:1119–1131. [Pmc Free Article]
- [18] Bowen Bc, Fraser K, Kochan Jp, Pattany Pm, Green Ba, Quencer Rm. Spinal Duralarteriovenous Fistulas: Evaluation With Mr Angiography. *Am J Neuroradiol*. 1995;16:2029–2043. [Pmc Free Article]
- [19] Si-Jia G, Meng-Wei Z, Xi-Ping L, Et Al. The Clinical Application Studies Of Ct Spinal Angiography With 64-Detector Row Spiral Ct In Diagnosing Spinal Vascular Malformations. *Eur J Radiol*. 2009;71:22–28. [Pubmed]
- [20] Freudenstein D, Duffner F, Ernemann U, Rachinger J, Grote E. Recurrence Of A Cerebral Arteriovenous Malformation After Surgical Excision. *Cerebrovasc Dis*. 2001;11:59–64. [Pubmed]