Spinal dural arteriovenous fistula masquerading as a herniated disc: illustrative case

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BACKGROUND Spinal dural arteriovenous fistula (SDAVF) is a rare disorder with an unknown etiology. Often, the clinical presentation and imaging findings are misleading, causing this condition to be mistaken for other entities, such as demyelinating or degenerative spinal lesions.

OBSERVATIONS The authors report a challenging case of SDAVF in which the patient’s symptoms were initially thought to be attributable to a herniated disc based on his imaging studies at another institution. He sought the authors for a second opinion, which yielded a confirmed diagnosis of SDAVF. Due to his rapidly progressive neurological manifestations, he underwent a surgical division of the fistula using intraoperative video angiography via indocyanine green injections. His symptoms progressively improved over a 3-month period. He regained full sphincter control by 4 months, which gave him a better recovery than seen in other patients with SDAVFs, who do not generally fully regain sphincter control.

LESSONS SDAVF is a critical spinal vascular pathology that should not be overlooked in the differential diagnosis of any patient presenting with signs of progressive myelopathy. Despite its associated vague initial clinical symptoms, SDAVF typically, but not always, demonstrates a characteristic imaging appearance on magnetic resonance (MR) imaging studies; therefore, MR angiography is still required for definitive diagnosis. Surgical treatment for SDAVF is almost always definitive and curative.

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KEYWORDS arteriovenous fistula; spinal cord disorder; dura mater

The spinal cord is supplied by an arterial anastomotic network consisting of the anterior spinal artery and paired posterior spinal arteries, in addition to the anterior and posterior radicular arteries that arise from the vertebral, posterior intercostal, lumbar, or sacral arteries, depending on the level of the spinal cord. The anterior and posterior radicular arteries enter the spinal canal through the intervertebral foramina to supply the spinal nerve roots and may extend intradurally to anastomose with the anterior or posterior spinal arteries (Fig. 1). Spinal veins have a configuration similar to that of the arteries. There are usually three anterior and three posterior spinal veins that lie on the surface of the spinal cord and drain into the epidural venous plexus, which drains into the segmental veins but also communicates with the intracranial venous sinuses through the foramen magnum.1

Overall, spinal vascular malformations (SVMs) are relatively rare lesions that account for approximately 10% of central nervous system vascular malformations.2 SVMs are usually classified on the basis of their vascular angioarchitecture and nidal location. Spinal dural arteriovenous fistula (SDAVF) is the most common type of SVMs, accounting for 70%–80% of spinal cord arteriovenous shunt disorders, with the majority of patients being between the ages of 60 and 70.2 SDAVFs are abnormal direct connections between radicular arteries and radicular veins in the dura of an adjacent nerve root sleeve, with approximately 85% involving a single transdural

ABBREVIATIONS ICG = indocyanine green; MR = magnetic resonance; MRI = magnetic resonance imaging; SDAVF = spinal dural arteriovenous fistula; SVM = spinal vascular malformation.

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arterial feeder and 15% involving multiple arterial feeders originating at a single level or multiple levels unilaterally or bilaterally. In consequence, the direct arterial inflow into the coronal venous plexus, which does not have valves, raises the pressure within it (Fig. 2). With time, the cord’s venous drainage decreases, causing venous congestion and intramedullary edema, a condition known as congestive or venous hypertensive myelopathy. There is also the possibility of spinal cord ischemia or infarction, which can manifest as limb sensory abnormalities, decreased muscle strength, and sphincter dysfunction. However, when SDAVF is diagnosed promptly, it can be effectively treated by blocking the fistula and interrupting the pathological changes in the spinal vein hypertension before irreversible changes in spinal nerve function occur. Due to the rare entity of SDAVFs, diagnosis can easily be overlooked, leading to continued progressive neurological deterioration. Therefore, in order to prevent permanent neurological
disorders such as gait disorder or sphincter dysfunction, early diagnosis and treatment of SDAVs is critical. Although the diagnosis of SDAVs is possible with clinical judgment and magnetic resonance imaging (MRI) studies, the gold standard for the diagnosis is still selective spinal angiography, which not only is used for localization of the vascular lesion but also can guide its obliteration and elimination.

In this report, we present a case of a patient with SDAVF who presented with signs and symptoms of rapidly progressive myelopathy.

Illustrative Case

History
A man in his 50s presented with a 3-week history of progressive unsteadiness. He reported having a painful sensation over the previous 3 months that had started in the feet and gradually ascended into the legs. Intermittently, he also found it difficult to pass urine. His medical history was unremarkable, and he was not receiving regular medication.

Neurological Examination
On examination, the patient was alert and orientated. In the lower extremities, his tone was increased, his power was markedly decreased, and his reflexes were brisk. Proprioception was impaired in the toes, with a positive Babinski sign bilaterally. The Romberg’s test result was negative, and there were no cerebellar signs. A sensory level was identified at the level of T3. The patient’s upper limbs were normal. Three days before his presentation, the patient was seen at another institution, where he was told, on the basis of imaging studies of his spine, that his symptoms were attributable to a herniated disc at the thoracic region and that surgery (discectomy) was required. Therefore, he decided to see us for another opinion before undergoing surgery.

Formulating the Problem Representation and Building the Differential Diagnosis
With the relevant findings of the upper motor neuron signs in his lower extremities, the T3 sensory level, and the positive Babinski signs, a lesion in the upper thoracic spinal cord was suspected, and the impaired proprioception in this context suggested dorsal
column involvement, which collectively makes a diagnosis beyond a herniated disc more relatable.

**Radiological Investigations**

The patient underwent MRI of the whole spine. He also was scheduled for a bladder scan before and after voiding, which did not reveal any significant residual volume. MRI studies revealed an area of diffuse high signal in the spinal cord, representing medullary edema, extending from the T3–4 level to approximately T12 (Fig. 3A). Prominent vessels were also observed around the cord between these levels, as indicated by the small areas of hypointensity (flow voids) on the cord surface (red arrow). A mild disc bulge is also noted (green arrow), MR angiogram (B) and catheter angiogram (C) show the intercostal arteries (white arrows) and a prominent tortuous early filling midline draining vein (blue arrows), which has an abnormal direct connection to a right intercostal artery (yellow arrows). Intraoperative intradural view (D) of the arterialized vein (blue arrowhead) associated with a right-sided nerve root; note the engorged dorsal venous plexus. Standard ICG injection study (E) shows shunting into the spinal venous system (blue arrowhead indicates the beginning of the arterialized radicular vein into the dorsal venous plexus). Variation in flow between arterialized radicular vein (red, F), the distended dorsal veins over the cord (yellow), and the parenchymal veins (green) is seen.
present but was not compressing the spinal cord and was therefore not related to the presenting symptoms as we had assumed. The reported findings were highly suggestive of an underlying spinal vascular abnormality. Therefore, the patient was scheduled for a magnetic resonance (MR) angiogram (Fig. 3B), which revealed an abnormal direct connection between a right-sided intercostal artery and a midline draining vein. On the basis of these findings, we were able to make a diagnosis of SDAVF, which was later confirmed by catheter angiography (Fig. 3C), which demonstrated the fistula at the level of T7 on the right side, between a radicular artery arising from the posterior intercostal artery and the central vein of the spinal cord, consistent with the diagnosis of SDAVF.

Hospital Course
The patient had rapidly progressive symptoms; therefore, he was offered urgent treatment to obliterate the fistula before any permanent damage occurs to the spinal cord. The options were surgical division of the fistula or endovascular embolization. This patient underwent surgical division of the fistula to avoid the approximately 50% chance of recurrence if he underwent endovascular embolization.7 After the appropriate level of the fistula was determined, the dura was cut and longitudinally opened to expose the spinal cord and the associated nerve roots, along with the arterIALIZED vein (Fig. 3D). The dorsal venous plexus was engorged, and the draining vein was thickened because of the chronic effects of arterialization, and it was easily distinguishable from other normal vascular vessels. Using indocyanine green (ICG), intravenous video angiography was performed, and fistulous drainage into the intradural vessel was confirmed (Fig. 3E, F). The subarachnoid vein draining the fistula was then clipped with a titanium hemoclip (Fig. 4A), and postligation ICG injection showed normalization of venous flow (Fig. 4B, C). The dura was then reflected, and the fistulous site was observed as a small cluster of tiny arteriovenous channels, from which emerged the draining vein that entered the subarachnoid space alongside the adjacent nerve root (Fig. 4D). The cluster with its surrounding dura was then cauterized to ensure that the fistula did not remain and transmit its flow to an adjacent level. The intradural vein was then carefully cauterized and sectioned within the dural layer, and the hemoclip was left in place as a future radiographic marker for level reference if needed.

Postoperative Course
The postoperative MRI scan showed a marked reduction in the amount of cord swelling (Fig. 4E), and the postoperative MR angiogram showed that the fistula was no longer evident (Fig. 4F). The patient's symptoms progressively improved over a 3-month period, and he regained full power and sphincter control by 4 months. Given that most patients with SDAVFs do not fully regain sphincter control, our patient appears to have fared better than most, given his marked rapid improvement.

Discussion
The classification of SVMs has evolved with radiological and surgical advances that have allowed more accurate descriptions of the pathological processes involved.8 A commonly used classification system based on the work of American, English, and French investigators is as follows. Type 1 lesions are dural arteriovenous fistulas in which a segmental (radicular) artery forms a fistula with a draining vein at the dural sleeve of a nerve root. Type 2 (glomerus) lesions consist of a compact intramedullary nidus. Type 3 (juvenile) lesions are large intramedullary lesions that may extend extradurally or extraspinally. Type 4 lesions are intradural, extramedullary arteriovenous fistulas with a direct connection between the vascular supply of the spinal cord (usually the anterior spinal artery) and a vein.

Observations
Characterization of SDAVFs
Our patient had a type 1 arteriovenous malformation (a SDAVF), which is the commonest type of SVMs.6 SDAVFs can easily result in severe morbidity if left undiagnosed and untreated in the early stages.9 They occur due to a direct connection between a radicular artery and the coronal venous plexus. The arterial branches merge with the venous system like a glomerular net at the fistula located in the dural envelope of the nerve root.10,11 As a consequence of arterializing the venous system in this manner, venous hypertension, venous occlusions, ischemia, and edema develop, causing progressive myelopathy.7,12,13 Spinal cord edema begins in the most dependent part of the cord; hence, symptoms are usually of ascending nature, which explains why our patient’s symptoms started in the feet.13–15

FIG. 4. Intraoperative view (A) in which a titanium hemoclip is placed on the subarachnoid vein draining the fistula. Postligation ICG injection studies (B, C) show normalization of venous flow; the dilated dorsal veins (blue) now appear as ghostlike remnants of their former arterialized state. The dura is reflected (D), and the fistulous site is observed as a small tuft of tiny arteriovenous channels, from which exits the draining vein that enters the subarachnoid space alongside the adjacent nerve root. Postoperative T2-weighted MRI scan (E) shows a marked reduction in the amount of cord swelling. MR angiography study (F) shows a no longer evident fistula or central vein.
Diagnostic Dilemmas

SDAVFs, especially in their early stages, are often misdiagnosed as acute encephalomyelitis or intramedullary spinal cord neoplasms, and, on rare occasions, they masquerade as a nonsignificant coexisting degenerative spinal cord disorder, as in our patient. These misdiagnoses are often due to the lack of specific appearance and the atypical imaging findings. Therefore, some patients cannot be clearly diagnosed for a prolonged period of time, which can delay their treatment and may lead to irreversible damage to spinal cord functions. SDAVFs can be detected by standard spinal MRI modalities, which are the first step of choice for investigating a progressive myelopathy; however, the gold standard for diagnosis is spinal angiography, which can accurately define the lesion and guide the most appropriate intervention. As seen on MRI, SDAVFs are characterized by enlargement of the spinal cord in the lower thoracic region and conus medullaris; hyperintensity on T2-weighted imaging across multiple segments; and serpiginous, enlarged intradural vessels along the cord’s dorsal and ventral aspects, as demonstrated in our case study. Gadolinium-enhanced MRI is often helpful in highlighting the dilated intradural vascular channels and can even demonstrate cord enhancement; however, it is not indispensable for radiological diagnosis.

According to the literature, in several cases, high T2 signals in the conus or vascular flow voids in the intradural space were missed on initial imaging, only to be detected at follow-up imaging after the patient’s symptoms progressed. Moreover, there are cases in which the cord signal was hyperintense on T2-weighted images and the patient was diagnosed with transverse myelitis or neuromyelitis optica, despite the presence of flow voids, explaining why these lesions are often misdiagnosed or undiagnosed and why MR angiography should be performed.

Management Options

The treatment options for SDAVFs are either a surgical division of the fistula or an endovascular embolization. According to the literature, the success rate using surgery is higher than that of endovascular embolization. In a single-institution series with meta-analysis, 98% of patients treated surgically had their fistulas fully obliterated, compared with 46% in whom obliteration was achieved with endovascular embolization. The authors of another study found that endovascular embolization was less successful than surgical treatment in 156 patients with SDAVFs over the past 30 years. Although embolization has reached a success rate of 77%...
thanks to advancements in endovascular techniques, it is still less successful than surgery.9

Surgical Treatment

The surgical option for SDAVFs is almost always definitive and curative, with the best results generally obtained in patients treated before advanced neurological deterioration occurs, because acceptable recovery of preexisting deficits is often limited. Modern imaging technologies have enabled SDAVF surgeries to be conducted with no higher systemic stress than minor neurosurgical procedures, such as laminectomy for lumbar canal stenosis. When the procedure is performed by an experienced neurosurgeon, the operative risks are minimal with almost guaranteed fistulous obliteration and immediate relief from symptoms of venous congestion. The surgical procedure is performed with the patient under general anesthesia (Fig. 5). Intraoperative monitoring via motor and sensory evoked potential technique is recommended throughout the full procedure. To avoid hypotension or cord hypoperfusion during the procedure, the mean arterial pressure should also be maintained between 60 and 80 mm Hg before the fistulous clipping step.

Surgical Outcome

In terms of motor functions, approximately 97% of patients continue to show improvement (74% to 82%) or stabilization (14% to 22%) 1 year after SDAVF treatment. However, the recovery rate for sphincter dysfunctions is lower (5% to 58%).8,12,19,21 Behrens and Thron21 showed that the most pronounced improvement was in muscle strength (approximately 67%), followed by achieved walking distance (approximately 57%). Noticeable deteriorations in male potency (approximately 28%) and pain (approximately 24%) were observed. No change or an absence of symptoms was reported by 71% for pain, 67% for sphincter dysfunction, and 67% for sensory disturbance.21 Neurological outcomes after the surgical management of SDAVFs are closely related to the preoperative neurological function of the patient (what is lost versus what is intact), the duration of the manifesting symptoms, and the extent of medullary edema (congestion) in T2-weighted MRI studies. Patients with poor preoperative neurological functions are reported to show less benefit from surgery, but their neurological deterioration is more likely to stabilize.21

Lessons

SDAVFs are critical spinal vascular disorders that should not be overlooked in the differential diagnosis of any patient who presents with the clinical manifestations of progressive myelopathy. They generally present with vague clinical symptoms, often mistaking them for other entities of spinal disorders. Most cases demonstrate a characteristic imaging appearance on MRI studies, but still an MR angiogram is required to confirm the diagnosis. Despite the considerable advances that have been contributed to the field of imaging and endovascular technology, SDAVFs still pose a major therapeutic challenge. Despite the absence of precise guidelines for the treatment, either endovascular embolization or surgery or both have been proved to be effective options. Surgical treatment for spinal SDAVFs is almost always definitive and curative. In the modern era, operative procedures for SDAVFs are no more systemically stressful than minor neurosurgical procedures, such as discectomy or laminectomy, when performed by experienced neurosurgeons using modern imaging technologies.

References


**Disclosures**
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