

Unusual presentation of myelopathy in a previously healthy man

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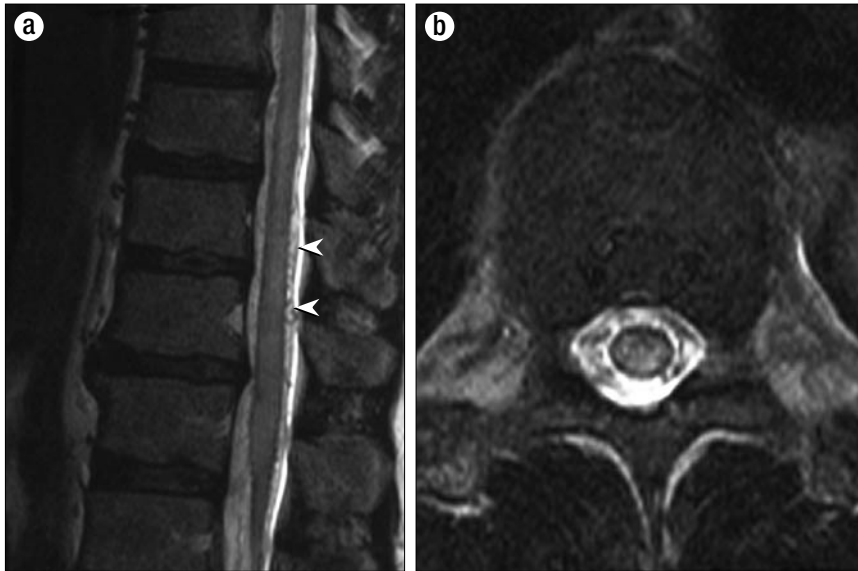


Figure 1. T2-weighted MR sequences. (a) Sagittal image of the thoracic spine demonstrates increased cord signal centrally with dorsal intradural flow voids (arrowheads). (b) Axial image through the thoracic spine demonstrates increased cord signal centrally.



Figure 2. Sagittal T1-weighted MR sequence of the thoracic spine demonstrates intradural flow voids over the dorsal aspect of the cord (arrowheads).

A previously healthy 58-year-old white man presented with progressive bilateral lower-extremity weakness. His symptoms had been intermittent for approximately 9 months, during which time he also reported several falls without loss of consciousness. He also described a history of bowel and bladder changes, including urinary urgency and occasional anal incontinence, as well as tingling within the dermal distribution of his genitalia. A lumbar puncture was done, which revealed a slightly elevated protein level, a normal glucose level, a normal white blood cell count, and no red blood cells. His cerebrospinal fluid was negative for oligoclonal bands. The patient then underwent diagnostic imaging of his brain and spinal cord. A presumptive diagnosis of transverse myelitis was made, but he was subsequently referred to Baylor University Medical Center for further diagnostic testing.

Clinical examination revealed normal mental status and speech, decreased sensation to light touch and pinprick within the lower extremities, and decreased vibratory sense in the lower extremities. Reflexes were symmetric throughout, with the exception of decreased reflexes in both ankles. Cranial nerve examination was normal, and motor examination was unremarkable. Additional laboratory tests revealed normal complete blood count, coagulation profile, and creatinine level. Aspartate aminotransferase and alanine aminotransferase levels were slightly elevated.

Magnetic resonance (MR) images of the thoracic and lumbar spine were reviewed (*Figures 1 and 2*).

What is the most likely diagnosis based on the clinical history and imaging findings? What are differential considerations? What is the appropriate management?

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DIAGNOSIS: Spinal dural arteriovenous fistula (AVF).

DISCUSSION

Spinal vascular malformations have traditionally been classified into four entities: type I, spinal dural AVFs; type II, intramedullary glomus malformations; type III, extensive juvenile malformations; and type IV, intradural perimedullary AVFs (1). A more recent publication has proposed a new classification scheme for spinal cord vascular lesions, with emphasis on location, pathophysiology, and treatment. According to this system, type I arteriovenous malformations are simply referred to as intradural dorsal AVFs and are subclassified into type A (those with a single feeding artery) and type B (those with multiple feeding arteries) (2). Intradural dorsal AVFs are the most common spinal arteriovenous malformation and represent true fistulas, arteriovenous communications without intervening capillary beds (1–3). Spinal AVFs typically occur between a dural branch of a segmental artery and an intradural vein (1–3). These abnormal arteriovenous connections result in venous hypertension and spinal cord congestion, with resulting myelopathy (1–4).

Clinical presentation

Spinal dural AVFs occur much more frequently in men and typically in those older than 40 years (1–5). Neurologic sequelae are the result of venous hypertension, secondary to abnormally increased flow within the valveless intradural venous plexus

(1–5). Spinal dural AVFs most commonly occur in the thoracolumbar region, typically between T4 and L3, and consequently, neurologic deficits are more marked in the lower extremities (2–4). Typical clinical presentation includes progressive myelopathy, bowel or bladder dysfunction, and sexual dysfunction (4). The constellation of these symptoms resulting from vascular malformation-induced venous hypertension has been referred to as the Foix-Alajouanine syndrome, as first described by Foix and Alajouanine in 1926 (3, 6).

Imaging

The MR imaging characteristics of spinal dural AVFs have been well documented (1, 3, 5, 7). A sensitive but relatively nonspecific finding on T2-weighted sequences is increased cord signal centrally, secondary to the effects of venous hypertension (4) (Figure 1). Postgadolinium-enhanced T1-weighted sequences may demonstrate mild cord enhancement at the level of the lesion (3, 5). Serpentine intradural flow voids, corresponding to the dilated venous plexus, can be seen on both T1- and T2-weighted MR sequences (Figures 1 and 2), a finding more specific for spinal vascular malformations (3, 5, 7). Recent reports have demonstrated the utility of MR angiography in localizing the level of spinal dural AVFs (5, 7). Spinal digital subtraction angiography, however, remains the gold standard in definitive localization and diagnosis of spinal dural AVFs (1, 3, 5, 7) (Figure 3).

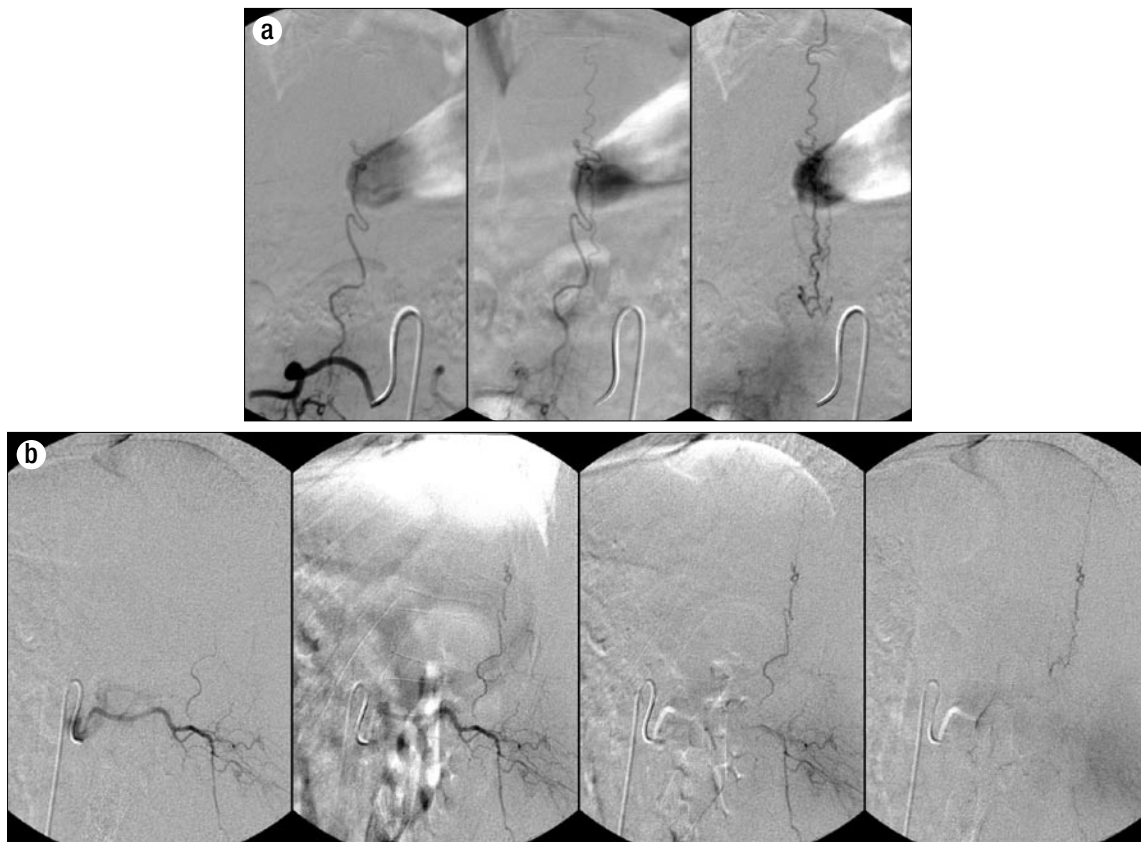


Figure 3. Spinal arteriogram of the patient in this case. (a) Anteroposterior projection. Early, mid, and late arterial phase images demonstrate a single enlarged feeding artery arising from the right L1 segmental artery, with a dorsal arteriovenous fistula at the lower thoracic spine. (b) Lateral projection. Representative arterial phase images again demonstrate a single enlarged feeding artery with an arteriovenous fistula of the dorsal aspect of the cord.

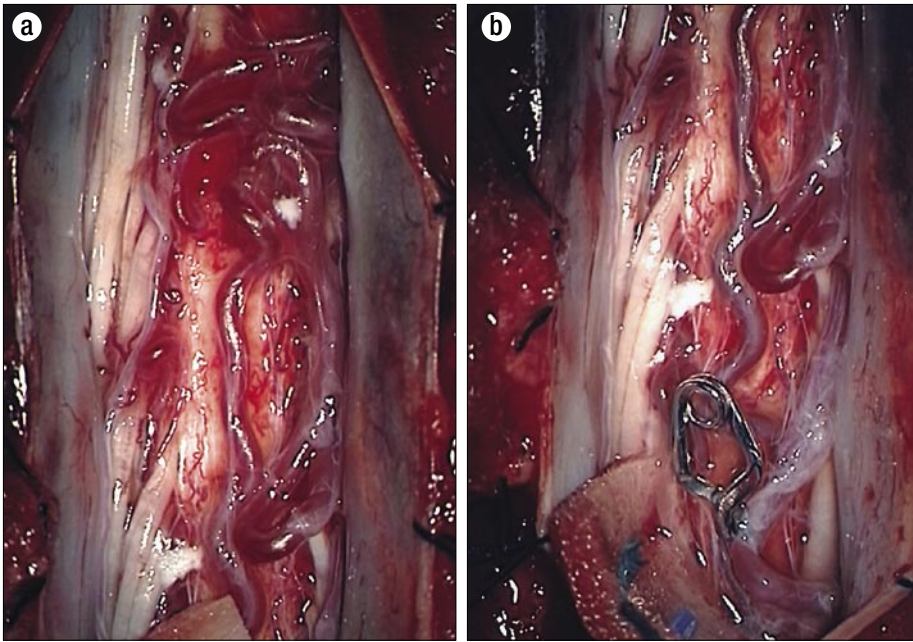


Figure 4. Intraoperative photomicrographs. (a) Preligation image demonstrates a single enlarged feeding vessel with an intradural dorsal arteriovenous fistula. (b) Postligation image shows the surgical clip on the parent vessel.

Differential diagnosis

The differential diagnosis of progressive myelopathy includes many diseases, such as Guillain-Barre syndrome, disc herniation, transverse myelitis, and intramedullary tumor (2, 3, 5). The classic clinical history, patient demographics, and MR imaging characteristics should alert the clinician and radiologist to the diagnosis of spinal dural AVF (1–5, 7).

Management

Treatment of dural AVFs involves interruption of the fistula by occlusion of either the supplying artery or the draining vein (2, 3). This occlusion can be achieved by either endovascular arterial embolization or by surgical interruption of the fistula (1–3). Endovascular embolization of dural AVFs is performed using acrylic polymers. While endovascular techniques are less invasive, the overall recurrence rates have been higher than for direct surgical interruption of the fistula (1–3). Endovascular embolization, however, can be a useful adjunct to surgery, especially in those patients with rapid neurologic decompensation who are already undergoing a diagnostic arteriogram to localize the level of the dural AVF (3). Surgical treatment involves laminectomy at the site of the vascular malformation and intraoperative localization of the draining medullary vein. The draining vein is subsequently coagulated and divided (1, 3, 4).

A multilevel spinal arteriogram of the patient in our case showed a single enlarged feeding artery arising from the right L1 segmental artery, leading to a dorsally located AVF at the T11 level, with early arterialized veins egressing both superiorly and inferiorly (Figure 3). He subsequently underwent a right-sided trilevel laminectomy from T11 to L1. Angiographic findings were confirmed surgically (Figure 4a), and the feeding artery and two large draining veins were surgically interrupted (Figure 4b). Afterward, his neurologic deficits were markedly improved.

CONCLUSION

Spinal dural AVFs can be an uncommon cause of myelopathy, and while their identification can be a diagnostic challenge, their management is relatively straightforward (1–5). The clinical and imaging characteristics

should lead the clinician to the correct diagnosis, and prompt clinical intervention should be pursued to avoid severe and/or progressive debilitating neurologic deficits (1–4).

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