Spinal dural arteriovenous fistula with intramedullary cord hemorrhage

Diagnostic challenges

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Practical Implications

A myelopathy with fluctuations should prompt consideration of a spinal dural arteriovenous fistula; steroid induced worsening may be a clue.

Spinal dural arteriovenous fistula (SDAVF) consists of an acquired arteriovenous shunt between a dural artery and perimedullary veins.¹ This results in arterialization of radicular veins, venous hypertension, and decreased spinal cord perfusion.¹ The condition has a male predominance and presents as a myelopathy in the fifth or sixth decade. Initial misdiagnosis is common.¹⁻³ We present a man with a SDAVF and associated intramedullary cord hemorrhage.

Case report

A 36-year-old man was admitted with a 4-month history of lower extremity numbness and weakness. His symptoms began with left followed by right foot paresthesias. Subacute onset of lower limb weakness 2 months after symptom onset prompted him to seek medical attention locally. There was no sphincteric dysfunction. Spine MRI showed a centrally located T1 hyperintensity (figure, A) with surrounding susceptibility-related hypointensity (figure, B) at T9–T10 and edema-related central T2 hyperintensity of the cord from T7 to L1 (figure, B). CSF studies were remarkable only for a slightly elevated protein. A vascular myelopathy was suspected. MRI showed abnormal perimedullary vascular flow voids suggesting a SDAVF. A cord arteriovenous malformation (AVM) and cavernous malformation were also in the differential diagnosis. Magnetic resonance angiography (MRA) and 2 spinal angiograms did not identify a vascular abnormality. A diagnosis of transverse myelitis or hemorrhagic demyelination was then proposed.

A 5-day course of IV methylprednisolone was given for presumed transverse myelitis. The patient reported lower extremity “stunning” immediately following steroid administration. He used the word stunning to describe worsening of his motor and sensory deficits. Days later, he noted recovery from the steroid-induced worsening. This was mistakenly interpreted as steroid-related improvement. This recurred with a repeat steroid trial.

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Figure MRI and angiography findings in spinal dural arteriovenous fistula with intramedullary hemorrhage

Sagittal T1-weighted (A) and T2-weighted (B) 1.5T MRI of the thoracic spine show a centrally located area of intramedullary spinal cord hyperintensity at T9–10 (A, B: arrow) with surrounding susceptibility-related hypointensity (B: dotted arrow) and edema (B: arrowheads) extending from T7 to L1. No definite abnormal vascularity was noted following administration of a contrast bolus. A 3T sagittal (C) and reconstructed coronal (D) thoracic spine fast imaging employing steady-state acquisition series shows multiple intradural tortuous dorsal and ventral flow voids (arrows). A left T3 intercostal artery injection during a spinal angiogram using DynaCT (E) shows a dural arteriovenous fistula (arrow) in the region below the left T3 pedicle. The fistula is seen draining into an intraspinal radiculomedullary vein (arrow) that extends superiorly and inferiorly as perimedullary veins (dotted arrows) away from T3 (F).

Four months following symptom onset, he was referred to us. Examination revealed moderately severe spastic paraplegia. He was wheelchair-bound with minimal distal left lower limb movement. Pinprick perception was impaired below T12 and vibratory sensation was absent at the toes. Repeat MRI was performed at 3T field strength. Fast imaging employing steady-state acquisition (FIESTA) sequences showed multiple tortuous intradural ventral and dorsal flow voids (figure, C and D). Given the presence of epidural lipomatosis and a relatively compressed dorsal subarachnoid space, these magnetic resonance series gave greater confidence regarding the presence of a vascular abnormality than did standard images. Repeated spinal angiography revealed a SDAVF on left T3 intercostal artery injection (figure, E and F); this segmental artery had not been catheterized on the prior angiograms. A T3 laminectomy was performed with SDAVF ligation. Immediate postoperative MRI demonstrated diminished distal cord edema. Ten days following his procedure, the patient was ambulating with a walker. At 2-month follow-up telephone contact, the patient reported continued improvement, including the ability to take a few steps unaided.

DISCUSSION

The clinical course of SDAVF is typically progressive and is due to spinal cord venous hypertension. SDAVFs rarely bleed. Subarachnoid hemorrhage⁴ and hematomyelia⁵ have been reported but are exceedingly rare. In over 200 SDAVFs evaluated at our institution over the past 3 decades, this is the first to present with cord hemorrhage. Our patient's acute worsening was due to intramedullary hemorrhage. This occurred in the area of lower thoracic cord edema and was likely due to severe venous hypertension. A hemorrhagic presentation is much more common with cord AVMs.¹ Patients with a SDAVF may show worsening with exercise, upright posture, and Valsalva maneuver.⁶ Also reported is steroid-induced worsening,⁶ which was seen in our patient twice following steroid administration. The presence of steroid-induced worsening may represent a novel means of redirecting the diagnosis from an inflammatory myelitis to a SDAVF. It has been suggested that steroid-induced worsening may be due to the hypertensive effects of hypercortisolemia.⁶

Diagnosis first relies on MRI that shows central cord hyperintensity on T2-weighted images and increased perimedullary flow voids, most commonly dorsal to the cord.² In our patient, these flow voids were better appreciated on 3T FIESTA imaging (figure, C). FIESTA sequences have a very high spatial and contrast resolution for intrathecal vessels relative to CSF. Catheter angiography remains the gold standard of diagnosis.² Despite SDAVF being in the initial diagnostic consideration, 1 MRA and 2 spinal angiograms did not definitively diagnose or localize the fistula. Most SDAVFs arise from segmental arteries between T6 and L2.¹ The SDAVF may be remote from cord parenchymal abnormalities; our patient had a left T3 fistula with the cord parenchymal abnormalities being most evident from T7 to L1. This emphasizes the need for spinal angiography to interrogate every possible arterial contributor to the spine if a fistula is not initially found but is still suspected.

The improvement in our patient was striking. This reflects the fact that deficits in SDAVF are a consequence of venous hypertension rather than cord infarction. Surgical correction may result in improvement even with longstanding severe deficits.^{3,7}

The diagnostic challenge in our patient was related to the young age at presentation, acute worsening, presence of spinal cord hemorrhage, and the failure of fistula detection on initial angiograms.

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