Absence of abnormal vessels in the subarachnoid space on conventional magnetic resonance imaging in patients with spinal dural arteriovenous fistulas

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Spinal dural arteriovenous fistula (DAVF) is an uncommon condition that can be difficult to diagnose. This often results in misdiagnosis and treatment delay. Although conventional MRI plays an important role in the initial screening for the disease, the typical MRI findings may be absent. In this article, the authors present a series of 4 cases involving patients with angiographically proven spinal DAVFs who demonstrated cord T2 prolongation on conventional MRI but without abnormal subarachnoid flow voids or enhancement. These cases suggest that spinal DAVF cannot be excluded in symptomatic patients with cord edema based on conventional MRI findings alone. Dynamic Gd-enhanced MR angiography (MRA) was successful in demonstrating abnormal spinal vasculature in all 4 cases. This limited experience provides support for the role of spinal MRA in patients with abnormal cord signal and symptoms suggestive of DAVF even when typical MRI findings of a DAVF are absent.

KEY WORDS • spinal dural arteriovenous fistula • magnetic resonance angiography • perimedullary flow voids

Although spinal DAVF is the most common variation of spinal vascular malformations, accounting for 60%–80% of cases, they are still rare entities that affect only an estimated 5–10 patients per million per year. They are slow-flow arteriovenous shunts characterized by an abnormal connection between a radicular artery and vein that is found most often in, or near, the dura of a nerve root sleeve. The resulting venous congestion and hypertension, first suggested by Aminoff et al. in 1974, eventually results in a progressive congestive myelopathy from the decrease in the arteriovenous pressure gradient, cord ischemia, and eventual infarction with neuronal loss. Hassler and Thron confirmed this hypothesis by demonstrating a venous pressure in a DAVF of approximately 70% of the patient’s systemic arterial pressure. The description by Marie et al. of a subacute necrotizing myelopathy is now widely accepted to represent the same pathological process.

Spinal DAVF is considered an acquired disease whose underlying cause remains unknown. Patients most often present in the 5th or 6th decade of life, with a strong male predominance of approximately 5:1 in reported series. Although the majority of spinal DAVFs are found in the mid-to-lower thoracic spine, venous congestion and subsequent myelopathy often begins in the caudal aspect of the spinal cord irrespective of the actual fistula level. Patients consequently most often present with nonspecific symptoms involving changes in sensation and strength in the lower extremities, lower-extremity pain sometimes exacerbated by exercise, and disturbances in gait. These symptoms overlap significantly with those of more common diseases such as spinal canal stenosis and polyneuropathy. As the myelopathy progresses, patient often will experience continued loss of lower-extremity strength leading to paraplegia, as well as develop abnormalities in micturition, defecation, and sexual function.

Conventional MRI is frequently the initial diagnostic examination in patients with a spinal DAVF, suspected or otherwise, and this represents a critical decision point for...
the diagnosis.\textsuperscript{10,13,17,24} The MRI findings of spinal cord T2 prolongation, intramedullary enhancement, and prominent vessels in the subarachnoid space as demonstrated by flow voids on T2-weighted imaging and enhancement on postcontrast T1-weighted imaging are considered characteristic for this disease.\textsuperscript{11,14,22,24} Although this combination of findings on conventional MRI is thought to be both fairly sensitive and specific for the diagnosis of a spinal vascular malformation, these findings may not all be present in a significant proportion of patients.\textsuperscript{11,13,14,22,24}

We present a series of 4 cases involving patients with proven spinal DAVF who demonstrated spinal cord edema and enhancement without abnormal flow voids or enhancement in the subarachnoid space on conventional MRI. Three patients presented at our academic referral center, and the fourth presented at another academic practice. Three patients were male and one was female, and their ages ranged from 57 to 72 years. All patients were evaluated by conventional MRI, consisting of sagittal T1-weighted, sagittal and axial T2-weighted, and Gd-enhanced sagittal and axial T1-weighted imaging. The diagnosis of spinal DAVF was subsequently confirmed by conventional spinal angiography, and all patients went on to be treated with surgical, endovascular, or multidisciplinary approaches. Dynamic Gd-enhanced MRA was performed prior to catheter angiography in 3 patients and was successful at suggesting the diagnosis of a spinal DAVF by demonstrating abnormal vessels on the surface of the spinal cord; MRA also helped to target subsequent catheter angiography in these patients by correctly identifying the fistula level. In a fourth patient, dynamic Gd-enhanced MRA was performed to evaluate for residual flow in a cervical DAVF after an attempted endovascular embolization; MRA correctly identified the residual fistula, which was subsequently closed.

Case Reports

Case 1

This 70-year-old man with a history of diabetes, peripheral vascular disease, and arthritis presented to our institution with a 4-year history of progressive lower-extremity pain and weakness. The patient’s initial symptoms consisted of bilateral lower-extremity pain on exertion, and he was subsequently diagnosed with claudication secondary to peripheral vascular disease. He underwent lower-extremity arterial angioplasty on 2 separate occasions for these symptoms 4 and 2 years prior to presenting to our institution. The patient had also received regular Synvisc knee injections for osteoarthritis with temporary relief of his lower-extremity pain. At presentation, the patient was using a walker to ambulate and had bilateral hip flexor weakness of (4+/5), as well as slight weakness of left knee extension (4+/5). A Romberg sign was present, and the patient had a wide-based, unsteady gait. He also had markedly decreased vibration and proprioception sensation to the midshins bilaterally. Both temperature and light touch sensation in the lower extremities were intact. Deep tendon reflexes were 1+ at the patella; there was no right ankle jerk, and the left ankle jerk was 1+.

Conventional MRI demonstrated a long segment of increased intramedullary T2 prolongation in the thoracic cord beginning at the T5–6 level and extending to the conus. There was no abnormal flow void or enhancement noted in the subarachnoid space. The presence of a DAVF, however, was considered based on the patient’s symptoms, and he therefore underwent dynamic contrast-enhanced MRA. That examination demonstrated abnormal vessels dorsal to the inferior thoracic cord and conus medullaris. A prominent vessel was evident extending to the left L-1 neural foramen, suggesting the level of the fistula. Follow-up spinal angiography confirmed the presence of a spinal DAVF originating from the left T12–L1 level with enlargement of the coronal venous plexus (Fig. 1). More precise localization was not possible due to tortuosity and atherosclerosis involving the aortic segmental arteries. The patient subsequently underwent a successful microsurgical resection of the fistula. On follow-up examinations his lower-extremity weakness and sensory changes remained stable, likely due to the long-standing duration of his myelopathy prior to treatment, but no progression was evident.

Case 2

This 72-year-old man presented to an outside institution with a 2-month history of progressive upper- and lower-extremity myelopathic symptoms of transient paresthesias, as well as mild gait difficulty. Conventional MRI of the cervical spine performed at the outside hospital at 2 time points demonstrated abnormal T2 prolongation throughout the cervical and upper thoracic spinal cord, as well as anterior intramedullary enhancement in the cervical cord. The patient was referred to our institution for a spinal cord biopsy because of concern for an underlying spinal cord tumor. A review of the outside MRI studies revealed no evidence of abnormal flow void or enhancement in the cervical or thoracic subarachnoid space. Cervical spinal canal stenosis secondary to degenerative disease was present, however.

Based on our previous experience and the patient’s symptoms, spinal angiography was performed to evaluate the possibility of a spinal vascular malformation. A cervical DAVF was found—supplied by an intradural branch of the right vertebral artery and with venous drainage both anterior and posterior to the cervical cord. An attempt was subsequently made to embolize the fistula. However, following the intervention there was clinical concern for continued patency of the DAVF, which was confirmed by follow-up dynamic contrast-enhanced MRA demonstrating enlarged, tortuous vessels along the surface of the cervical cord (Fig. 2).

This case was subsequently managed with a multidisciplinary approach. After the failed endovascular embolization, microsurgical ligation of an arterialized vein at the C-1 level was performed, but the patient’s symptoms did not improve, and a follow-up conventional angiogram demonstrated an additional, previously unrecognized, arterial feeder arising from the intradural right vertebral artery. This second arterial feeder was successfully embolized with Onyx, and a final postintervention angiogram demonstrated no evidence of residual DAVF. The patient has subsequently undergone extensive physical therapy.
Absence of typical MRI findings in patients with spinal DAVF

with improvement in his gait, to the point where he can ambulate with a cane or walker, and resolution of his paresthesias.

Case 3

This 57-year-old man with a medical history significant for Lyme disease presented to our institution with a 4-month history of progressive lower-extremity paresthesias, extending to the perineum, and gait instability. Conventional MRI of the spine demonstrated a long segment of abnormal, intramedullary, T2 prolongation involving the thoracic spinal cord with associated faint enhancement on Gd-enhanced T1-weighted imaging. No abnormal flow voids or abnormal enhancement were evident in the subarachnoid space.

Dynamic contrast-enhanced MRA, however, demonstrated prominent, tortuous vessels anterior to the midthoracic cord suggestive of a vascular malformation. In addition, MRA suggested potential fistula levels on the right at T7–8 and on the left at T6–7. Conventional spinal angiography demonstrated a DAVF that was supplied by both a radicular branch of the right T-8 intercostal artery, with the artery of Adamkiewicz arising from the left T-7 intercostal artery (Fig. 3). The fistula was subsequently successfully embolized using a combination of 30% n-butyl 2-cyanoacrylate glue and 70% Lipiodol. Four-year clinical follow-up

Fig. 1. Case 1.  A and B: Sagittal thoracic T2-weighted (A) and Gd-enhanced T1-weighted (B) images demonstrating a long segment of T2 hyperintensity and intramedullary enhancement in the thoracic spinal cord extending to the conus medullaris in a 70-year-old man with gradual onset of bilateral lower-extremity weakness and alteration of sensation. There is no evidence of abnormal flow voids or enhancement in the subarachnoid space to suggest abnormal spinal vasculature.  C: Dynamic contrast-enhanced MR angiogram demonstrating abnormal, dilated, and tortuous vessels extending along the dorsal aspect of the thoracic spinal cord raising the possibility of a spinal DAVF. Magnetic resonance angiography was also suggestive of a possible fistula location on the left at the L-1 level.  D: Image subsequently obtained by catheter angiography, confirming the diagnosis and demonstrating the feeding radicular artery arising from the left T12–L1 level.

Fig. 2. Case 2.  A and B: Sagittal cervical-thoracic T2-weighted (A) and Gd-enhanced T1-weighted (B) images demonstrating a long segment of T2 hyperintensity throughout the cervical and superior thoracic spinal cord with anterior intramedullary enhancement in the cervical spine in a 72-year-old man with upper and lower extremity myelopathy symptoms who was referred to our institution for spinal cord biopsy due to concerns for an intramedullary tumor. There is no evidence of abnormal flow voids or enhancement in the subarachnoid space to suggest abnormal spinal vasculature.  C: Dynamic contrast-enhanced MR angiogram obtained after attempted endovascular embolization demonstrating abnormal, dilated, and tortuous vessels extending along the ventral and dorsal aspects of the cervical cord suggestive of residual spinal DAVF.  D: Image obtained by catheter angiography confirming the MRA findings and demonstrating arterial supply from a branch of the intradural right vertebral artery.
demonstrated improvement in the patient’s lower-extremity paresthesias with no residual gait abnormality.

Case 4

This 66-year-old woman had progressive lower-extremity weakness and paresthesias over the course of several months. A conventional MRI examination demonstrated a long segment of abnormal T2 prolongation in the thoracic spinal cord that extended to the conus medullaris along with mild intramedullary enhancement. No abnormal flow voids or enhancement were identified in the subarachnoid space. However, dynamic contrast-enhanced MRA demonstrated prominent, tortuous vessels along the dorsal aspect of the thoracic spinal cord, which—along with the symptoms—suggested a spinal DA VF with possible localization of the fistula at the right T-12 level. Follow-up conventional spinal angiography confirmed the presence of a spinal DA VF supplied by the right T-12 segmental artery (Fig. 4). The patient was treated successfully with surgical ligation of the fistula.

Discussion

A delay in the diagnosis of spinal DA VF can be attributed to both the rarity of the diagnosis and the nonspecific quality of the presenting symptoms. Jellema et al. found in their review of the literature that the delay between symptom onset and diagnosis ranged between 12 and 44 months. Unfortunately, it is not unusual for the patient’s symptoms to significantly progress during that time interval. These symptoms include worsening lower-extremity weakness as well as the onset of additional symptoms, such as micturition, defecation, and sexual function. Successful treatment requires occlusion of the draining, arterialized, intradural vein, which has been found to provide a lasting and durable treatment of the symptoms of spinal DA VFs. Currently, surgical ligation, endovascular embolization, and a multidisciplinary approach are all used for the treatment of spinal DA VFs.

Once the diagnosis of a spinal DA VF has been made and the fistula has been successfully closed, a majority of patients, up to 80%–90% in some series, will demonstrate either improvement or stabilization of symptoms, particularly lower-extremity strength. Aghakhani et al. showed that even patients with complete paraplegia at the time of treatment can demonstrate a significant improvement in function following intervention. This represents a dramatic divergence from the natural history of spinal DA VF, which consists of progressive deterioration, as demonstrated by Aminoff and Logue, who showed that 50% of patients will be wheelchair-bound 3 years after presentation. While a few studies have described a possible late deterioration of patient function on longer-term follow-up, these findings have served as an impetus to increase knowledge of spinal DA VF in the general medical community in the hope of reducing delays in diagnosis and thereby improving patient outcomes.

Spinal cord edema on conventional MRI has been found to be very sensitive for the diagnosis of spinal DA VF, with reported sensitivity close to 100%. However it is a nonspecific finding that can be attributed to much more common entities such as demyelination, trauma, and spinal canal stenosis secondary to degenerative disease of the spine. Our experience shows that even using current MR technology, the more specific findings of abnormal vasculature in the subarachnoid space may be absent in some patients with spinal DA VFs on conventional MR images. Thiex et al. reported a similar experience in a patient with a spinal DA VF in whom conventional MRI failed to demonstrate abnormal vessels in the subarachnoid space. The results of earlier and larger published series support our assertion, with some reporting only 45%–55% of spinal DA VF patients dem-
onstrating abnormal spinal vasculature on conventional MRI. However, this lower range of reported sensitivities may be attributable, at least in part, to the limitations of less sophisticated MRI units in earlier studies. The diagnosis of a spinal DAVF therefore cannot be excluded when spinal cord edema is encountered on conventional MRI in the appropriate clinical setting. However, the lack of specificity of spinal cord edema and symptoms alone are frequently insufficient to justify conventional spinal angiography, given its small, but significant, risks and complexity.

Although dynamic Gd-bolus MRA may have a role in patients with typical findings of DAVF because it can reduce both fluoroscopy time and contrast agent volume,10,20,26,31 necessary on conventional angiography, our small case series suggests that it may have a very important role in establishing the diagnosis of spinal DAVF. Other published reports demonstrating high sensitivity and reasonably high specificity of dynamic Gd-enhanced MRA for detection of spinal DAVFs support this assertion.3,10,26

Clinical history and the suspicion of a spinal DAVF should ultimately remain the deciding factors that determine whether one should proceed with spinal angiography. However, in cases involving patients who do not present with diagnostic MRI findings or typical symptoms of a spinal DAVF, MRA examination may provide compelling evidence to go on to invasive conventional angiography.

Disclosure

The authors report no conflict of interest concerning the material or methods used in this study or the findings specified in this paper.

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