Deterioration After Surgical Treatment of Spinal Dural Arteriovenous Fistula Associated With Spinal Perimedullary Fistula
—Case Report—

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Abstract

Spinal dural arteriovenous fistulas (SDAVFs) are the most common type of vascular malformations of the spine and are defined as abnormal arteriovenous shunts within the dura. SDAVFs are considered to be acquired and should be distinguished from congenital intradural perimedullary arteriovenous fistulas (PMAVFs). A 32-year-old female presented with both SDAVF and PMAVF, manifesting as a slowly progressive paraparesis over a 6-month period. Initial spinal angiography demonstrated an SDAVF in the sacral region and was terminated with incomplete demonstration of all segmental arteries. The fistula was obliterated by surgery and the patient showed transient postoperative improvement followed by delayed deterioration 2 months later. Magnetic resonance (MR) imaging showed many hypointense flow voids around the cord. The second angiography verified a PMAVF in the lumbar region and complete obliteration of the SDAVF. The fistula was closed by surgery and the patient improved slightly. Surgical results of SDAVFs are generally good. Therefore, if a patient fails to improve or deteriorates further after surgery with persistent perimedullary vessel abnormalities on MR imaging, the possibility of reopening of the fistula or the presence of another fistula should be considered and repeat angiography must be performed, especially if the initial angiography was incomplete.

Key words: spinal cord, arteriovenous fistula, dural fistula, perimedullary fistula

Introduction

Spinal dural arteriovenous fistulas (SDAVFs) are the most common type of vascular malformations of the spine and are defined as abnormal arteriovenous shunts within the dura supplied by the dural arteries and drained by the perimedullary veins. Intradural drainage of the fistula leads to venous hypertension and swelling as well as edema within the spinal cord.⁵,⁸,¹² SDAVFs are usually acquired⁴,¹¹,¹² and present as single lesions.²,⁶,¹²,¹⁵ SDAVF treatment involves complete obliteration of the fistula, and the results of surgery are generally good.⁵,⁸,¹³ However, after treatment, if the patient’s condition fails to improve or deteriorates further with persistent perimedullary vessel abnormalities on magnetic resonance (MR) imaging, the possibility of reopening of the fistula or the presence of multiple fistulas should be considered.¹,²,⁵,¹⁰,¹²,¹³

We present a case of SDAVF associated with perimedullary AVF (PMAVF), and describe the clinical presentation, neuroradiological findings, and treatment procedures.

Case Report

A 32-year-old female suffered slowly progressive paraparesis over a 6-month period. Her past history was unremarkable. T₂-weighted MR imaging taken at another hospital revealed a slight enlargement of the spinal cord (from T9 to L1) with a central hyperintense area, and many hypointense flow voids on the surface of the spinal cord and cauda equina (Fig. 1A). Three-dimensional computed tomography (CT) angiography showed an abnormal network around the spinal cord and cauda equina (Fig. 1B, C). SDAVF was suspected, and T10-T11 laminectomy was performed to confirm the diagnosis, but the fistula could not be identified epidurally and intradurally. Her paraparesis and impairment of urinary function slowly worsened postoperatively, and subsequently she was referred to our hospital.

Neurological examination showed mild weakness and bilateral sensory impairment of the lower limbs, more pronounced on the left. She could walk with the aid of a cane. The leg reflexes were markedly increased and bilateral Babinski signs were present, as well as significant
impairment of bowel and bladder functions. Spinal angiography demonstrated SDAVF fed by the left lateral sacral artery and drained by the ascending perimedullary veins (Fig. 2A–C). Most of the segmental arteries were catheterized, but selective angiography of the 9th intercostal artery was not performed to exclude the presence of multiple fistulas. Furthermore, angiography of the lower thoracic aorta showed no apparent abnormalities of the left 9th to 11th intercostal arteries (Fig. 3A). Concurrent PMAVF remained undetected on initial angiography.

S1–S3 laminectomy was performed, and the arterialized draining vein was obliterated epidurally and intradurally near the end of the dural sac (Fig. 2E, F). After surgery, the patient showed mild improvement in her symptoms; she could walk almost freely. However, her neurological status deteriorated again after 2 months. She developed weakness of both legs and the symptoms became as severe as prior to the operation. Follow-up MR imaging showed cord swelling and many serpiginous hypointense flow voids around the spinal cord, indicating that the fistula was still patent (Fig. 4A, B). Because of the recurrent deterioration in her neurological status and persistent pathological vessels on the MR images, second angiography was performed 3 months after the second surgery, which showed the completely occluded fistula with preservation of the anterior spinal artery (Fig. 3D). Postoperative MR imaging revealed improvement in the cord swelling and disappearance of the flow voids around the spinal cord (Fig. 4C). The patient showed mild improvement in motor weakness and sensory impairment in both legs. However, gait disturbance, and bladder and bowel dysfunction continued postoperatively. She was transferred to another hospital for rehabilitation.

**Discussion**

Spinal arteriovenous malformations (AVMs) can be divided into dural AVFs and intradural AVMs, each having different pathological, clinical, and radiological features. SDAVFs are defined as abnormal arteriovenous shunts within the dura supplied by dural arteries and drained by perimedullary veins. SDAVFs constitute approximately 70% to 80% of all spinal AVMs. SDAVFs are acquired lesions, similar to intracranial dural fistulas, and show symptoms of myelopathy due to increased venous pressure in the coronal venous plexus. Patients usually present with progressive myelopathy associated with motor and sensory deficits, and bowel, sexual, and bladder dysfunction, and mainly affect men aged 50 years and above. A majority of the lesions occur in the thoracolumbar region, whereas sacral involvement is rare.

Intradural AVMs are further classified as intramedullary AVMs, where the nidus is located within the spinal cord parenchyma, and PMAVFs, where the nidus is absent but a direct arteriovenous shunt on the pial surface of the spinal cord is present. PMAVFs originate from the spinal artery and are characterized by the presence of a direct fistula drained by an enlarged venous network, and constitute approximately 10% to 20% of all spinal AVMs. PMAVFs are considered to be congenital lesions, and most patients present with slowly progressive myelopathy due to venous hypertension. In the present case, initial spinal angiography with incomplete demonstration of all segmental arteries showed an SDAVF in the sacral region fed by the left lateral sacral artery and drained intradurally by an arterIALIZING draining vein. The second angiography verified a small and single PMAVF supplied by the anterior spinal artery from the 9th intercostal artery and drained by a venous network. Both fistulas seemed to be quite different in nature.

Although the exact pathogenesis of SDAVFs is not clear, venous thrombosis and hypertension are considered to be major factors governing the formation of SDAVFs. Therefore, a single SDAVF or PMAVF is postulated to aggravate elevated pressure, venous stagnation, and subsequent thrombosis, resulting in formation of another SDAVF. SDAVF is usually single, and the frequency of double or multiple fistulas is reported to be approximately 1% to 2%. The association of a PMAVF with an SDAVF is even less common, with cases in only four patients (three males and one female, aged 32–61 years), including

![Fig. 1 A: Preoperative sagittal T2-weighted magnetic resonance image revealing a slight enlargement of the spinal cord with a central hyperintense area, and many hypointense flow voids on the surface of the spinal cord and cauda equina. B, C: Three-dimensional computed tomography angiograms showing an abnormal vascular network around the cord. The right 11th intercostal artery is considered to be a feeding artery (arrow, C).](image-url)
Fig. 2 A–C: First spinal angiograms, anteroposterior view (A, C) and lateral view (B), revealing a spinal dural arteriovenous fistula fed by left lateral sacral artery. D: Second spinal angiogram showing no opacification of the fistula. E, F: Intraoperative photographs showing the distal end of the dural sac is opened and the arterialized draining vein is obliterated epidurally and intradurally with Weck titanium clips (E), with the epidural draining vein (arrow, F).

Fig. 3 A–D: First lower thoracic aortogram (A) showing the left 9th to 11th intercostal arteries to have no apparent abnormalities. Second lower thoracic aortograms, arterial phase (B) and venous phase (C), showing a perimedullary arteriovenous fistula fed by the anterior spinal artery from the left 9th intercostal arteries. Retrospective analysis of the first and second aortograms identified a serpentine vessel along the cord (arrow, A), consistent with the feeder part of a concurrent perimedullary arteriovenous fistula verified by the second angiography (B, C). Third lower thoracic aortogram (D) shows obliteration of the fistula. E, F: Intraoperative photographs showing a network of draining veins and the fistula is occluded with a microdissector.

the present patient (Table 1). The incidence of concurrent spinal AVMs of a different nature may be underestimated because complete spinal angiography is not systematically performed.

MR imaging is useful for the assessment of patients with progressive myelopathy. Furthermore, this noninvasive...
procedure can provide an initial diagnosis of spinal AVM. T2-weighted MR imaging reveals slight enlargement of the spinal cord with hyperintense central area and enlarged tortuous epidural veins presenting as flow voids, as the epiphenomena of SDAVFs. MR imaging showed findings consistent with the presence of SDAVF in 146 of 147 patients (99.3%). T2-weighted MR imaging showed hyperintense areas involving the lower spinal cord and conus medullaris, as well as variable length of perimedullary vessel abnormalities that are usually prominent on the surface of the spinal cord and conus in 126 patients (85.7%).

Table 1  Reported cases of spinal dural arteriovenous malformation (SDAVF) associated with perimedullary arteriovenous malformation (PMAVF)

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age (yrs)/Sex</th>
<th>Presentation</th>
<th>Type</th>
<th>Level</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Dam-Hieu et al. (2001)</td>
<td>49/M</td>
<td>myelopathy, improvement after 2nd ope.</td>
<td>a) SDAVF</td>
<td>rt T6</td>
<td>SO</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>b) PMAVF</td>
<td>rt L1</td>
<td>SO 3 wks after the 1st ope.</td>
</tr>
<tr>
<td>2</td>
<td>Morgalla et al. (2004)</td>
<td>61/M</td>
<td>progressive paraparesis, recurred weakness 2 mos after the 1st ope.</td>
<td>a) SDAVF</td>
<td>rt T9</td>
<td>SO</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>b) PMAVF</td>
<td>lt T7*</td>
<td>SO 2 mos after the 1st ope.</td>
</tr>
<tr>
<td>3</td>
<td>Krings et al. (2006)</td>
<td>35/M</td>
<td>progressive myelopathy</td>
<td>a) SDAVF</td>
<td>rt T6</td>
<td>SO of both in one stage SO**</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>b) PMAVF</td>
<td>rt T5</td>
<td>SO**</td>
</tr>
<tr>
<td>4</td>
<td>Present case</td>
<td>32/F</td>
<td>progressive paraparesis, recurred weakness 2 mos after the 2nd ope.</td>
<td>a) SDAVF</td>
<td>sacrum</td>
<td>SO</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>b) PMAVF</td>
<td>lt T9*</td>
<td>SO 4 mos after the 1st ope.</td>
</tr>
</tbody>
</table>

*Not revealed on the 1st angiogram. **Not revealed on the 1st angiogram due to incomplete angiography. ***Persistent arterial flow on Doppler sonography after surgical obliteration (SO) of the fistula. F: female, M: male, ope.: operation.

Although the process of locating SDAVFs using spinal angiography is often tedious, complete angiography is the gold standard to diagnose and localize the arteriovenous shunt. The technique requires precise catheterization of all intersegmental arteries from the craniocervical junction to the sacrum. Angiographic investigation of the fistula can be important from the venous drainage point of view. In the present case, the concurrent PMAVF was not clearly detected on initial angiography because of incomplete demonstration of all segmental arteries. In clinical practice, many neuroradiologists probably stop injecting additional arteries once they have discovered the AVF. However, even after a fistula has been located, selective angiography should be completed until all segmental arteries are demonstrated to be clear of concurrent AVF. The artery of Adamkiewicz should also be visible, as absence may indicate the presence of SDAVF-induced venous hypertension. In one case, an SDAVF at T9 on the right was first detected by spinal angiography. Following surgical ligation of the fistula, the patient showed initial improvement, only to deteriorate again 2 months later. On repeat angiography, a spinal PMAVF was revealed, which was missed on the initial angiography despite the injection of the intersegmental artery. The presence of a second fistula was suspected due to decreased flow in the common draining veins after the first fistula was obliterated. The dural fistula may have obscured the other fistula because of its higher shunt volume. 

The treatment goal is complete obliteration of the fistula, and the results of surgical treatment of SDAVFs are generally good. Surgery resulted in complete exclu-
sion of the fistula at the first attempt in 146 (95%) of 154 patients, and 141 (96.6%) of 146 patients experienced improvement (120 patients, 82.2%) or stability (21 patients, 14.4%) of motor function at last follow up compared with the preoperative status. Only 6% of patients showed subjective or objective worsening of preoperative signs and symptoms at the time of discharge, which persisted at follow up. Therefore, if the patient’s neurological status fails to improve or deteriorates despite treatment, further investigation should be carried out. Reopening of the treated SDAVF, as well as the possible existence of a second fistula, should be considered in cases of incomplete angiography, and persistent clinical and MR findings. The presence of double fistulas remained undetected on angiograms in 2 of the 4 reported patients, who showed transient postoperative improvement followed by delayed deterioration. This observation suggests that decrease of flow in the common draining veins occurs after the first fistula is closed. However, the shunt volume of the second fistula could open up due to improved venous drainage. Following closure of the second fistula, the patients showed improvement, except for one in whom two fistulas were closed by single-stage surgery. These outcomes indicate that ligation of one fistula may not be sufficient to improve clinical symptoms.

The present case indicates that the characteristic signs of SDAVF associated with PMAVF are as follows: differences in the extent of venous drainage on MR images and spinal angiograms, persistent arterial signals in the venous structures seen on Doppler ultrasonography performed after surgical obliteration of the fistula, persistent flow voids seen on postoperative MR images, and recurrence of clinical symptoms after surgical obliteration. Repeat complete angiography should be performed if a concurrent AVF is suspected.

References


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Neurol Med Chir (Tokyo) 52, July, 2012