Spinal Epidural Arteriovenous Fistula: a Unique Pathway into the Perimedullary Vein

A Case Report

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Summary

Spinal dural arteriovenous fistulas (AVFs) are rare vascular malformations in the spinal dura, fed by dural branches of the radicular arteries, and drain primarily into intradural venous plexuses. They may cause elevated medullary venous pressure and produce a progressive myelopathy.

We describe a case of AVF in the epidural space of the previous surgery site of L3 and it showed a unique complex venous pathway into the perimedullary vein, leading to classic clinical symptoms of venous congestion in the spinal cord. The shunt was draining into bilateral epidural venous plexus and then to the paravertebral veins at the level of L2. The venous outflow entered to the epidural space again and finally refluxed into the intradural perimedullary vein.

Introduction

The literature on spinal vascular malformations contains a lot of confusing terminology and classification. Oldfield et al¹ classified spinal vascular malformations into 4 types. However they did not represent spinal arteriovenous malformations (AVMs) and AVFs confined to the epidural space in their classification. Spetzler et al² proposed a modified classification of spinal cord vascular lesions to in-

clude extradural variants. Lesions are divided into three broad primary categories, neoplasms, aneurysms, and arteriovenous lesions. The third category, spinal cord arteriovenous lesions are divided into AVMs and AVFs. The fistulae are classified according to their location as extradural or intradural, and the intradural fistulae are subdivided into dorsal and ventral types. Geibprasert et al³ reported a new classification of dural AVFs according to craniospinal epidural venous anatomical bases: most of the spinal epidural AVFs belong to the ventral epidural group and rest of them are in the dorsal epidural group.

Recently, several literature reports have described a special group of AVFs in the epidural space⁴⁻⁸. They are rare vascular malformations in the epidural space and may cause elevated medullary venous pressure causing progressive myelopathy⁹. The angiographic findings of venous drainage into perimedullary vein of our case were unique and complex.

Case Report

A 65-year-old man presented with a one day history of painful paresthesias on his feet and difficulty walking. On examination we observed almost paralysis of his feet. Sensory examination showed almost absent vibration at toes, reduced at ankles, and mildly reduced at knees. He had lost control of urinary sphincter

functions on admission. There had been no motor and sensory change of the upper extremities.

He had a history of L2-4 laminectomy and discectomy of L2-3, L3-4 for back pain four years before presentation and coronary artery bypass graft eight months before.

On MR imaging, spinal cord revealed high signal intensity at the lower portion and serpentine perimedullary signal voids presented on T2-weighted images (Figure 1) and enhanced on contrast-enhanced T1-weighted images, suggesting congestive myelopathy by spinal vascular malformation.

Spinal angiography was performed from T3 to L4 bilaterally and both internal iliac arteries. On spinal angiogram, there was an AV shunting at the level of L3 lumbar vertebral body on the right side. This region was the site of previous laminectomy. AV shunting was fed by the right anterior segmental artery of L3 lumbar artery. The venous drainage was to epidural vein at the level of L2-3 on the right side, this epidural vein was going up and across the midline and drained to both paravertebral veins at L2 level. These veins were going down to segmental lumbar veins of L3 level. These lumbar veins entered back into the epidural venous plexus of L3 level and eventually drained into the perimedullary vein at the level of L3 on the left (Figure 2A-C).

The L3 feeder and proximal portion of the draining vein were embolized with n-butylcyanoacrylate mixture (Figure 2D).

Following the procedure, the patient's neurological deficits were improved. Paresthesia of the lower extremities and gait instability were resolved completely. The urinary sphincter function was slowly recovered. Follow-up MR images revealed improved spinal cord edema and serpentine signal voids around spinal cord on T2-weighted images disappeared.

Discussion

Spinal epidural AVFs are rare, and only a few case reports have been published in the literature^{2,4-8,15}. The pathophysiology remains unsettled, but these lesions are likely acquired. Some authors have described an association with neurofibromatosis, specifically in the cervical levels, presenting as vertebrovertebral fistulas¹⁰⁻¹². Some reports questioned the relationship with previous surgery or trauma⁷. In our

case, the patient had a history of spinal stenosis which had been treated by L2-4 laminectomy and discectomy four years prior to presenting with myelopathy. Surgical interruption of epidural venous drainage may predispose to venous thrombosis and subsequent abnormal recanalization. How an AVF develops is still unresolved, similar to the intracranial acquired dural AVMs. Compromised outflow of the epidural venous plexus subsequently induces reflux to the intradural veins and eventually to the perimedullary veins. The clinical presentations are the same as classical spinal dural AVFs, resulting in congestive myelopathy.

Lasjaunias et al¹³ recently described a new classification of spinal arteriovenous shunts according to the location of the fistulas. The classical spinal dural fistulas are categorized as "lateral epidural fistulas". Those shunts are lo-

Figure 1 MRI T2-weighted image reveals high signal intensity in lower portion of spinal cord with serpentine signal voids at perimedullary region (arrows) suggestive of congestive myelopathy.

Figure 2 A-C) Superselective angiograms of right L3 lumbar artery demonstrate a AV shunting (arrow) in the epidural space fed by a right anterior intraspinal artery. The venous drainage is to the epidural vein at the level of L3 on the right side. This epidural vein is draining up to L2 level and to both paravertebral veins (were partially cut off, small arrows) at L2 level, then drains back to the epidural venous plexus at L3 level. Note later visualization of the intradural vein (arrows). D) Radiograph shows n-butylcyanoacrylate cast within the epidural AVF located in the superior margin of previous laminectomy.

cated in either ventral or dorsal epidural space and named accordingly. The ventral epidural AVFs are the ones with retrograde venous drainage to the intradural veins when communicating channels of epidural to paravertebral venous plexuses are interrupted, such as our case. The fistulas are supplied by epidural

branches of segmental arteries and their initial venous components are epidural venous plexuses. The symptoms of epidural AVFs are related to the pattern of venous drainage. If they are exclusively epidural, they may present with local pain or radicular pain. But if there is retrograde drainage from the epidural veins into the intradural radicular veins, the patients present with progressive myelopathy as in cases of classical, so-called lateral epidural AVFs. Our case presented with symptoms of lower leg paraparesis, loss of sphincter functions and rather rapidly progressing myelopathy. There have been reports about a reflux impeding mechanism from epidural veins to the intradural venous system, and substantial narrowing and bending of the vessel at the transdural course appropriate to act as an antireflux device¹⁴. However some cases of spinal epidural AVFs with an insufficiency of the reflux-impeding mechanism showed venous reflux into the perimedullary vein, resulting in congestive myelopathy.

Krings et al¹⁵ reported the spinal epidural AVF at the level of L3 with perimedullary drainage, without any history of trauma or surgery in this patient. In their case, the pattern of venous flow was descending, crossing the midline, and ascending on the other side L3 level and retrograde drainage from the epidural venous plexus into a radicular vein. They reported it is interesting to note that the epidural shunt was more extensive, crossing the midline and filling multiple compartments of the plexus before retrogradely entering the radicular vein. In our case, venous flow was unique. From the point of the shunt at the level of L3, it drained to the L2 level and then connected to the segmental paravertebral vein of L2 level bilaterally. These L2 veins drained to the L3 segmental veins. Interestingly, L3 segmental veins drained back into the ventral epidural space. They eventually connected to the L3 radicular vein on the left side.

We assume this complex venous flow into perimedullary vein in our case was due to previous surgery at this level that may have injured the epidural connective tissue involving epidural veins, predisposing to venous thrombosis thus leading to an AVF and complex retrograde venous drainage into the perimedullary vein.

Any AVFs with progressive myelopathy have to be treated as soon as diagnoses are

made. Surgical disconnection of the intradural radicular vein is the curative method, so is endovascular occlusion of the fistula and proximal part of its venous drainage with liquid embolic material. It is essential to advance embolic material to the venous side to prevent recanalization. For the epidural AVF, it is not necessary to penetrate to the radicular vein. Occlusion of the initial part of the epidural venous

plexus is sufficient to cure the shunt as in our case¹⁶.

Our case illustrates that the spinal epidural AVFs can have delayed complex retrograde venous flow into the perimedullary vein. It is important to follow venous drainage long enough to document opacification of any intradural vein, thus make a correct diagnosis and treat such devastating symptoms.

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