Spinal epidural arteriovenous fistulas associated with progressive myelopathy

Report of four cases

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The authors report the cases of four patients who presented with progressive myelopathy (one patient had been asymptomatic for 25 years) due to spinal epidural arteriovenous fistulas (AVFs). Clinical symptoms and magnetic resonance imaging findings were similar to those of dural AVFs. In contrast to dural AVFs, angiography showed that the lesions were fed by multiple vessels and drained in one case in multiple veins. Perimedullary venous drainage was visible in three of the four cases. All fistulas were cured by embolization; arterial access was used in two cases and venous in two. The authors’ aim in this paper is to emphasize the differences between dural and epidural AVFs in terms of their physiopathology and angioarchitecture as well as the therapeutic strategy.

KEY WORDS • epidural arteriovenous fistula • embolization • venous occlusion

There are many reports about spinal dural AVFs with perimedullary venous drainage. These fistulas provide a connection between the medullary draining veins and arteries and lead to progressive myelopathy. In addition, the thrombosis of radiculomeningeal veins that is likely the origin of these fistulas contributes to the decrease in medullary venous outflow and the increase in venous hypertension. Spinal epidural AVFs are rare vascular malformations in the epidural space, are fed by dural or epidural branches, and drain primarily into epidural venous plexuses. They may cause elevated medullary venous pressure and produce a progressive myelopathy.1-11

We present four cases of spinal epidural AVFs causing progressive myelopathy and we compare the physiopathology, angioarchitecture, and therapeutic strategy with those of spinal dural AVFs.

Case Reports

Case 1

This 60-year-old woman with a history of T-12 fracture and T11-L1 laminectomy presented with symptoms of progressive myelopathy that had lasted 1 year. Spinal MR imaging showed a hyperintensity and swelling of the conus medullaris with abnormal, slightly dilated vessels posterior to the spine typical of a spinal dural AVF with perimedullary venous drainage (Fig. 1A). Spinal angiography revealed an epidural AVF fed by the right and left T-12 portions of the lumbar artery draining in an epidural vein, with reflux into the left T-12 radiculomeningeal vein and then into the posterior perimedullary veins (Fig. 1B and C).

Selective catheterization of the left T-12 artery was performed, and a 0.4-ml mixture of Glubran and Lipiodol (25% dilution) was injected (Fig. 1D and E). Angiograms and MR images obtained 3 months later revealed normal findings, but the lower-limb deficit and dysesthesia persisted at the 10-month follow-up clinical examination.

Case 2

This 68-year-old man presented with progressive myelopathy that had lasted 10 months. Three months before presentation, he had undergone a prostatectomy. Spinal MR imaging revealed signs typical of a spinal dural AVF with perimedullary venous drainage (Fig. 2A). Spinal angiography revealed an epidural AVF fed by the left and right L-2 arteries that drained into one right epidural vein and then into the posterior perimedullary veins (Fig. 2B and C).

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Case 3

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Selective catheterization of the left T-12 artery was performed, and a 0.4-ml mixture of Glubran and Lipiodol (25% dilution) was injected (Fig. 1D and E). Angiograms and MR images obtained 3 months later revealed normal findings, but the lower-limb deficit and dysesthesia persisted at the 10-month follow-up clinical examination.
Lipiodol (dilution of 25%) with passage of the glue into the origin of the draining vein. The patient experienced a transient clinical improvement, but his symptoms worsened after 2 months. The MR images obtained 3 months later showed persistent signal abnormalities.

Spinal angiography revealed the persistence of a fistula draining into one of the three previously visible veins (Fig. 2E), and a second embolization was performed with catheterization of the right L-3 artery and a 0.4-ml injection of Glubran and Lipiodol (dilution of 25%), but unfortunately the glue did not reach the origin of the vein. Clinical improvement was incomplete, and 3 months later the MR images again revealed abnormal findings. Another angiography study was performed and revealed a persistent epidural fistula that was fed by the right L-2 artery (Fig. 2F). Selective catheterization of this pedicle allowed a 1.1-ml injection of Onyx (Microtherapeutics Inc.) over 15 minutes, and the Onyx was placed into the first 2 cm of the draining vein. Three months later the patient had recovered completely. Findings on spinal angiograms and MR images were normal.

Case 3

This 69-year-old man presented after experiencing progressive myelopathy for 1 month. Spinal MR images (not shown) were obtained, which revealed typical signs of dural AVF with perimedullary venous drainage. Angiography showed an epidural AVF that was fed by the right and left portions of the T-12 artery and drained into the azygos vein (Fig. 3A). Reflux into the right T-11 radiculomeningeal and perimedullary veins was visible (Fig. 3B). The ASA arose from the left L-1. Embolization was initially undertaken by catheterization of the right T-12 artery and a 1.2-ml injection of Onyx over a period of 30 minutes; a small volume of Onyx reached the venous side. Angiography of the right and left T-12 arteries showed the persistence of the slow-flow fistula. At 1 month, there was no clinical improvement and the MR imaging findings were still abnormal. Angiography showed the persistence of the fistula arising from the left (Fig. 3C) and right T-12 and right L-1 arteries. Embolization was then performed through a venous approach. Retrograde catheterization of the azygos vein with a 6-F catheter was done but was extremely difficult due to the presence of the valves. A microcatheter was then introduced within the venous pouch (Fig. 3D). Seven coils were finally placed, and the fistula was completely occluded (Fig. 3E). At 3 months the patient had completely recovered. Findings on spinal MR imaging were normal.
Case 4

This 75-year-old woman presented with an epidural fistula that had been diagnosed 25 years earlier during a cardiology consultation concerning a lumbar pulsatile bruit. This diagnosis was confirmed by angiography. No reflux into the medullary veins was visible. Because the patient was asymptomatic, no treatment was undertaken. Six months before the present admission, she experienced progressive myelopathy. Angiography showed an epidural fistula fed by the right T-11, T-12, and L-1 (Fig. 4A) and left T-12 and L-1 arteries, draining into the azygos vein as well as the perimedullary venous system (Fig. 4B). The ASA arose from the left L-1 artery, which also fed the fistula. Because of this, arterial embolization was considered too dangerous.

Retrograde venous catheterization was then considered. However, catheterization of the azygos vein was again extremely difficult and microcatheter navigation failed to reach the origin of the draining vein.

One month later, another attempt was undertaken in which we performed arterial embolization by catheterization of the right L-1 artery and injected 3.6 ml of Onyx over a period of 57 minutes (Fig. 4C). Unfortunately, the Onyx did not reach the origin of the draining vein, and the injection was stopped because of the risk of retrograde migration of Onyx into the left L-1 spinal artery. We then decided to attempt a venous approach and finally were able to reach and occlude the vein with coils (Fig. 4D). Two months later, the patient had completely recovered.

Discussion

Spinal dural AVFs with perimedullary venous drainage correspond to a single microscopic transdural fistula fed by radiculomeningeal arteries that drains into an intradural vein. The fistula is embedded in the dura covering the nerve root as well as in the adjacent dura mater. It drains in a retrograde way through the medullary veins to the co-
nal venous plexus, which becomes dilated, tortuous, and elongated. Increased venous pressure is responsible for progressive myelopathy. The main site of interference with cord function due to high-pressure blood in the coronal venous plexus is in the region of the spine (thoracic, lumbar, or sacral) where the fistula is located. On T2-weighted MR images the conus medullaris appears swollen and hyperintense due to venous congestion and serpentine linear structures around the surface of the cord corresponding to dilated perimedullary veins.

Epidural AVFs are much rarer, and only a few case reports have been published in the literature. Their physiopathology is still not clear, but these lesions are likely always acquired. Some authors have described an association with neurofibromatosis and previous surgery or trauma. In our four patients, only one suffered a prior T-12 fracture that had been treated with a T11–L1 laminectomy. The fistula developed 1 year later at the level of T-12. The patients in the three other cases had no remarkable medical history, and no patient had suffered back pain before the myelopathy occurred.

In spinal epidural AVFs, the fistula is located in one of the epidural venous compartments. It is fed by dural or epidural branches and drains primarily into epidural venous plexuses. In our four cases, the fistula was fed by two arteries in two cases, and three and five arteries in the remaining two cases. The numerous osseous and epidural branches of the intersegmental arteries as well as anastomosis with upper or lower segments or contralateral arteries explain why these fistulas, located in the epidural space, are frequently fed by multiple arteries. This differs from dural AVFs.

The clinical characteristics of epidural AVFs depend on the pattern of venous drainage. If they are exclusively epidural, they may present with pulsatile bruit (as in Case 4) or with chronic local pain or compressive radiculopathy or myelopathy. In cases in which there is retrograde drainage from the epidural veins into a radiculospinal vein and then to the medullary veins, the patients may present with progressive myelopathy as in cases of dural AVFs.

Of our four patients, three presented initially with progressive myelopathy. In the fourth patient the AVF was revealed by a pulsatile bruit detected in the lumbar region. In this patient, the spinal angiograms showed no medullary drainage, and she was followed up for 25 years. When she suffered myelopathy, the angiogram showed the medullary drainage that was not initially present. In one of the four cases, the reflux within the medullary vein was not visible but the injection into the ASA showed no opacification of venous drainage in the spinal cord after more than 20 sec-

![Fig. 3. Case 3. Spinal angiograms. Images showing an epidural AVF fed by right (A) and left T-12 arteries draining into the azygos vein (single arrow in B) with reflux into the perimedullary veins (double arrows in B). Onyx embolization through the right T-12 artery created insufficient passage into the origin of the draining vein, causing persistence of the fistula. Image obtained at the 1-month follow up (C) confirming that the residual fistula originated from the left and right T-12 and right L-1 arteries. Retrograde access through the azygos vein (D) was possible. Complete occlusion was attained after coil embolization (arrows in E).](image)
The congestion of the medullary vein even without any retrograde drainage could explain the absence of visualization of the medullary drainage, clinical symptoms, and MR imaging signs.

Concerning MR imaging, our four patients presented with signs typical of a spinal AVF with swelling and hyperintensity of the spinal cord and conus medullaris on T₂-weighted imaging sequences. In addition, serpentine

Fig. 4. Case 4. Spinal angiograms. Images showing an epidural fistula fed by the right T-11, T-12, L-1, and left T-12 and L-1 arteries draining into the azygos vein (arrow in A) with reflux into the perimedullary venous system (arrows in B). Onyx embolization failed to occlude the vein (arrows in C). Retrograde catheterization of the azygos vein was performed. Complete occlusion was obtained after coil insertion at the origin of the draining vein (arrows in D).
Spinal epidural arteriovenous fistulas

curvilinear structures around the surface of the cord and meningeal contrast enhancement were seen. Even retrospectively nothing could differentiate these epidural AVFs from dural AVFs.

Treatment of dural AVFs consists of occluding the origin of the draining vein by using either embolization or direct surgery. Embolization may fail when the feeding artery is small and does not allow distal catheterization or efficient injection of the embolization agent at the origin of the vein. Failure to reach the vein always produces small arterioles and persistence of a fistula that requires surgery.

We attempted an arterial approach in our four patients, but it succeeded in only two. Additionally three arterial embolization procedures were required in one patient (Case 2) to obtain complete closure of the fistula. The use of Onyx after two glue embolization procedures had failed allowed a much longer injection time and complete filling of the draining vein.

Onyx is a new nonadhesive agent. A nonadhesive liquid decreases the risk of the catheter being glued to anatomical structures and allows a more durable injection of a much larger amount of agent delivered in a single injection. Nevertheless, the use of Onyx in Cases 3 and 4 failed to attain a complete occlusion of the fistula because the draining vein could not be completely occluded. The numerous feeding vessels and their small size made occlusion of the vein extremely difficult. Additionally, all four fistulas were located at the thoracolumbar junction. The presence of the ASA of the conus medullaris rendered the arterial injection dangerous and it did not allow a very long injection time of Onyx due to the risk of migration of the agent by anastomosis into the origin of the spinal artery.

Unlike a spinal dural AVF, an epidural spinal AVF may be treated in the same manner as a dural cranial fistula through a venous access. Venous access is less dangerous than arterial access when considering the risk of spinal artery occlusion. However, transvenous catheterization is difficult. Placement of a guiding catheter in the azygous vein may be tricky due to the presence of the valves. Moreover, microcatheter navigation across to the venous side of the fistula is difficult due to the tortuosity of the small meningeal connections within the paraspinal longitudinal veins. In the two cases (Cases 3 and 4) in which venous access was attempted, two attempts were necessary in one before the origin of the draining vein was reached. Finally, similar to the treatment of intracranial dural AVFs, we decided to place coils, which allowed immediate and definitive occlusion of the fistula. Three of our patients completely recovered clinically even though the fistulas were treated 1 year after they had been diagnosed.

We did not perform surgery in our cases, but it is an efficient therapeutic option for treating these types of fistulas. Krings and colleagues reported on a patient with progressive myelopathy and a spinal epidural AVF that drained into the epidural plexus and the perimedullary veins (similar to our Case 3). Direct surgery was performed, and both the epidural fistula zone and the radicular vein were coagulated. A complete obliteration of the fistula was confirmed on angiography. In only one report a combined endovascular and surgical approach of such fistulas was described; Pirouzmand et al. used arterial glue injection followed by direct surgery to treat a sacral epidural AVF fed by the sacral lateral artery that drained into the epidural venous plexus. Thus, surgery may be considered as a therapeutic option for epidural AVFs with a complex angioarchitecture.

Based on our four cases and our review of the literature, the main differences between dural and epidural AVFs in terms of angioarchitecture, clinical presentation, MR imaging signs, and treatment are summarized in Table 1.

Conclusions

Despite our limited number of cases some aspects of epidural spinal AVFs are of interest. These lesions may be associated with clinical symptoms and MR imaging signs similar to dural AVFs. In addition, they can be fed by multiple arteries, and retrograde flow within the perimedullary veins may or may not be visible. Furthermore, arterial embolization is difficult and potentially dangerous because of the numerous feeding vessels and risk of migration of the embolic agent into the ASA if the origin of the vessel is close to the fistula. Finally, venous access to these fistulas is difficult but can effect a complete and definitive occlusion.

Disclosure

Dr. Cognard has a contract with ev3, Inc., of which Microtherapeutics is a division, to train European physicians to use Onyx in animal models and patients.

References


TABLE 1

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<tr>
<th>Characteristic</th>
<th>Symptoms and Clinical Features</th>
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<tr>
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<td>Spinal Dural AVF</td>
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<tr>
<td>symptom</td>
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