Spinal Epidural Arteriovenous Fistulas with Unusual Manifestation of Sudden Onset of Severe Neurological Deficits: Case Report

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Abstract

Spinal epidural arteriovenous fistulas with perimedullary venous drainage cause venous hypertension, and usually manifest as slowly progressive myelopathy. We treated two patients presenting with sudden onset of severe neurological deficits. Moreover, in Case 1, the venous drainage was exclusively epidural and no perimedullary venous drainage was present. Angiographic findings of this patient were characterized by a slow-flow fistula with marked retention of the epidural venous drainage. Rapidly progressing thrombosis of the epidural venous plexus may have caused the sudden onset of the symptoms. In Case 2, hematomyelia may also be possibly associated with the sudden onset of the symptoms. Early diagnosis and treatment are essential to achieve favorable outcome in such cases because venous congestion results in irreversible venous infarction within a short period.

Key words: congestive myelopathy, endovascular therapy, spinal epidural arteriovenous fistula, unusual manifestation

Introduction

Spinal epidural arteriovenous fistulas (AVFs) occur as a single fistula between the radicular artery and vein within the dural sleeve of a nerve root, and drains retrogradely toward the perimedullary vein, resulting in venous hypertension and progressive myelopathy. In contrast, spinal epidural AVFs occur as multiple fistulas within the epidural space fed by the dural branches, and draining primarily into the epidural venous plexus.1,2,5,6,8,9,12,14,16) Spinal epidural AVFs can be classified into two types based on the angiographic findings and clinical presentations: high-flow fistula that mainly causes compressive myelopathy or radiculopathy due to the dilated epidural venous plexus;2,5,8) and slow-flow fistula that is usually asymptomatic because the anti-reflux system in the radicular vein within the dural sleeves hinders retrograde flow toward the perimedullary veins from the epidural veins.9) However, if perimedullary venous drainage is present in some pathological circumstances, congestive myelopathy as well as spinal dural AVFs can occur.1,6,8,12,14,16)

Case Reports

Case 1: A 24-year-old man presented with sudden onset of complete tetraparesis. He had a 6-month history of intermittent nuchal pain after neck injury caused by a fall. T1- and T2-weighted magnetic resonance (MR) imaging taken 4 hours after the onset revealed no abnormalities. His symptoms persisted and he was referred to the neurology department in our hospital 4 days after the onset. Neurological examination showed complete tetraparesis, sensory disturbance, and bladder-bowel dysfunction. T2-weighted MR imaging 4 days after the onset showed an intramedullary high intensity lesion in the spinal cord at the C2-C5 levels and in the C5 vertebral body. Abnormal signal voids were not recognized (Fig. 1A). MR imaging with contrast medium showed no enhanced lesions (Fig. 1B). Steroid pulse therapy was performed, but was not effective, so he was referred for neurosurgery. T2-weighted MR imaging 12 days after the onset revealed...

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Fig. 1  Case 1. A: T2-weighted MR image 4 days after the onset showing high intensity lesions in the C3-C5 spinal cord with surrounding edema and in the C5 vertebral body (asterisk). No abnormal flow voids are recognized. B: T2-weighted MR image with contrast medium 5 days after the onset showing no enhanced lesion. C: Right vertebral angiogram 12 days after the onset showing an epidural AVF fed by the right C2 segmental artery (large arrow). Anterior spinal artery (small arrow) and posterior spinal artery (small open arrow) are patent. D: Left vertebral angiogram showing an epidural AVF mainly fed by the left C3 segmental artery (large open arrow). E, F (antero-posterior views, E: arterial phase, F: late venous phase) and G (lateral view): Transarterial embolization of the right C2 segmental artery was performed 12 days after the onset. Angiogram of the right C2 segmental artery showing epidural AVF draining into the right posterior internal vertebral venous plexus and descending to the right C3 with marked retention. Note that no perimedullary venous drainage is present. H: T2-weighted MR image 8 days after the embolization showing disappearance of the surrounding edema and persistent high intensity core. I: Four months later, transarterial embolization of the left C3 segmental artery was performed. Angiogram of the left C3 segmental artery showing the epidural AVF draining into the right posterior internal vertebral venous plexus, descending near to the left C4-5 intervertebral foramen, and ascending and then crossing the midline (not shown) with marked retention. Note that no perimedullary venous drainage is present. Arrowhead indicating the tip of the microcatheter, open arrowheads indicating the draining vein (E-G, I). AVF: arteriovenous fistula, MR: magnetic resonance.

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progression of the spinal cord lesion that consisted of a central high intensity core and surrounding high intensity lesion. Right vertebral angiography 12 days after the onset demonstrated an AVF at the C1-C2 levels in the right posterior epidural space, mainly fed by the right C2 segmental artery, draining into the right posterior internal vertebral venous plexus, and descending to the C3 level of the spinal cord with marked flow retention. The anterior spinal artery (ASA) and posterior spinal artery were patent (Fig. 1C). Left vertebral angiography demonstrated an AVF at the C1-C3 levels in the left posterior epidural space, mainly fed by the left C3 segmental artery, draining into the left posterior internal vertebral venous plexus, ascending and crossing the midline, and descending near to the C4-5 intervertebral foramen (Fig. 1D).

Transarterial embolization of the right C2 segmental artery was performed 12 days after the onset (Fig. 1E-G). A 1.5F Marathon catheter (ev3 Neurovascular, Irvine, California, USA) was navigated into the right C2 segmental artery. Perimedullary venous drainage was not recognized. After coil embolization of the muscle branch, a 33% mixture of N-butyl-2-cyanoacrylate (NBCA) (B. Braun, Melsungen, Germany) and lipiodol was injected, and the shunt flow significantly decreased. After the embolization, the tetraparesis gradually improved on the left, but bladder-bowel dysfunction did not recover. T1-weighted MR imaging 8 days after the embolization revealed disappearance of the surrounding edema, but the high intensity core persisted (Fig. 1H). Four months later, transarterial embolization of the left C3 segmental artery was performed using NBCA because of the persistence of the AVF on follow-up angiography (Fig. 1).

**Case 2:** A 55-year-old man presented with sudden onset of right nuchal pain followed by right complete hemiplegia. T1-weighted MR imaging 2 days after the onset revealed intramedullary high intensity lesion in the spinal cord at the C1-C7 levels with low intensity lesion. Abnormal signal voids were not recognized (Fig. 2A). MR imaging with contrast medium showed no enhanced lesions. Right vertebral angiography demonstrated an AVF at the C3-C4 levels in the right anterior epidural space, mainly fed by the right C4 segmental artery, and draining into the right anterior internal vertebral venous plexus with perimedullary venous drainage (Fig. 2B). Left vertebral angiography demonstrated an AVF at the C3-C4 levels in the left anterior epidural space, mainly fed by the left C4 segmental artery, and draining into the left posterior internal vertebral venous plexus.

Transarterial embolization of the right C4 segmental artery was performed 11 days after the onset (Fig. 2C-E). A 1.2F Magic catheter was navigated into the right C4 segmental artery. Right C4 segmental angiography revealed an epidural AVF which drained superiorly into the ASV. However, this segmental artery also supplied blood flow to the ASA, so we abandoned transarterial embolization. T2-weighted MR image taken 24 days after the onset showed high intensity lesion coinciding with the low intensity lesion on the T1-weighted imaging (Fig. 2F). T2* MR imaging at the C4 level showed a low intensity lesion indicating hemorrhage (Fig. 2G).

Fifty days after the onset, surgical disconnection of the intradural draining veins was performed. Right C2-C4 hemi-laminectomy was performed. The intradural draining veins ascending along the right C4 ventral nerve roots and the radiculomedullary artery connecting with the ASA were identified (Fig. 2H), and the draining veins were coagulated. The epidural AVF and draining veins were left untouched. Postoperative angiography showed disappearance of the perimedullary venous drainage and faint residual fistula of the epidural space (Fig. 2I). T2-weighted MR imaging 1 month after the operation revealed marked decrease of the spinal cord edema (Fig. 1J). After the operation, the right hemiparesis gradually improved and he could walk with a cane one month later, but the paresis of the upper limb did not improve.

**Discussion**

The present two cases were characterized by sudden manifestation of neurological deficits mimicking spinal cord infarction, whereas spinal epidural AVFs usually cause slowly progressive myelopathy. A previous case of epidural AVF with acute paraplegia may have involved thrombosis of the epidural venous plexus with incomplete recanalization and some shunts due to insufficient collaterals, leading to increased venous pressure and reflux into the intradural medullary veins.14)

In our Case 1, the venous drainage was exclusively epidural and no perimedullary venous drainage was present. Angiographic findings of this patient were characterized by a slow-flow fistula with marked retention of the epidural venous drainage. We speculate that the increased epidural venous pressure caused by the thrombosis, as evidenced by the marked retention of the draining veins, interfered with the venous outflow in the spinal cord, resulting in venous congestion despite the absence of perimedullary venous drainage. The rapidly progressing venous thrombosis apparently caused the sudden onset of symptoms, and the antecedent nuchal pain was a sign of thrombosis of the epidural venous plexus. In our Case 2, hematomyelia demonstrated by high intensity on T1-weighted and low intensity on T2*-weighted MR imaging was also possibly associated with the sudden onset of hemiparesis.

In our Case 1, T1- and T2-weighted MR imaging showed a high intensity lesion in the vertebral body. Vertebral body infarction is known as a confirmatory sign of spinal
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cord ischemia. If the radiculomedullary artery supplying the ASA and the anterior and posterior central branches supplying the vertebral body originate from the same segmental artery, spinal cord infarction and vertebral body infarction can occur simultaneously. Cheng et al. reported seven of twenty patients diagnosed as spinal cord infarction had concomitant vertebral body infarctions, and four of them had aortic diseases. Most of the vertebral body infarctions were seen in the thoracolumbar regions, which are territories of Adamkiewicz artery, and were adjacent to their cord lesions. Therefore, our initial diagnosis of Case 1 was spinal cord infarction based on the absence of perimedullary venous drainage and the presence of vertebral body infarction. However, venous hypertension or congestion of the vertebral venous plexus can also cause venous infarction of the spinal cord and the vertebral body as a result of venous return disorder in the radicular veins from the spinal cord and the basivertebral vein from the vertebral body. No case of venous infarction of the vertebral body resulting from spinal dural or epidural AVFs has been reported. We speculate that the cause of the high intensity lesions in both the spinal cord and vertebral body was venous congestion, because patency of the ASA was confirmed by spinal angiography, and the improvement of the symptoms coincided with the disappearance of the surrounding edema after embolization.

Dural AVFs initially cause venous hypertension and congestion, and result in venous infarction according to the progression of the lesion. Venous hypertension and congestion result in vasogenic edema and are reversible, whereas venous infarction results in cytotoxic edema and

Fig. 2 Case 2. A: T2-weighted MR image 2 days after the onset showing marked spinal cord swelling and high intensity lesions in the C1-C7 spinal cord with low intensity lesions. No abnormal flow voids are recognized. B: Right vertebral angiogram 11 days after the onset showing an epidural AVF fed by the right C4 segmental artery draining into the right anterior internal vertebral venous plexus with perimedullary venous drainage. C-E: Serial angiograms of the right C4 segmental artery showing early filling of the anterior spinal artery (arrowheads) supplied by the radiculomedullary artery (C), and then an epidural AVF draining into the right anterior epidural venous plexus and ascending to the anterior spinal vein (D, E: open arrowheads). Note the diamond shape of the anterior spinal artery (D: dotted circle) and a connection with the epidural veins (E: arrow) and perimedullary veins of the cranio-cervical junction (E: open arrow). F: T1-weighted MR image 24 days after the onset showing high intensity lesion coinciding with the low intensity lesion on the T2-weighted image. AVF: arteriovenous fistula, MR: magnetic resonance.
is irreversible. Follow-up MR imaging of the present two cases showed that the spinal cord became atrophic and the core of the high intensity lesion persisted. These areas seem to have already suffered irreversible venous infarction within a short period. Early diagnosis and treatment are essential to achieve favorable outcome for patients with rapid progression of neurological symptoms.

Occlusion of the origin of the draining vein using either embolization or direct surgery can result in complete cure of spinal dural AVFs. However, spinal epidural AVFs commonly have multiple feeding arteries, fistulas, and draining veins, so complete obliteration by transarterial embolization using NBCA seems to be difficult because NBCA hardly penetrates into the large venous lake in the epidural space. Surgical interventions combined with or without embolization may be required in most cases. Recently, embolization of spinal epidural AVFs using Onyx has been reported. Onyx is a non-adhesive liquid embolic material, and can penetrate the large venous lake and perimedullary vein, resulting in high cure rate. The ASA is sometimes difficult to differentiate from the ASV in cases of spinal epidural AVFs draining to the ASV, as in our Case 2. The ASA is identified by earlier filling than the ASV, a typical diamond shaped, and connections with the other segmental arteries. In contrast, the ASV is identified by connections with the other medullary veins or epidural veins, and deeper location than the ASA in the lateral projection. Precise analysis of the angiographic findings is necessary to avoid complications associated with the embolization.

**Conclusion**

Two unusual cases of spinal epidural AVFs manifested as sudden onset of severe neurological deficits. Rapidly progressing venous thrombosis of the epidural venous plexus apparently caused the sudden onset of symptoms. Early diagnosis and treatment are essential to achieve favorable outcome in such cases because venous congestion results in irreversible venous infarction within a short period.
References


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