Spinal intraosseous epidural arteriovenous fistula with perimedullary drainage obliterated with Onyx embolization: case report

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The authors report an extremely rare case of spinal intraosseous epidural arteriovenous fistula (AVF) with perimedullary vein reflux causing symptoms of myelopathy. The intraosseous fistula tracts were completely obliterated with Onyx embolic agent, resulting in a total resolution of symptoms. The unique features of this case include the rare location of the fistula in the vertebral body and the association of the fistula with a compressive fracture. Imaging studies confirmed these hemodynamic findings and provided clarity and direct evidence regarding the association of epidural AVF formation with the vertebral compressive fracture. The authors also propose a possible disease evolution based on the previously adduced reflux-impending mechanism.

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A SPINAL dural arteriovenous fistula (AVF) is the most common spinal vascular lesion, accounting for 70% of all arteriovenous shunts of the spine.^{7,10} In very rare situations, the fistula tract occurs extradurally between the radicular artery branches and the epidural venous plexus in a condition referred to as a “spinal epidural arteriovenous fistula.” This lesion is very rare, and only a limited number of cases have been reported in the literature to date.^{1–4,8,13,15,18} Here we describe an extremely rare case of intraosseous spinal epidural AVF with perimedullary vein reflux causing symptoms of myelopathy. The intraosseous fistula was located in a fractured vertebral body and was totally obliterated using Onyx embolization. We present the unique imaging features of the intraosseous communications, clarify the relationship between the traumatic compressive fracture and the extradural AVF formation, and propose our pathophysiological hypothesis for such rare cases.

Case Report

History and Examination

A 57-year-old man who performed heavy labor presented with a history of T-6, T-9, T-12, and L-2 compression fractures due to occupational injuries sustained before 2005 and mild low-back pain lasting for more than 1 year. After accidentally falling from a tree in 2009, the patient experienced acute deterioration in his symptoms of low-back pain with paresthesia. His Medical Research Council Scale score for muscle strength was Grade 3 for the bilateral iliopsoas muscles, Grade 4 for the quadriceps muscles, Grade 2 for the hamstring muscles, and Grade 2 for the dorsiflexor muscles. The knee and ankle jerks were more brisk than the tendon jerks in the arms, and the plantar responses were positive. Urine retention and feces incontinence were also noted. Spinal T2-weighted MRI revealed hyperintensity and edema of the spinal cord with

ABBREVIATION AVF = arteriovenous fistula; LES = liquid embolic system.

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numerous abnormal dilated vessels indicating spinal dural AVF with perimedullary venous drainage (Fig. 1A and B). Spinal angiography showed an epidural AVF fed by the left T-12 intercostal artery with intraosseous fistula tracts directly connecting to the basivertebral vein and draining to the anterior internal (epidural) vertebral venous plexus with reflux into the perimedullary (intradural-extradural) veins (Fig. 1C and D).

Treatment

Embolization was performed via selective catheterization from the left T-12 intercostal artery into the fistula, and 0.6 ml of Onyx LES (liquid embolic system, Covidien Ltd.) was injected over a 12-minute period. The proximal parts of the venous receptacle and each intraosseous fistula tract were totally occluded in one session, and postembolization angiography showed complete obliteration of the AVF (Fig. 2A and B).

Posttreatment Course

The patient's symptoms resolved the day after embolization, and he was able to walk with aid. Follow-up MRI 5 days later revealed the complete disappearance of abnormal cord signal intensity and the resolution of dilated vessels. Multidetector CT with multiplanar reconstruction clearly confirmed the intraosseous communication of the AVF (Fig. 3). The patient was discharged 1 week later, and rehabilitation of the bilateral lower extremities was arranged. Three- and 6-month clinical follow-ups with MRI (Fig. 2C) and 1-year digital subtraction angiography studies showed complete remission of the neurological symptoms with no abnormality or residual fistula.

Discussion

The pathophysiological mechanism of spinal epidural AVF remains unclear, although causes such as neurofibromatosis, previous surgery, or trauma have been reported. The epidural venous plexus extends as a continuous vertical, thin-walled valveless system from the skull base to the sacrum and drains the intervertebral vein, radicular veins, and spinal veins toward the azygous and hemiazygous venous systems.¹⁷ Tadie et al.¹⁶ described an anti-backflow system within the transdural course of the radicular vein resulting from a substantial narrowing and bending of the vein while crossing the dura mater.¹⁴ The spinal epidural fistula may generally slowly progress and are asymptomatic owing to a sufficient reflux-impending mechanism at the dural sleeves.⁵ However, a patient may present with symptoms of cord compression if there is enlargement of the epidural veins. Uncommonly, if an episode such as break down of the reflux-impending mechanism takes place,^{9,14,16} the shunt flow may drain retrogradely into the perimedullary veins, causing subsequent venous congestion and spinal cord edema, resulting in progressive myelopathy.

We assumed that the patient in our case had existing vertebral compressive fractures and may have had asymptomatic epidural AVF for a period of time given the sufficiency of the reflux-impending system. After his most recent accident, the anti-backflow mechanism may have failed, resulting in drainage into the paraspinal venous system because of thrombophlebitis and/or thrombosis of the outflow veins, and then flowed retrogradely from the internal plexus into the perimedullary veins, resulting in venous congestion and spinal cord edema.

An intraosseous extradural AVF with perimedullary

FIG. 1. Preembolization studies. Sagittal T2-weighted (A) and T1-weighted Gd-enhanced (B) MR images of the thoracolumbar spine reveal numerous dilated perimedullary abnormal vascular structures (arrows), which raise the suspicion of a spinal dural or perimedullary AVF. The high signal intensity within the spinal cord (A) indicates spinal cord edema. Early (C) and late (D) phase, anteroposterior views, with selective angiography at the left 12th thoracic intercostal artery, reveal an extradural AVF (arrowheads) draining into the basivertebral vein, the segmental vein, the intervertebral vein, and the internal plexus with reflux into the tortuous perimedullary veins (arrows).

FIG. 2. Post-Onyx embolization studies. Scout film (A) and selective (B) angiograms (anteroposterior views, obtained at the left 12th thoracic intercostal artery) show total obliteration of the fistula and proximal drainage vein (basivertebral vein). A 6-month follow-up sagittal T2-weighted MR image (C) of the thoracolumbar spine reveals the disappearance of the perimedullary venous structures and normal spinal cord signal intensity.

reflux, as in the present case, is extremely rare, with fewer than 5 cases reported in the literature.^{3,6} The therapeutic goal for spinal epidural AVF is to obliterate the shunt and proximal venous communication while maintaining the patency of the distal parts of the vein and proximal artery by using transarterial or transvenous embolization, direct surgery, or a combination of the two.^{1,9} Surgical treatment is preferred if small feeders prohibit distal catheterization

or if the fistula is close to a major artery such as the anterior spinal artery. We attempted embolization via an arterial approach in our patient using the Onyx LES, which incorporates a new nonadhesive liquid embolic agent that can decrease the risk of gluing the catheter to anatomical structures and allows a more durable injection of greater volumes of agent delivered in a single session. This agent is mainly used for the treatment of cerebral dural fistulas, and some studies have used this agent for spinal dural AVFs, but only 3 reports^{12,13,15} have described the application of this agent for spinal epidural AVFs. A study by Silva et al.¹⁵ documented 2 cases of epidural AVF treated using Onyx embolization, but treatment failed because of incomplete occlusion of the draining vein and the risk of agent migration into the spinal artery via anastomosis. Nogueira et al.¹¹ reported that endovascular treatment with Onyx was feasible, safe, and highly effective, as it allows more controllable penetration of the embolic agent into the fistula and draining vein.

Several interesting clinical points, unlike those in other studies, existed in our case. For example, the fistula in our case had an intraosseous course associated with compressive fracture, and imaging studies confirmed these hemodynamic findings and provided clarity and direct evidence regarding the association between epidural AVF formation and trauma (compressive fracture). Because our patient had incurred a compressive fracture before symptom exacerbation, we hypothesized that the fistula developed in the vertebral body and was fed by the left T-12 intercostal artery connecting to the basivertebral vein. At this stage, symptoms may be caused by mass effect and can be tolerated. After a period of time or following an accident, the reflux-impending mechanism may be destroyed, resulting in retrograde flow into the perimedullary vein, causing medullary venous hypertension and cord edema. In this case, we performed transarterial embolization of the completely occluded intraosseous fistula tracts and proximal drainage veins using Onyx, which incorporates an embolic agent whose use is feasible, safe, and highly

FIG. 3. Post-Onyx embolization studies. Multidetector CT scans with multiplanar reconstruction, axial (left) and sagittal (right) views, clearly confirm an intraosseous epidural fistula in the fractured T-12 vertebrae. Note the linear opacities are Onyx within the fistula and basivertebral vein. Figure is available in color online only.

effective, allowing for good controlled penetration. In addition, it provides excellent contrast for the follow-up CT of embolized fistulas owing to high density effects.

Conclusions

Spinal intraosseous AVF draining into the epidural venous plexus with reflux into the intradural perimedullary veins and causing myelopathy is an extremely rare finding and may be associated with traumatic compressive fracture. Transarterial endovascular surgery is an effective and noninvasive therapeutic method for this vascular disease. Use of the Onyx LES is thought to be feasible and safe, allowing for good controlled penetration to achieve complete obliteration of the fistulas.

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Author Contributions

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