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Case report

Treatment of perimedullary arteriovenous fistula of the spinal cord by superselective neuroendovascular therapy: A case report and literature review

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1. Introduction

Spinal cord arteriovenous malformation (SCAVM) is a rare disease accounting for 2%–5% of lesions within the spinal canal [1]. In general, neurosurgeons and radiologists are eventually consulted for treatment of this disease; however, in most cases, patients initially present to orthopedic surgeons with progressive myelopathy caused by venous hypertension [2,3]. The duration of symptoms before diagnosis is usually long [3], probably due to the rarity of the disease. Moreover, the functional outcomes have been shown to worsen with symptom duration prior to diagnosis [4]. Orthopedic surgeons should therefore consider this rare lesion to make an early diagnosis.

In 1971, Di Chiro et al. proposed the first classification for spinal arteriovenous shunt disease [5]. Since then, many surgeons have attempted to develop new and more useful classifications, and these attempts continue into the present. For example, Qureshi et al. proposed a new and easy-to-use scheme for angiographic classification of SCAVM [6], but no universal classification system has yet been developed for SCAVM. The most widely used classification system at present was created in the 1990s by combining systems from various reports; it distinguishes 4 categories of SCAVM: type I, dural arteriovenous fistula (AVF); type II, intramedullary glomus arteriovenous malformation (AVM); type III, juvenile or combined AVM; and type IV, intradural perimedullary AVF. We reviewed previous reports on SCAVM and show the most general classification and characteristics of each type in Table 1 [2,7–20].

Among SCAVMs, type IV or perimedullary AVFs are especially rare, accounting for only approximately 15%–40% of all SCAVMs [2,21–23]. Perimedullary AVFs are arteriovenous shunts without a nidus of abnormal vessels between the feeding artery and the draining vein. They are located on the surface of the spinal cord (just under the pia) and are fed by the anterior spinal artery (ASA) and/or the posterior spinal arteries (PSAs) [24]. In previous reports, the trend in the male-to-female ratio in perimedullary AVFs was unclear, whereas the age of onset ranged from 20 to 40 years (average approximately 35 years); this age is younger in comparison with dural AVFs [7,9,10,21]. Compared with dural AVFs, perimedullary AVFs are generally more difficult to treat because of their complicated anatomy and physiology [25]. Current treatment options include open microsurgery and endovascular embolization; however, endovascular therapy has gained acceptance as a first-line treatment. Surgical treatment is typically selected when the endovascular approach fails or is nearly impossible [9,10].

Here, we encountered a difficult-to-treat case of perimedullary AVF in the ventral lower thoracic cord. Cure with traditional treatment options (conventional endovascular embolization or open microsurgery) was difficult, but we were eventually able to cure this patient's perimedullary AVF using the new microcatheter-based highly selective embolization technique, which has rapidly progressed in recent years. In this paper, we report our experience for the benefit of other orthopedic spine surgeons.

2. Case report

A 35-year-old woman developed low back pain and left leg pain and numbness. Four weeks after onset, she was unable to stand unassisted and was admitted to a hospital. She had severe bladder dysfunction and muscle weakness in the proximal part of both legs. Magnetic resonance imaging (MRI) showed swelling of the spinal

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Table 1
Traditional classification and characteristics of spinal cord arteriovenous malformations (SCAVMs).

Traditional type	I	II	III	IV
Structural features	Dural AVF	Intramedullary glomus AVM	Juvenile or combined AVM	Intradural perimedullary AVF
Spetzler classification	Intradural dorsal AVF	Intramedullary AVM	Extradural–intradural AVM	Intradural ventral AVF
Subclassification	a: Single feeder, b: Multiple feeders	Compact, Diffuse	—	a: Small shunt, b: Medium shunt, c: Large shunt
Feeder	Radiculomeningeal artery	ASA, PSA, Radiculopial artery	ASA, PSA, Radiculopial artery	ASA, PSA, Anterior radiculomedullary artery, Posterior radiculomedullary artery
Drainer	Posterior perimedullary vein, Anterior erimedullary vein, Epidural vein	Perimedullary vein	Perimedullary vein	Anterior perimedullary vein, Posterior perimedullary vein
Median age	Approximately 60 years	Approximately 25 years	Approximately 25 years	Approximately 35 years
Male/female	5:1	Unclear (more common in men)	Unclear (more common in men)	Unclear
Frequent site	Midthoracic to mid-lumbar	Cervical, Thoracolumbar	Cervical, Thoracolumbar	Lower thoracic to mid-lumbar
Pathophysiology	Venous hypertension	Hemorrhage	Hemorrhage	Venous hypertension
Typical symptoms	Progressive myelopathy (relatively rapid progression)	Acute myelopathy or pain, Progressive myelopathy (gradual progression)	Acute myelopathy or pain, Progressive myelopathy (gradual progression)	Progressive myelopathy (relatively gradual progression)
Recommended primary treatment	Embolization	Embolization	Palliative embolization	Type a: Surgery Type b: Surgery Type c: Palliative embolization

AVF: ateriovenous fistula, AVM: arteriovenous malformation, ASA: anterior spinal artery, PSA: posterior spinal artery.

cord and flow void in the lower thoracic spine (Fig. 1a). She was initially treated with rest and steroid injection.

She was referred to a specialist and was transferred to our hospital to confirm the diagnosis. Contrast-enhanced computed tomography (CT) angiography showed abnormally dilated, tortuous vessels along the thoracic spinal cord. A high-density mass thought to be an A-V shunt was seen in the ventral aspect of the spinal cord at the T10 level (Fig. 1b and c). According to our standard practice, we consulted a general endovascular treatment team (well trained in conventional endovascular embolization) at our hospital. Thirty-four days after onset, the treatment team performed spinal angiography, which revealed a perimedullary AVF at the anterior surface of the spinal cord. The feeder was found to branch from the descending ASA. The ASA was connected to the enlarged anterior artery (artery of Adamkiewicz), which arose from the left T10 segmental artery (Fig. 2). The fistula was associated with an intranidal aneurysm (Fig. 2: asterisk). In the venous phase, marked venous congestion with dilatation of spinal veins was seen (image not shown). The ASA was also opacified from the Rt. T12 segmental artery connected with the origin of the feeder. Because they were unable to traverse the artery of Adamkiewicz with the tip of a catheter, and because of the risk of occluding the ASA or Adamkiewicz artery, they felt it would not be possible to perform this treatment. In addition, if we performed an open operation, the location of the lesion would make performance of delicate maneuvers (such as hemostasis) difficult to nearly impossible.

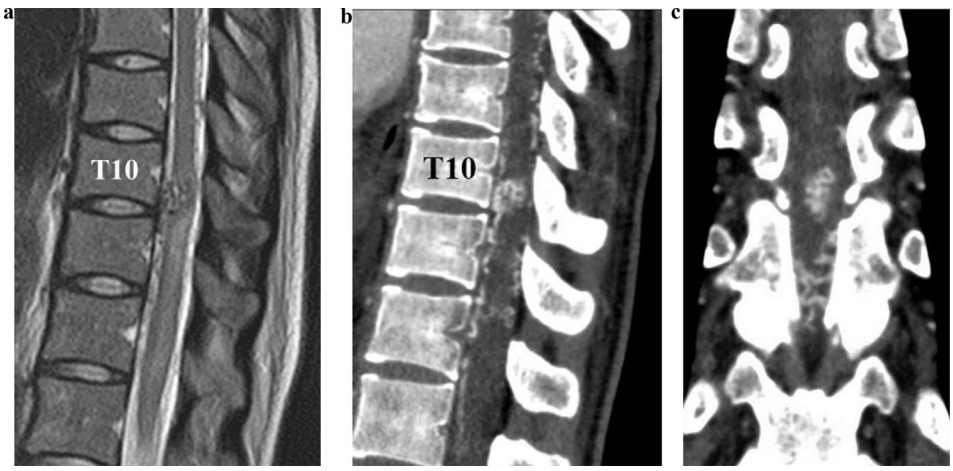

Fig. 1. (a) MRI of the lower thoracic spine before treatment. Sagittal T2-weighted image shows serpentine flow voids (congestive venous network) posterior to the thoracic spinal cord, with abnormal intramedullary signal intensity at the T10 level. Edema (high intensity) of the lower thoracic spinal cord is also seen. (b, c) Contrast-enhanced CT angiography of the lower thoracic spine before treatment. Sagittal and coronal image shows abnormal, dilated vessels along the thoracic spinal cord at the T10–11 level and a high-density mass thought to be the abnormal A-V shunt.