JOURNAL OF NEUROSURGERY:

J Neurosurg Case Lessons 8(8): CASE2457, 2024 DOI: 10.3171/CASE2457

Spinal epidural arteriovenous fistula with an intraosseous shunt arising in a compression fracture vertebra: illustrative case

Natsuki Akaike, MD, Masakazu Okawa, MD, PhD, Akira Ishii, MD, PhD, Takayuki Kikuchi, MD, PhD, Yukihiro Yamao, MD, PhD, Yu Abekura, MD, PhD, Hirofumi Tsuji, MD, Ryo Akiyama, MD, So Matsukawa, MD, and Yoshiki Arakawa, MD, PhD

Department of Neurosurgery, Kyoto University School of Medicine, Shogoin Kawahara-Cho Sakyo-ku, Kyoto, Japan

BACKGROUND Spinal epidural arteriovenous fistulas (SEAVFs) with intraosseous shunts are rare, and their underlying pathophysiological mechanisms remain unclear.

OBSERVATIONS A female in her 70s presented with rapidly progressive weakness in both lower extremities and urinary retention. Lumbar spine magnetic resonance imaging revealed spinal cord edema and flow voids due to venous dilation and compression fractures of the L1 and L2 vertebral bodies. Spinal angiography revealed ventral and dorsal somatic branches of the lumbar arteries at L1 and L2 flowing into the shunt. High-resolution cone-beam computed tomography revealed a shunt within the compression-fractured vertebral body bone of L2. The intravertebral shunt blood flowed into the ventral epidural venous plexus (VEVP) and returned into the perimedullary vein (PMV). Transarterial embolization was performed using N-butyl cyanoacrylate and Onyx-18 for feeder L1 and feeder L2, respectively. Onyx-18 was injected from the VEVP into the PMV, and complete occlusion of the shunt was achieved. The patient showed symptomatic improvement postoperatively.

LESSONS Vertebral compression fractures are common but rarely associated with SEAVFs.

https://thejns.org/doi/abs/10.3171/CASE2457

KEYWORDS spinal epidural arteriovenous fistula; compression fracture; embolization

Spinal epidural arteriovenous fistulas (SEAVFs) are rare spinal vascular lesions, often misdiagnosed as spinal dural arteriovenous fistulas (AVFs) given their similar clinical symptoms and vascular structures. Advances in imaging techniques, such as three-dimensional rotational spinal angiography, and an improved understanding of the anatomy of spinal vascular lesions have led to an increasing frequency of SEAVF diagnoses.¹ The pathophysiological mechanism of SEAVFs remains unclear, although causes such as neurofibromatosis, previous surgery, or trauma have been reported.² Shunts of SEAVFs are mainly located in the ventral epidural venous plexus (VEVP) and are primarily fed by the dorsal somatic branch of the lumbar artery.³ Osseous involvement is occasionally seen in SEAVFs, and these are reported as spinal osseous epidural AVFs, an extremely rare sublineage of SEAVFs.⁴ Bone erosion by the epidural

venous pouch or varix secondary to SEAVFs is common in osseous involvement. Here, we describe the extremely rare case of an intraosseous SEAVF involving a vertebral compression fracture with perimedullary reflux causing myelopathy symptoms. The intraosseous fistula was located in the fractured body. We aim to present the unique imaging features of intraosseous communication, clarify the relationship between traumatic compressive fractures and extradural AVF formation, and propose a pathophysiological hypothesis for such rare cases.

Illustrative Case

History and Examination

A woman in her 70s with progressive paraparesis was admitted to our hospital. Neurological abnormalities included weakness (manual

ABBREVIATIONS AVF = arteriovenous fistula; CT = computed tomography; MMT = manual muscle testing; MRI = magnetic resonance imaging; NBCA = N-butyl cyanoacrylate; PMV = perimedullary vein; SEAVF = spinal epidural arteriovenous fistula; TAE = transarterial embolization; VEVP = ventral epidural venous plexus. INCLUDE WHEN CITING Published August 19, 2024; DOI: 10.3171/CASE2457. SUBMITTED January 27, 2024. ACCEPTED March 22, 2024.

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FIG. 1. A: Sagittal T2-weighted MRI showing a high signal suggestive of spinal cord congestion and a flow void around the spinal cord (*arrows*). **B:** Sagittal T2-weighted MRI showed compression fractures in the L1 and L2 vertebrae, with bony erosion within the L2 vertebra (*arrowhead*).

muscle testing [MMT] grade 2/5), dysuria, and diarrhea. Computed tomography (CT) angiography of the lumbar region revealed abnormal darkening of the epidural venous plexus at the L2 level of the spine. Lumbar magnetic resonance imaging (MRI) showed a T2 high signal suggestive of spinal cord congestion and flow voids around the spinal cord (Fig. 1A), as well as compression fractures of the L1 and L2 vertebral bodies, which were asymptomatic, with an unknown time of onset (Fig. 1B). The patient was treated for suspected spinal arteriovenous malformation.

Treatment

Diagnostic angiography and endovascular treatment were performed with the patient under general anesthesia. Hyoscine butylbromide was used to suppress intestinal peristalsis. Lumbar arteriography of the left L1 and L2 revealed an AVF (Fig. 2A-C). High-resolution cone-beam CT showed a bone defect in the compression-fractured L2 vertebra (Fig. 2D) and a shunt in a similar area, forming a venous pouch (Fig. 2E). The lesion was diagnosed as an osseous SEAVF with perimedullary drainage. The L2 level showed the ventral and dorsal somatic branches perfusing the shunt through the vertebral body. The anterior spinal artery was also delineated from the left 9th intercostal artery but was not associated with the lesion. Since L2 was the main feeder, a branch of L1 was embolized with 20% N-butyl cyanoacrylate (NBCA; Fig. 3A). A Rebar catheter (Medtronic) was then guided posteriorly to the vertebral body near the shunt through the dorsal somatic branch of the L2 segmental artery, from which Onyx-18 (Covidien) was injected. Transarterial embolization (TAE) was performed using Onyx-18 and injected from the VEVP to the perimedullary vein (PMV) using the plug-and-push technique (Fig. 3B). The final angiogram revealed complete fistula obliteration (Fig. 3C).

After 3 months, no recurrence of the lesion was observed. Postoperative lumbar MRI revealed the disappearance of the flow void (Fig. 4). The patient's muscle strength in the bilateral lower extremities improved to MMT grade 4/5 and she could walk with a cane, and symptoms of cystorectal disability improved.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

This patient's most distinctive feature was a fistula located within a fractured vertebral body. CT revealed the shunt area as a bone defect image, which was a characteristic imaging finding. This case involved a spinal vascular lesion with an intraosseous shunt in the epidural space. It was first reported by Chul Suh and colleagues in 2004 as a spinal osseous epidural AVF, which is considered a subtype of SEAVF.⁴ SEAVFs associated with vertebral compression fractures are extremely rare, with only a few reported cases in the literature.⁵⁻¹¹ Table 1 provides the details of 8 cases, including this case. There were 5 males and 3 females, aged 57-74 years, with a mean age of 69 years. Seven patients underwent TAE alone or in combination with transvenous embolization using coils, NBCA, or Onyx. Complete shunt occlusion was observed in all cases. One case of direct surgery was reported wherein complete shunt occlusion was not achieved, but reduction of the venous pouch was achieved.9 No treatment-related complications were reported. All patients obtained improvement in postoperative symptoms, and treatment prognosis was favorable.

SEAVFs can be categorized into type A and type B based on the presence or absence of intradural venous drainage.¹² Type A SEAVFs, which are diagnosed in patients in their 6th decade of life, typically present with venous congestive myelopathy in the thoracolumbar and lumbar regions due to intradural venous reflux. Type B SEAVFs, which are usually diagnosed in patients in their 3rd decade of life, present with compressive myelopathy or radiculopathy in the cervical and upper thoracic regions due to compression of the thecal sac or root sleeves by an enlarged extradural venous plexus. Previous reports described 6 cases of type A and 1 of type B SEAVFs. The present case involved a spinal osseous epidural AVF with perimedullary drainage and was classified as type A.

Geibprasert et al. introduced a new classification of dural AVF based on craniospinal epidural venous anatomy,¹³ outlining 3 venous subgroups based on the relationship between epidural venous spaces and afferent veins from the bone and central nervous system. This classification includes the following: 1) ventral epidural or osteocartilaginous epidural group, 2) dorsal epidural or osteomembranous epidural group, and 3) lateral epidural or leptomeningeal epidural group. SEAVF aligns with the ventral epidural or osteocartilaginous epidural groups, where dural AVF shunts in these areas mainly involve the epidural space. These shunts often directly contact adjacent osseous structures, potentially invading or recruiting blood from them. In this case, we hypothesized that a vertebral compression fracture was linked with developing an intraosseous shunt.

Herein, we present a pathophysiological hypothesis explaining the development of intraosseous shunts and progressive myelopathy. The feeders of the SEAVF are the dorsal branch and ventral



FIG. 2. Left L1 arteriography, anteroposterior (**A**) and lateral (**B**) projections, showing the SEAVF (*white arrow*) fed by the ventral and dorsal somatic branches of the lumbar artery and a venous pouch, with shunt blood flow into the VEVP and back into the PMV (*black arrow*). Left L2 lumbar arteriography, anteroposterior projection (**C**), showing a high-flow SEAVF (*white arrow*) fed by the ventral and dorsal somatic branches of the lumbar artery and a venous pouch, with shunt blood flow into the VEVP and back into the PMV (*black arrow*). Left L2 lumbar arteriography, anteroposterior projection (**C**), showing a high-flow SEAVF (*white arrow*) fed by the ventral and dorsal somatic branches of the lumbar artery and a venous pouch, with shunt blood flow into the VEVP and back into the PMV (*black arrow*). Axial high-resolution cone-beam CT image showing a compression fracture of the L2 vertebra, with internal bone erosion (*white asterisk*, **D**). Dorsal and ventral somatic branches are depositing blood into a shunt, and a venous pouch (*black asterisk*, **E**) is consistent with the bone erosion image.

somatic branches of the lumbar artery, which both supply the vertebral body. Initially, compression fracture-induced damage to the vascular walls of these branches perforates the venous side, forming an intraosseous shunt. The spinal veins are guided from the intradural to the epidural venous plexus via the radiculomedullary vein or bridging vein and perfuse from the epidural venous plexus into the paravertebral veins, such as the ascending lumbar vein and azygos vein via the emissary vein, which drain out of the spinal canal. The radiculomedullary vein is physiologically narrowed and tortuous while crossing the dura mater and has an anti-backflow system.¹⁴ If an event disrupts this system, like the breakdown of the anti-backflow system, shunt flow can retrogradely drain into the PMVs, leading to subsequent venous congestion, spinal cord edema, and progressive myelopathy.⁸ Disruption of this system is believed to result from venous thrombus inflammation around the vertebral body.⁸ The compression fracture remained asymptomatic, and the time course of SEAVF development and the cause of disruption of the anti-backflow system remained unknown. We assumed



FIG. 3. A: TAE was performed using 20% NBCA for the feeder from the lumbar artery. **B:** The Rebar catheter was guided posteriorly to the vertebral body near the shunt through the L2 segmental artery, from which Onyx-18 was injected. TAE was performed using Onyx-18 and injected from the VEVP to the PMV (*white arrows*). **C:** The final angiogram revealed complete obliteration of the fistula.

that the patient in our case had preexisting vertebral compressive fractures, potentially harboring an asymptomatic SEAVF due to the deficient anti-backflow system. An objection to this hypothesis arises



FIG. 4. Postoperative lumbar MRI showing the disappearance of the flow void.

from studies suggesting that the anti-backflow system may decompensate under high-pressure conditions, allowing venous blood to flow in either direction.¹⁵ However, none of these studies were physiological, as they involved the injection of contrast agents or anatomical materials under artificial pressure and various adjuvant maneuvers. Gailloud proposed that the anti-backflow system likely involves physiological factors (e.g., muscular tone and cerebrospinal fluid pressure) alongside the purely morphological barrier established by the narrowing of the radiculomedullary vein at its transdural passage point.¹⁶

Endovascular treatment, direct surgery, or a combination of the two are options for SEAVF management; however, owing to vascular structure complexity, the optimal approach remains undetermined. Endovascular treatment is more frequently reported than direct surgery, given the complexity of anterior lesions, posing challenges for adequate exposure from a posterolateral approach. A review of TAE for SEAVF revealed low occlusion rates when shunt blood flow also flowed out of the dura mater: however. TAE occlusion was achieved in 90% of cases with only intradural regurgitation.1 Good results involve occluding the proximal side of the venous pouch and draining the vein with liquid embolic substances such as NBCA or Onyx.^{3,17} In high-flow lesions, completely filling and occluding the shunt from the entire affected vein with low-concentration NBCA is challenging. Recently, the usefulness of TAE using Onyx has been reported.8,17-19 With the goal of filling the venous plexus and occluding the proximal root vein, Onyx, which can be injected in a sufficient volume, may be useful. In this case, the feeder from L2, which was distant from the anterior spinal artery and had a high flow rate, was embolized with Onyx. TAE was performed using Onyx, injected from the VEVP to the PMV, and complete shunt occlusion was achieved. Despite the need to monitor the mass effect in the spinal canal and avoid straying into vessels feeding the spinal artery and nerve roots, even high-flow lesions like this one can be cured, making TAE with Onyx a viable treatment option.

Lessons

We present the rare case of an intraosseous SEAVF secondary to a thoracolumbar compression fracture with perimedullary venous reflux causing progressive myelopathy. The fistula was located in the fractured vertebral body. It was a high-flow lesion, and TAE with Onyx was radical treatment.

TABLE 1. Summary c	of reported	l cases of	SEAVFs associat	ted with verte	bral compression fractu	res						
				Duration From Onset								
	Age	Level of	Level of	Until		Type of	Draining		Treatment			
Authors & Year	(yrs)/Sex	Fistula	Fractured VB	Treatment	Symptom	SEAVF	Route	Treatment	Result	Complication	Recurrence	Outcome
Gjertsen et al., 2010 ⁵	67/M	T12	T12	Several yrs	Myelopathy	A	ParaW, PMV	TAE (coil), direct embolization (NBCA)	8	None	None	GR
Jin et al., 2010 ⁶	68/F	L	L1	9 yrs	Myelopathy	A	ParaVV, PMV	TAE (NBCA)	8	None	None	GR
Imajo et al., 2015 ⁷	74/F	L4	L4	3 mos	Back pain	в	ParaW	TAE (coil)	8	None	None	GR
Ou et al., 2015 ⁸	57/M	T12	Т6, Т9, Т12, L2	>5 yrs	Back pain, myelopathy	A	PMV	TAE (Onyx)	8	None	None	GR
Shimizu et al., 20219	71/F	T12	T12	1.5 yrs	Myelopathy	A	ParaVV, PMV	Resection	РК	None	None	GR
Furuta et al., 2021 ¹⁰	71/F	T12	T12, L1	10 yrs	Myelopathy	A	PMV	TAE (NBCA)	8	None	None	GR
Baba et al., 2022 ¹¹	74/M	T12	T12	Unknown	Back pain, myelopathy	A	ParaVV, PMV	TVE, TAE (NBCA)	8	None	None	GR
Present case	70/F	L2	L1, L2	Unknown	Myelopathy	A	PMV	TAE (Onyx)	CO	None	None	GR
CO = complete obliteration	ו; GR =good	recovery; F	araVV = paravertebr:	al vein; PR =par	tial resolution; TVE = transver	nous embo	olization; VB = verte	bral body.				

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J Neurosurg Case Lessons | Vol 8 | Issue 8 | August 19, 2024 | 5

Disclosures

Dr. Arakawa reported grants from Philips, Otsuka, Chugai, Nihon Medi-Physics, Daiichi Sankyo, Stryker, Eisai, Japan Blood Products Organization, Ono Pharmaceutical, Taiho Pharma, Sumitomo Dainippon Pharma, Astellas Pharma, Incyte Biosciences, and Servier; and personal fees from Nippon Kayaku, Novocure, UCB Japan, Ono Pharmaceutical, Brainlab, Merck, Chugai, Eisai, Daiichi Sankyo, Carl Zeiss, and Nihon Medi-Physics outside the submitted work.

Author Contributions

Conception and design: Okawa, Akaike. Acquisition of data: Ishii, Yamao, Abekura, Tsuji, Akiyama. Analysis and interpretation of data: Abekura,

Akiyama. Drafting the article: Okawa, Arakawa. Critically revising the article: Okawa, Kikuchi, Akiyama, Arakawa. Reviewed submitted version of manuscript: Kikuchi, Yamao, Akiyama, Matsukawa, Arakawa. Approved the final version of the manuscript on behalf of all authors: Okawa. Administrative/technical/material support: Okawa, Matsukawa, Arakawa. Study supervision: Okawa, Arakawa.

Correspondence

Masakazu Okawa: Kyoto University School of Medicine, Sakyo-ku, Kyoto, Japan. okawam@kuhp.kyoto-u.ac.jp.