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Cervical Extradural Arteriovenous Fistula without Intradural Drainage Successfully Treated with Endovascular Treatment Using Both Transvenous and Transarterial Approach: Case Report and Review of Literatures

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Abstract

The classification of spinal extradural arteriovenous fistulas (AVFs) was reported based on a case series treated by microsurgery in 2009 and endovascular interventions in 2011. The present report describes a patient with extradural AVFs at the cervical spine manifesting gradual progressive radiculomyelopathy of bilateral upper extremities. Magnetic resonance imaging (MRI) revealed a mass sign from C1 to C4 at the right ventral side and the spinal cord was deviated to the left and indicated as a flow void sign. Diagnostic angiography revealed an extradural AVFs located at the C1–C4 level that was supplied by bilateral radicular artery from the vertebral artery (VA) and right ascending cervical artery (ACA). The shunting points were recognized multiply at C2/3 and C3/4 levels on the right. The transvenous embolization to the enlarged extradural venous plexus around the shunting points via right hypoglossal canal and the transarterial embolization against multi-feeders of the branch of left radicular artery, right ACA achieved complete occlusion of the lesions. His symptom was gradually recovered, and angiography performed 2 weeks after embolization showed no recurrence. When the arteriovenous shunts in the upper cervical spine were high flow shunts, transvenous approach via the hypoglossal canal might be one option for the treatment of spinal extradural AVFs.

Keywords: extradural arteriovenous fistula, cervical spine, endovascular treatment, anterior condylar emissary vein

Introduction

Spinal arteriovenous malformations (AVMs) including spinal arteriovenous fistulas (AVFs) were classified into four types based on the vascular supply, vascular drainage, and nidus location according to the traditional classification system.[1–4)] Dural AVFs are most common forms of spinal AVMs and extradural AVF accounts for approximately 1.6% of all spinal AVMs[5)] The classification of spinal extradural AVFs was reported based on the case series treated by microsurgery in 2009 and endovascular interventions in 2011[6–9)] Moreover, the review and treatment strategy of spinal extradural AVFs according to the venous drainage patterns were reported.[7,8)] Rangel-Castilla et al. classified that extradural AVFs have two distinct subtypes: extradural AVFs with intradural venous drainage (type A) and extradural AVFs

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with pure extradural drainage only (type B). The type B extradural AVF in the cervical spine is rare among extradural AVFs and over a dozen cases were reported in the literature[6–22] and almost cases were treated with transarterial approach. Takai et al. proposed that the goal of the treatment is the mass reduction of the venous plexus in patients with extradural AVFs without intradural venous drainage which was classified as type B. Because, the cause of compressive myelopathy is considered due to the compression from the enlarged extradural vertebral venous plexus.[12] Endovascular embolization of the feeding arteries was performed as the first-choice treatment to block arterial reflux into the extradural venous plexus. 66.7% (2/3 cases) could complete or nearly complete embolization with transarterial embolization.[12] Here, we report a case of a cervical spinal extradural AVFs without intradural venous drainage, which was successfully treated with the combination of transvenous and supplementary transarterial embolization.

Case Report

A 59 year-old man, who had a past history of myasthenia gravis, presented to our hospital. He had no traumatic episode and no prior cervical spine surgery. He had been aware of bilateral upper limbs pain, skillful movement disorders, and gait disturbance from approximately 5 months before and the symptoms had gradually progressed. Cervical X-ray showed a dilation of the right foramen at C3/4 and no instability. On magnetic resonance imaging (MRI), an epidural mass was recognized located around the right foramen of the C2–C4 compressing the spinal cord to the left (Fig. 1a–1c).

The mass was consisted with low signal intensity both on T1 and T2 suggesting a flow void of a vascular lesion. A part of the mass lesion was enhanced using contrast enhancement (Fig. 1d). 3D-computed tomography angiography indicated extradural AVFs on the right in the cervical spine with multi-feeders (Fig. 1e).

Angiogram indicated multi-shunting points at the epidural space of right foramens of C2/3 and C3/4. The right subclavian artery angiogram demonstrated a high flow arteriovenous shunts at the extradural space around the right foramen of C3/4 from multi-branches of the ascending cervical artery (ACA) which consisted the main portion of the AVF (Fig. 2a). Right vertebral artery (VA) angiogram demonstrated arteriovenous shunts at the extradural space around the right foramen of C3/4 from radicular artery of V1 portion (Fig. 2b). Left VA angiogram demonstrated arteriovenous shunts at the extradural space around the right foramen of C2/3 and C3/4 from radicular artery of V1 portion (Fig. 2c). The flow was drained to the extradural vertebral venous plexus without intradural and intracranial drainage. Cone beam CT images more clearly suggested that it was extradural AVF (Fig. 3). Based on the above

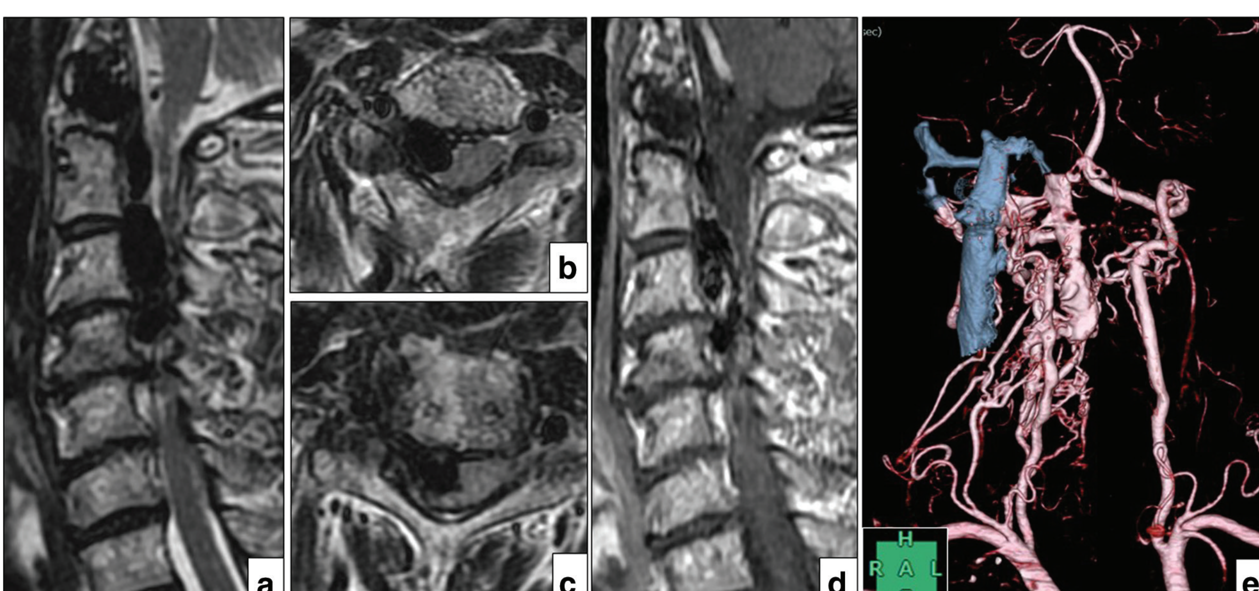

Fig. 1 Preoperative MR images and CTA image. (a) T2 on sagittal view, (b) T2 on axial at C2/3, (c) T2 on axial at C3/4, (d) Gd-enhanced T1 on sagittal view, and (e) 3D construction image (red: artery, blue: vein). CTA: computed tomographic angiography, Gd: gadolinium, MR: magnetic resonance, 3D: three-dimensional.

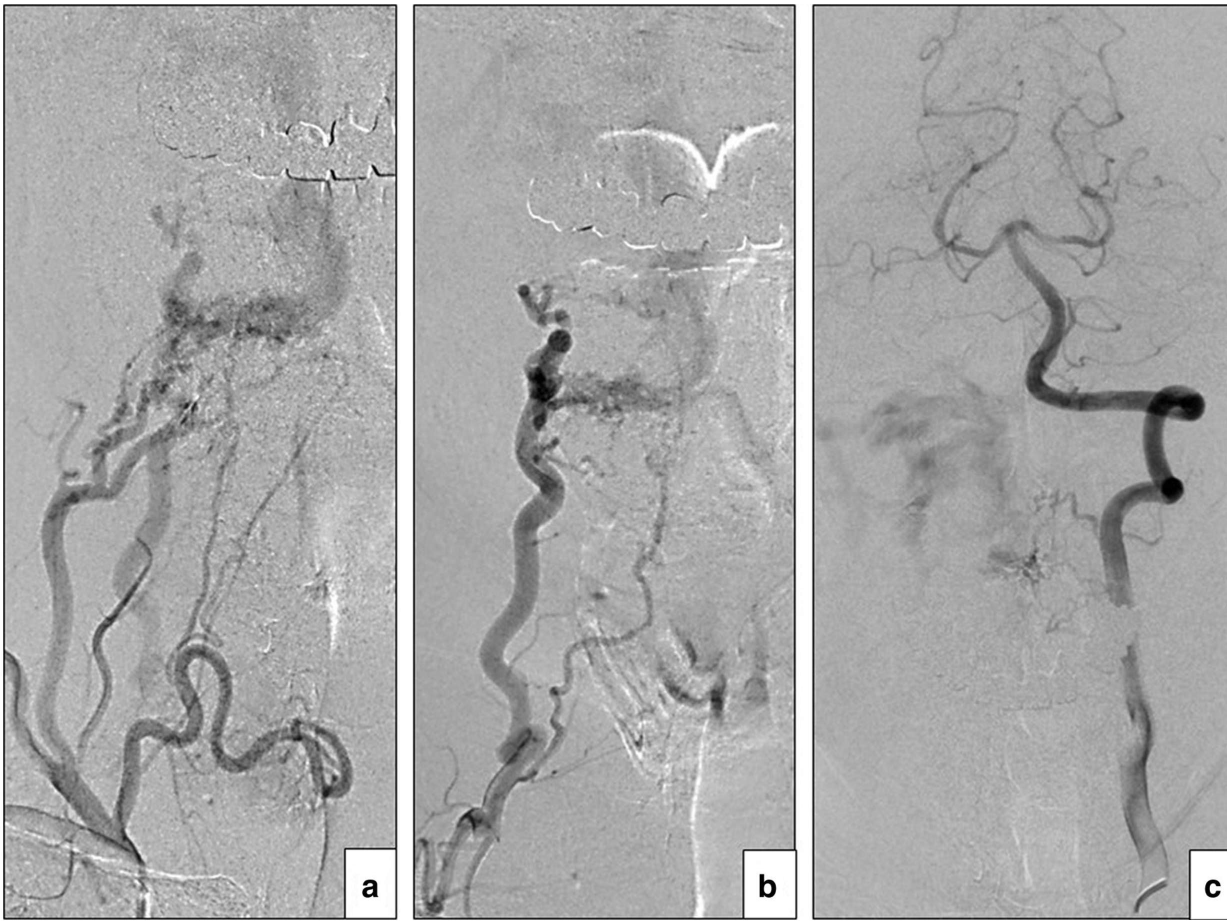

Fig. 2 Angiogram. (a) Right subclavian angiogram, (b) right VA angiogram, and (c) left VA angiogram.

findings, it was diagnosed as extradural AVF classified as type B1 by Rangel-Castilla L.

Endovascular treatment was performed under general anesthesia and systemic heparinization to maintain the activated clotting time at level above twofold to threefold of the baseline value. First, transvenous embolization was conducted. First, 6 French intermediate catheter assembled with 8 French guiding catheter system was placed in the right internal jugular vein via right femoral vein. The microcatheter (Excelsior 1018; Stryker, Fremont, CA, USA) was coaxially inserted into extradural space at C3/4 on the right side nearby the shunting point via anterior condylar emissary vein in the hypoglossal canal and coil embolization was performed (Fig. 4). After this procedure, most of the shunt volume decreased. Second, 5 French guiding catheter (Envoy; Cordis, Miami Lakes, FL, USA) was placed in the left VA via left femoral artery. The microcatheter (Excelsior SL-10; Stryker, Fremont, CA, USA) was coaxially inserted into the radicular artery at left V1 portion and coil embolization was done. Third, the same 5 French guiding catheter was repositioned in the origin of right ACA and the branches of right ACA embolized with n-butyl cyanoacrylate (NBCA; 33% concentration) to prevent the recurrence form the collateral branches. The combination of transvenous and transarterial embolization leaded complete arteriovenous shunts obliteration (Fig. 5). The systemic heparinization was naturally reversed because the AVF was not related to the intraspinal venous reflux. The myelopathy gradually recovered and the follow up angiogram 2 weeks later demonstrated complete shunt obliteration.

Discussion

Spinal dural AVFs commonly cause the progressive myelopathy because the arterial blood reflux into the intradural veins increase medullary venous pressure.[23] On the other hand, spinal extradural AVFs cause myelopathy or radiculopathy because abnormal direct connection between arteries and the extradural venous plexus within spinal canal causes venous hypertension, mechanical compression,

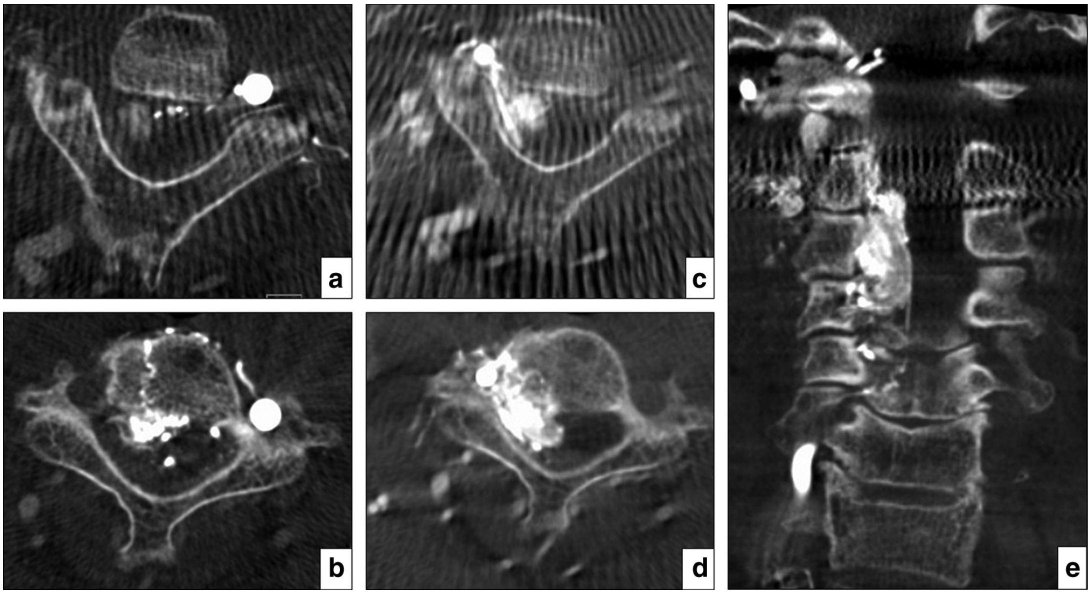

Fig. 3 Cone beam CT image with right and left VA angiogram showed the direct reflux to extradural vertebral venous plexus. (a) Axial image with right VA angiogram at C2 level, (b) axial image with left VA angiogram at C3 level, (c) axial image with right VA angiogram at C2 level, (d) axial image with right VA angiogram at C3 level, and (e) coronal image with right VA angiogram. CT: computed tomography, VA: vertebral artery.

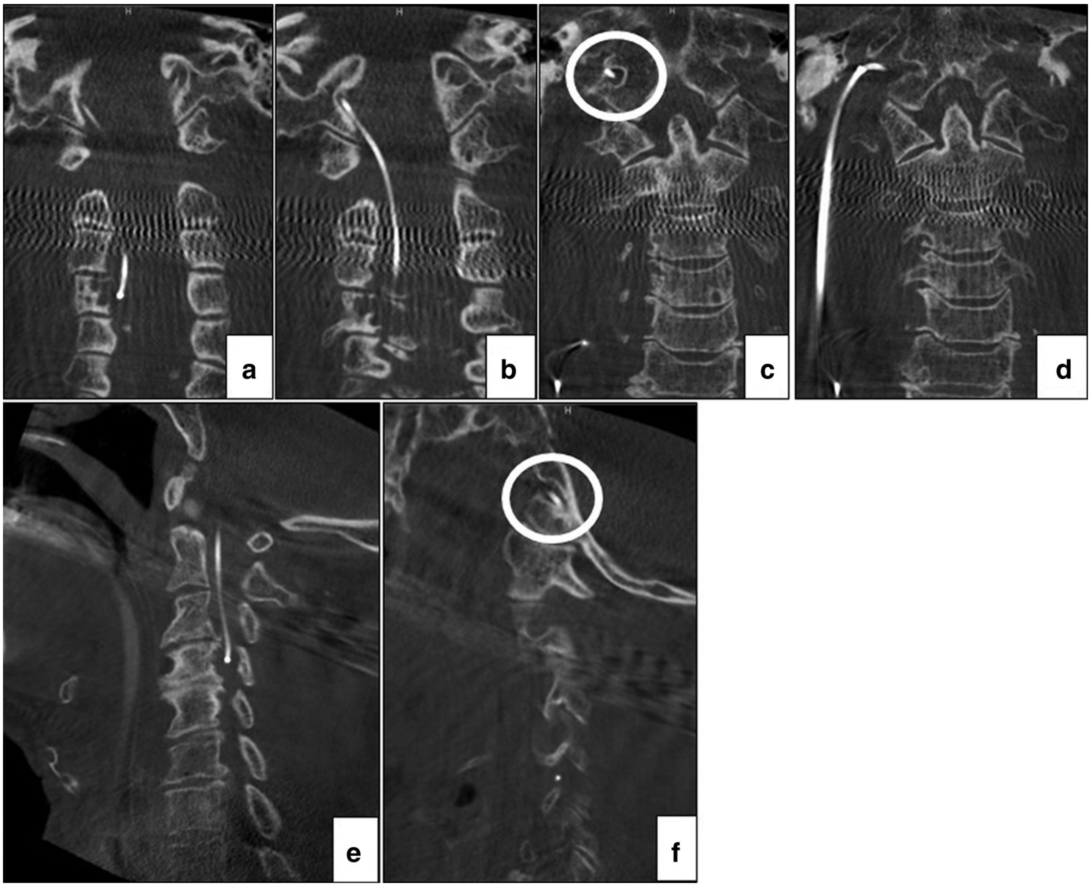

Fig. 4 Cone beam CT image demonstrated that the microcatheter was inserted into extradural space at C4 body level via hypoglossal canal. White ring: hypoglossal canal. (a–d) Coronal image, (e, f) sagittal image. CT: computed tomography.

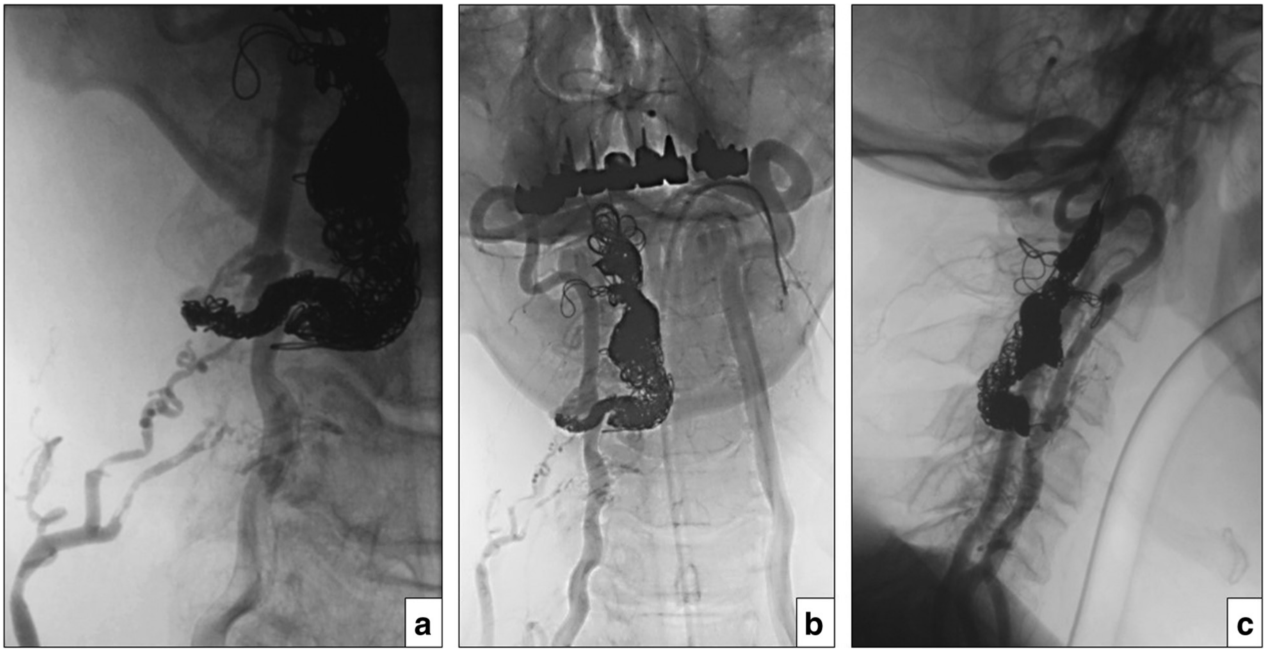

Fig. 5 Final angiogram. (a) Right subclavian angiogram. Arteriovenous shunts were obliterated. (b, c) Bilateral VA angiogram at the same time. Arteriovenous shunts were obliterated. VA: vertebral artery.

and vascular steal effect.[6,8,18,24–26] Extradural AVFs rarely causes hemorrhage.[18]

Kiyosue et al.[27] reviewed angiographic and clinical characteristics of epidural and dural AVFS in the thoracolumbar spine. In the review of epidural AVFs in 59 and dural AVFs in 108, epidural AVFs and dural AVFs showed similar symptoms and male predominance. There were no significant differences in age, sex, or symptoms. A history of spinal surgery or trauma was less frequently observed in the dural AVF (12%) than epidural AVFs (36%). Huang et al.[8] reviewed spinal extradural AVFs in 2013. In this review, 54 cases in cervical spine were summarized. As a pertinent history, neurofibromatosis type 1 of NF1, prior spinal surgery, and prior spinal trauma were 16/54 (30%), 5/54 (9%), and 7/54(13%), respectively. 50/54 (92%) showed extradural drainage only. This review was excellent study but contained many old reports and it is unclear whether its exact diagnosis of extradural AVF was correct.

In 2011, Rangel-Castilla et al.[7] classified that extradural AVFs have two distinct subtypes: extradural AVFs with intradural venous drainage (type A) and extradural AVFs with pure extradural drainage only (type B). In 2012, Takai et al.[28] analyzed and reviewed the spinal extradural AVFs with or without intradural venous drainage published between 1990 and 2011. In the review, posttraumatic and postoperative extradural AVFs were excluded because the spinal extradural AVF is defined as a spontaneous abnormal direct connection between an artery or arteries and the extradural venous plexus. According to the reviews, spinal extradural AVFS with intradural venous drainage (type A) are diagnosed in patients around the 6th decade of life and causes myelopathy due to venous congestion, which is similar to dural AVFs. On the other hand, spinal extradural AVFs without intradural venous drainage (type B) are diagnosed in patients around the 3rd decade of life and had a tendency to show a normal signal intensity of the cord with a severe mass effect due to an enlarged extradural venous plexus; they commonly occurred in the cervical and upper thoracic regions (18/22, 82%). Therefore, type B extradural AVFs present with compressive myelopathy or radiculopathy due to the compression of the thecal sac or root sleeves by the enlarged extradural venous plexus. We summarized the extradural AVFs without venous drainage (type B) in the cervical spine by adding the cases reported after 2012 (Table 1). In almost cases, transarterial approach is chosen for spinal extradural AVFs. In Huang W's review, 65 cases of extradural AVFs in whole spine among 101 cases underwent endovascular approach and the approach was transarterial in 87% of cases, transvenous in 11%, and a combination in 2%.[8] Embolization materials included NBCA in 34% of cases, coils in 34%, balloons in 23%, and Onyx in 17%. Takai et al.[12] reported that 66.7% (2/3 cases) could complete or nearly complete embolization

Table 1 Summary of clinical characteristics in patients with type B spinal extradural AVFs

Authors	Year	Age (years), sex	Presentation	Location of extradural veins	Feeders	Diffuse T2 high on MRI	Mass effect on MRI	Treatment	Occlusion of AVFs	Outcome
Brinjikji et al.	2020	13, F	Tetraparesis	C4–C7	VA, thyrocervical trunk	+	+	TAE (PVA)	Total	Improved
Takai et al.	2018	38, M	Tetraparesis	C3–C6	VA, ACA, DCA	–	+	TAE (NBCA, coils), microsurgery	Subtotal	Improved
		20, F	Epidural hematoma, Tetraparesis	C5-T1	ACA	–	+	TAE (NBCA, coils)	Total	Improved
		77, M	Epidural hematoma, Tetraparesis	C7	DCA	–	+	TAE (NBCA, coils)	Subtotal	Improved
Nakagawa et al.	2014	54, M	SAH	C1–2	VA, ASA	No description	No description	TAE (NBCA)	Subtotal	Improved
Puri	2014	45	Headache	C1–C2	VA	–	–	TAE (onyx)	Total	Improved
Rangel-Castilla et al.	2011	57, M	Myelopathy	C2-T1	ACA	–	+	TAE (onyx, coils)	Total	Improved
Wang et al.	2011	20, F	Myelopathy	C3–C7	VA, ACA	–	+	TAE (onyx, coils)	Total	Improved
Paolini et al.	2008	26, M	Myelopathy, NF1	C1–C5	VA,ECA,DCA	–	+	TAE (balloon, coils), microsurgery	Total	Improved
Tenjin, et al.	2005	72, F	Myelopathy	C6	VA	–	+	TAE (coils)	Total	Improved
Chuang et al.	2003	4, F	Epidural hematoma, Myelopathy	C6–C7	DCA	–	+	TAE (NBCA), microsurgery	Total	Improved
Kahara et al.	2002	38, M	Neck pain, NF1	C2–C4	VA	–	+	TAE (coils)	Total	Improved
Asai et al.	2001	24, M	Myelopathy	C5-T2	ACA, DCA	+	+	TAE (NBCA), microsurgery	Partial	Improved
Taylor et al.	2001	41, F	Radiculopathy	C1-T1	VA	–	+	TAE (coils)	Total	Improved
		44, F	Radiculopathy	C2–C7	VA	–	+	TAE (balloons)	Total	Improved
Goyal et al.	1999	68, M	Myelopathy	Cervical	VA, ECA, ACA, DCA	–	+	TVE (coils), 2 times	Partial	Improved
Szajner et al.	1999	48, F	Klippel-Trenaunary syndrome	C5–C7	VA, ACA, DCA	–	+	TAE (coils, NBCA), TVE (NBCA) 2 times	Total	Improved

ACA: ascending cervical artery, DCA: deep cervical artery, ECA: external carotid artery, NF1: neurofibromatosis type 1, VA: vertebral artery.

with transarterial embolization. In the lumbar spine, Kiyosue et al.[29] reported and classified the ventral epidural AVFs regarding the draining routes; the perimedullary venous drainage type (PM type), the paravertebral venous drainage type (PV type), and the combined perimedullary and paravertebral venous drainage type (PMPV type).[29] The type B extradural AVF in the cervical spine defined by Rangel-Castilla et al. was considered to correspond to the PV type by Kiyosue et al. and our case was also diagnosed as this type.

In our case, we tried transarterial embolization of the main feeders from ACA, but we judged that it was dangerous because the shunts were high flow and the NBCA and coils might flow over the shunting points into the jugular vein. Moreover, the coil embolization might result in only feeder occlusion and recurrence of the epidural AVFs because it could not obliterate the draining vein over multi-shunting points. Therefore, transvenous approach was performed as a first choice. Anterior condylar emissary vein in the hypoglossal canal from right jugular vein was chosen as the route to the enlarged cervical extradural venous plexus. Arteriovenous shunts from multi-branches of ACA almost disappeared. Next, transarterial embolization with coil and NBCA leaded complete obliteration of the AVFs. Theoretically, coil embolization into the venous plexus might be avoided because the coils may remain as a mass and may compress the dural sac and the root sleeve. However, the foramen at C3/4 on right was enlarged and the discontinuation of the arterial pulsation might improve myelopathy.

In this case, we did not choose microsurgical resection because the arteriovenous shunts supplied from bilateral VA and had multi-shunting points and was located at the ventral side. It would be very difficult to resect multi-feeders from bilateral VA and multi-shunts to control and stop bleeding even if complete removal of the facet joints was done with anterior or posterior approach.

Conclusion

Extradural AVF of the cervical spine is a rare disease and its treatment strategy should be discussed carefully. When it is an extradural AVFs without intradural venous drainage and cause compressive myelopathy due to an enlarged vertebral venous plexus, flow reduction is necessary. Transvenous approach via anterior condylar emissary vein in the hypoglossal canal might be one option when the arteriovenous shunts in the upper cervical spine has a high flow shunt.

Conflicts of Interest Disclosure

No potential conflicts of interest were disclosed.

References

1) Kim LJ, Spetzler RF: Classification and surgical management of spinal arteriovenous lesions: arteriovenous fistulae and arteriovenous malformations. *Neurosurgery* 59: S195–201; discussion S193-113, 2006
2) Mourier KL, Gobin YP, George B, Lot G, Merland JJ: Intradural perimedullary arteriovenous fistulae: results of surgical and endovascular treatment in a series of 35 cases. *Neurosurgery* 32: 885–891; discussion 891, 1993
3) Oldfield EH, Doppman JL: Spinal arteriovenous malformations. *Clin Neurosurg* 34: 161–183, 1988
4) Takai K, Kin T, Oyama H, et al.: The use of 3D computer graphics in the diagnosis and treatment of spinal vascular malformations. *J Neurosurg Spine* 15: 654–659, 2011
5) Bao YH, Ling F: Classification and therapeutic modalities of spinal vascular malformations in 80 patients. *Neurosurgery* 40: 75–81, 1997
6) Clarke MJ, Patrick TA, White JB, et al.: Spinal extradural arteriovenous malformations with parenchymal drainage: venous drainage variability and implications in clinical manifestations. *Neurosurg Focus* 26: E5, 2009
7) Rangel-Castilla L, Holman PJ, Krishna C, Trask TW, Klucznik RP, Diaz OM: Spinal extradural arteriovenous fistulas: a clinical and radiological description of different types and their novel treatment with Onyx. *J Neurosurg Spine* 15: 541–549, 2011
8) Huang W, Gross BA, Du R: Spinal extradural arteriovenous fistulas: clinical article. *J Neurosurg Spine* 19: 582–590, 2013
9) Asai J, Hayashi T, Fujimoto T, Suzuki R: Exclusively epidural arteriovenous fistula in the cervical spine with spinal cord symptoms: case report. *Neurosurgery* 48: 1372–1375; discussion 1375-1376, 2001
10) Kawabori M, Hida K, Yano S, Asano T, Iwasaki Y: Cervical epidural arteriovenous fistula with radiculopathy mimicking cervical spondylosis. *Neurol Med Chir (Tokyo)* 49: 108–113, 2009
11) Brinjikji W, Colombo E, Lanzino G: Clinical and angioarchitectural characteristics of spinal vascular malformations of the cervical spine. *J Neurosurg Spine* 32: 755–762, 2020
12) Takai K, Shojima M, Imai H, Saito N, Taniguchi M: Microsurgical and endovascular treatments of spinal extradural arteriovenous fistulas with or without intradural venous drainage. *World Neurosurg* 111: e819–e829, 2018
13) Nakagawa I, Park HS, Hironaka Y, Wada T, Kichikawa K, Nakase H: Cervical spinal epidural arteriovenous fistula with coexisting spinal anterior spinal artery aneurysm presenting as subarachnoid

hemorrhage--case report. *J Stroke Cerebrovasc Dis* 23: e461–e465, 2014
14) Puri AS, Telischak NA, Vissapragada R, Thomas AJ: Analysis of venous drainage in three patients with extradural spinal arteriovenous fistulae at the craniovertebral junction with potentially benign implication. *J Neurointerv Surg* 6: 150–155, 2014
15) Wang Q, Song D, Chen G: Endovascular treatment of high-flow cervical direct vertebro-vertebral arteriovenous fistula with detachable coils and Onyx liquid embolic agent. *Acta Neurochir (Wien)* 153: 347–352, 2011
16) Paolini S, Colonnese C, Galasso V, et al.: Extradural arteriovenous fistulas involving the vertebral artery in neurofibromatosis Type 1. *J Neurosurg Spine* 8: 181–185, 2008
17) Tenjin H, Kimura S, Sugawa N: Coil embolization of vertebro-vertebral arteriovenous fistula: a case report. *Surg Neurol* 63: 80–83, discussion 83, 2005
18) Chuang NA, Shroff MM, Willinsky RA, Drake JM, Dirks PB, Armstrong DC: Slow-flow spinal epidural AVF with venous ectasias: two pediatric case reports. *AJNR Am J Neuroradiol* 24: 1901–1905, 2003
19) Kahara V, Lehto U, Ryymin P, Helen P: Vertebral epidural arteriovenous fistula and radicular pain in neurofibromatosis type I. *Acta Neurochir (Wien)* 144: 493–496, 2002
20) Taylor CG, Husami Y, Colquhoun IR, Byrne JV: Direct cervical vertebro-venous fistula with radiculopathy and MRI changes resolving after successful endovascular embolisation: a report of two cases. *Neuroradiology* 43: 1118–1122, 2001
21) Goyal M, Willinsky R, Montanera W, terBrugge K: Paravertebral arteriovenous malformations with epidural drainage: clinical spectrum, imaging features, and results of treatment. *AJNR Am J Neuroradiol* 20: 749–755, 1999
22) Szajner M, Weill A, Piotin M, Moret J: Endovascular treatment of a cervical paraspinal arteriovenous malformation via arterial and venous approaches. *AJNR Am J Neuroradiol* 20: 1097–1099, 1999
23) Jellema K, Tijssen CC, van Gijn J: Spinal dural arteriovenous fistulas: a congestive myelopathy that initially mimics a peripheral nerve disorder. *Brain* 129: 3150–3164, 2006
24) Arnaud O, Bille F, Pouget J, Serratrice G, Salamon G: Epidural arteriovenous fistula with perimedullary venous drainage: case report. *Neuroradiology* 36: 490–491, 1994
25) Chul Suh D, Gon Choi C, Bo Sung K, Kim KK, Chul Rhim S: Spinal osseous epidural arteriovenous fistula with multiple small arterial feeders converging to a round fistular nidus as a target of venous approach. *AJNR Am J Neuroradiol* 25: 69–73, 2004
26) Alexander MJ, Grossi PM, Spetzler RF, McDougall CG: Extradural thoracic arteriovenous malformation in a patient with Klippel-Trenaunay-Weber syndrome: case report. *Neurosurgery* 51: 1275–1278; discussion 1278-1279, 2002
27) Kiyosue H, Matsumaru Y, Niimi Y, et al.: Angiographic and clinical characteristics of thoracolumbar spinal epidural and dural arteriovenous fistulas. *Stroke* 48: 3215–3222, 2017
28) Takai K, Taniguchi M: Comparative analysis of spinal extradural arteriovenous fistulas with or without intradural venous drainage: a systematic literature review. *Neurosurg Focus* 32: E8, 2012
29) Kiyosue H, Tanoue S, Okahara M, Hori Y, Kashiwagi J, Mori H: Spinal ventral epidural arteriovenous fistulas of the lumbar spine: angioarchitecture and endovascular treatment. *Neuroradiology* 55: 327–336, 2013

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