 doi: 10.2176/jns-nmc.2024-0030

*NMC Case Report Journal 11, 175-179, 2024*

# *Lower Cervical Dural Arteriovenous Fistula with a "Skip Lesion" in the Brainstem: A Case Report*

Yuki YASUOKA,[1] Takafumi MITSUHARA,[2] Shinya NABIKA,[1] Naohiko OHBAYASHI,[1] Asahi SAITO,[1] and Nobutaka HORIE[2]

[1]*Department of Neurosurgery, Matsue Red Cross Hospital, Matsue, Shimane, Japan*
[2]*Department of Neurosurgery, Graduate School of Biomedical and Health Sciences, Hiroshima University, Hiroshima, Hiroshima, Japan*

## Abstract

**Spinal dural arteriovenous fistulas (SDAVFs) are rare vascular malformations that can occur anywhere in the spine. Most SDAVFs lead to slow aggressive myelopathy due to venous congestion at a level adjacent to the shunt point. However, rare cases of localized brainstem edema without spinal cord lesions have been reported. In this study, we present a case of a lower cervical SDAVF that showed localized congestive edema of the medulla in the absence of an edematous change in the cervical spinal cord. The patient was a 57-year-old woman who experienced vertigo and vomiting without myelopathy that did not improve with conservative treatment. Magnetic resonance imaging (MRI) revealed high signal intensity in the left medulla on T2-weighted imaging (T2WI), while angiography revealed an SDAVF at the right C8 segmental level supplied by the right thyrocervical trunk. She underwent surgical interruption of the draining vein, which led to a rapid improvement in her symptoms. A subsequent follow-up MRI confirmed resolution of both the medullary edema and the dilated draining vein. SDAVFs may cause vertigo and vomiting, which are brainstem symptoms. Early diagnosis and surgical intervention are crucial for successful treatment outcomes.**

Keywords: arteriovenous fistulas, arteriovenous shunts, skip lesion

## Introduction

Spinal dural arteriovenous fistulas (SDAVFs) represent a subtype of spinal arteriovenous shunts that occur in individuals in their 50s and 60s. Distinguished by direct connections between the arteries and veins of the dura mater, SDVAFs lack capillary vessels. SDVAFs can occur anywhere along the whole spine, spanning from the cervical to the sacral spine, with thoracolumbar arteriovenous fistulas (AVFs) as the most prevalent type.[1-3] Most SDAVFs are supplied by meningeal branches from the radiculomeningeal artery and drain into a single intradural vein,[4] leading to slow aggressive myelopathy owing to venous congestion at the level adjacent to the shunt point.[5] The common symptoms are sensory and/or motor deficits ascending from the feet and mimicking polyneuropathy or radiculopathy.[6] However, rare cases of localized brainstem edema and skipping of the spinal cord have been reported. In this study, we describe a case of SDAVF in a 57-year-old woman with a shunt point at the C8 segmental level that showed congestive edema of the medulla without edematous changes in the cervical spinal cord. Moreover, we explain the etiology, clinical presentation, and imaging findings based on a literature review.

## Case Report

A 57-year-old woman without remarkable medical history experienced repetitive vertigo and vomiting for a month. There were no other symptoms reported, and both blood tests and neurological examinations yielded unremarkable results. Therefore, she was admitted to the otorhinolaryngology department. Her symptoms did not improve after conservative therapy for a week.

Received February 2, 2024; Accepted April 1, 2024

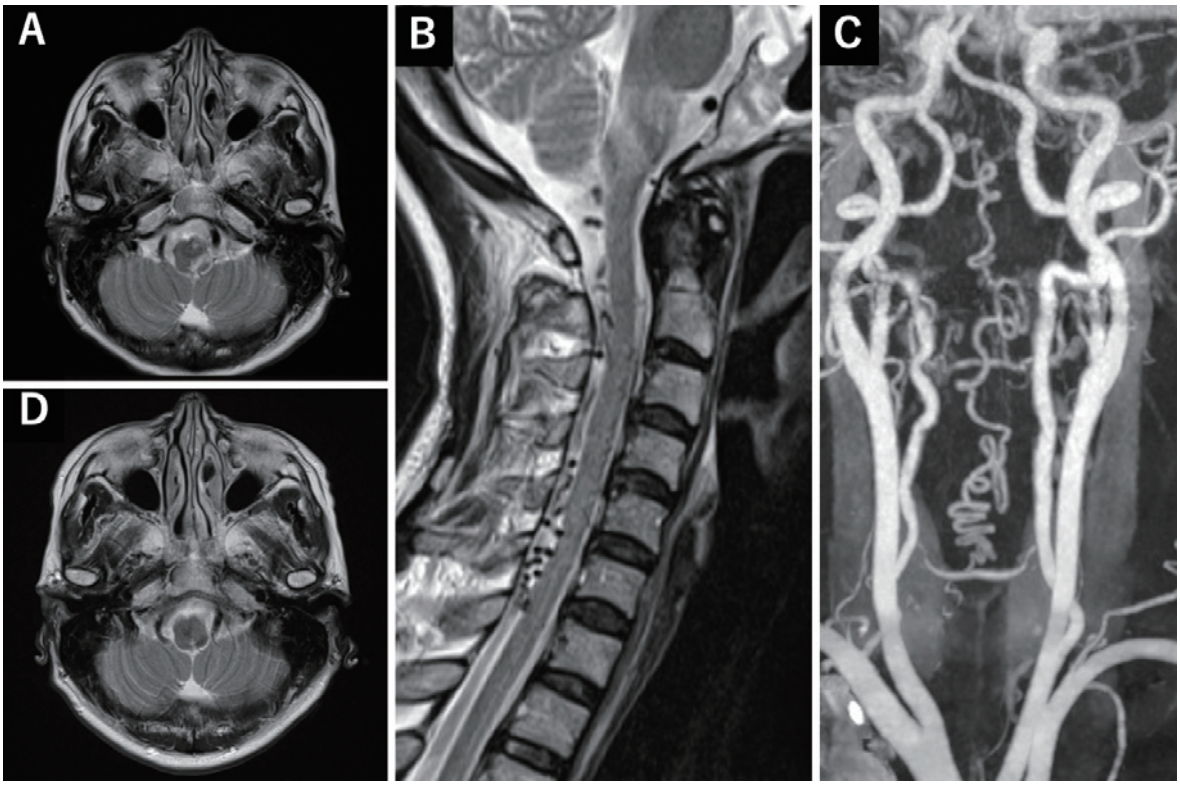


**Fig. 1**
**T2-weighted image (A) shows a hyperintense lesion in the left medulla.**
**Sagittal T2-weighted image (B) shows cervical flow voids and brainstem edema, skipping the spinal cord.**
**Maximum intensity projection image of cervical CT angiography (C) shows a dilated tortuous vein in the cervical region.**
**Follow-up MRI (D) shows that medullary edema resolved 3 months after surgery.**

Magnetic resonance imaging (MRI) of the brain and spine revealed high signal intensity in the left medulla on T2-weighted imaging (T2WI) (Fig. 1A). There were no additional edematous changes in the spinal cord; however, cervical MRI findings showed serpentine T2 flow voids around the spinal cord (Fig. 1B), which were demonstrated as a dilated tortuous vein on a computed tomography (CT) angiogram (Fig. 1C). Subclavian angiography revealed an SDAVF at the right C8 segmental level that was fed by the spinal branch from the right thyrocervical trunk. This SDAVF drained cranially into the ascending intradural vein (Fig. 2A, B).

The patient underwent surgical disconnection of the shunt 5 weeks after symptom onset. The procedure was performed using a C7 hemilaminectomy approach in the prone position in a hybrid operative room equipped with surgical instruments and X-ray fluoroscopy for interventional radiology. The intradural arterialized draining vein was immediately visible and closed with temporary clipping, and intraoperative angiography via the popliteal artery confirmed the disconnection of the arteriovenous shunt. The intracranial vein congestion disappeared after the vessel was coagulated using bipolar forceps. Vertigo and vomiting improved immediately after treatment, and the patient was discharged with no complications on the 18th postoperative day following rehabilitation. A follow-up MRI 3 months after surgery confirmed the resolution of medullary edema (Fig. 1D).

## Discussion

We herein describe a rare case of an SDAVF presenting with vertigo and vomiting due to localized brainstem edema. This study provides important clinical suggestions. Although recent reports have highlighted localized brainstem congestion in craniocervical junction (CCJ) arteriovenous fistulas,[7] there are limited instances of SDAVF where only the brainstem is affected, with the spinal cord near the shunt point remaining unaffected. To our knowledge, only four similar cases, including ours, have been reported in the literature.[8-10] Table 1 summarizes the previous case reports. Cordato *et al.* were the first to report a case of thoracic DAVF with positional vomiting as a brainstem symptom.[9] Shunt levels ranged from the cervical to the upper thoracic spine. In previous cases, the edema extended to the medulla bilaterally, and some patients had immediate gait disturbance.

This imaging finding of a “skip lesion” is related to the

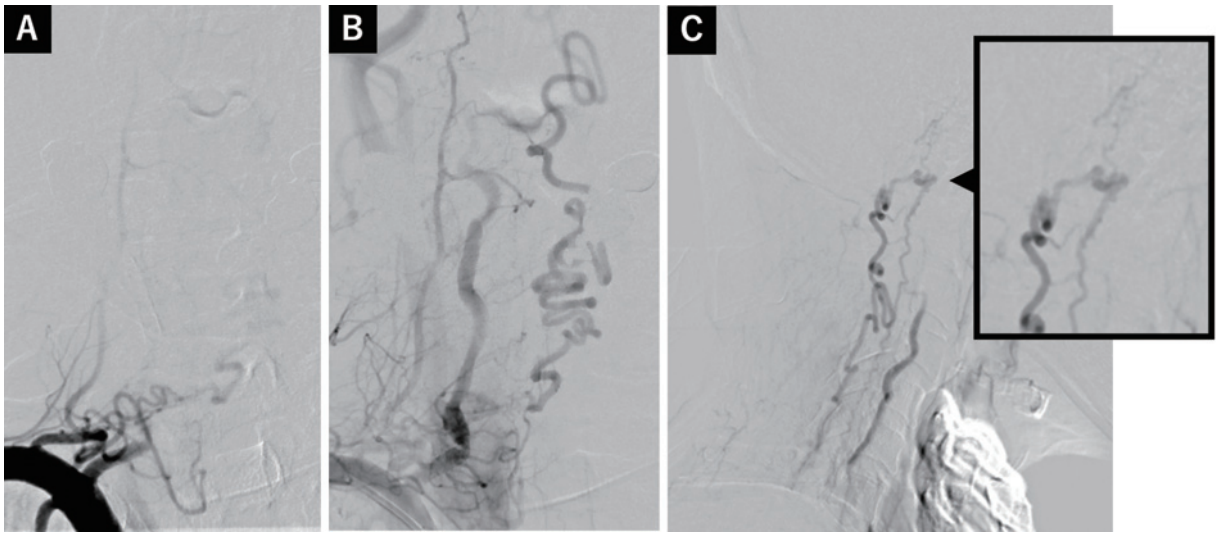


**Fig. 2**
**Preoperative right subclavian angiography (A and B) demonstrating an SDAVF supplied by the right C8 segmental artery. The lateral view (C) shows that the draining vein ascending the dorsal cervical cord turned around the ventral side via the coronal venous plexus at the level of the CCJ, dividing into the median anterior spinal vein and the median anterior medullary vein.**

**Table 1 Previous case reports of SDAVF with isolated brainstem edema**

| Case | Author | Year | Age | Sex | Symptom | Duration (days) | Shunt level | Location of brainstem edema | Treatment | Modified Rankin scale |
|---|---|---|---|---|---|---|---|---|---|---|
| 1 | Cordato | 2004 | 63 | M | Vertigo, vomiting | 21 | T4 | Bilateral medulla | Surgery | 1 |
| 2 | T. Sasaki | 2015 | 56 | M | Vomiting, hiccup, thermal hypoalgesia, gait disturbance | 4 | C4 | Bilateral medulla | Surgery | 1 |
| 3 | K. Sasaki | 2020 | 36 | M | Vomiting, dysphagia, gait disturbance | 4 | T5 | Bilateral medulla | Surgery | 1 |
| 4 | Present case | 2023 | 57 | F | Vertigo, vomiting | 34 | C8 | Left medulla | Surgery | 0 |

SDAVF: Spinal Dural Arteriovenous Fistula

theory that the cervical spinal cord is more resistant to venous congestion than other parts. The incidence of cervical SDAVF is low, accounting for less than 5% of all cases.[6,11,12] In addition, a relatively high prevalence of asymptomatic SDAVF in the cervical region has been reported.[13] The possible mechanisms by which the cervical spinal cord becomes resistant to venous congestion are as follows: First, the characteristic vascular network and anatomy seem to prevent the cervical spinal cord from increasing the venous pressure. Venous return from the cervical spinal cord has multiple epidural venous perfusion routes.[14] Second, the cervical spine has lower venous return resistance from the internal to external venous plexus than the thoracolumbar spine due to the differences in the number of valves.[8,15] Another theory is that the cervical spine is less affected by pulmonary changes in the cerebrospinal fluid (CSF) flow than the thoracolumbar spine.[13] In the present case, these factors may have allowed the intradural draining vein to ascend the dorsal of the cervical cord and reach the intracranial area without causing a venous stasis in the cervical spinal cord.

Furthermore, the symptoms and progression in this patient can be explained by imaging findings and brainstem anatomy. Congestive edema was limited to the left medulla oblongata near the floor of the fourth ventricle, which contains vomiting centers, such as the nucleus ambiguus, solitary nucleus, and reticular formation.[16] According to gadolinium-enhanced T1WI and MR cisternography, the intracranial drainage channel induced congestion in the vein of the left middle cerebellomedullary fissure (Fig. 3A, B). We believe that a lack of venous perfusion on the petrosal surface led to venous stasis in the medulla. Based on lateral angiography, the draining vein ascending to the level of the CCJ turned around the ventral side via the coronal venous plexus, dividing into the median anterior spinal vein and median anterior medullary vein (Fig. 2C). The bifurcation of the drainage pathway may have distributed the shunt blood flow and delayed the progression of

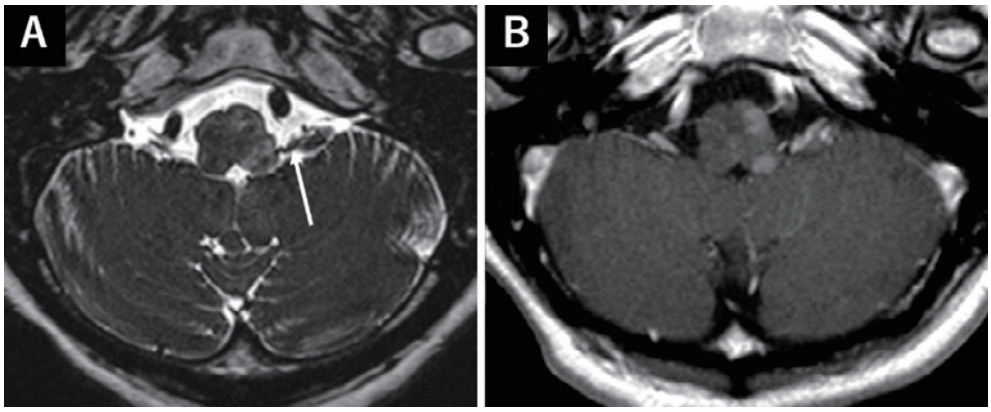


**Fig. 3**
**MR cisternography (A) shows a dilated vein of the left middle cerebellomedullary fissure (*white arrow*) running near the medullary edema.**
**T1-weighted image with gadolinium enhancement (B) shows contrast in the vessel and left medulla.**

intracranial congestion.

This patient's presentation with atypical symptoms underscored the challenge in diagnosing SDAVF promptly. The study emphasizes the need for spinal MRI in patients with brainstem lesions. Sagittal T2WI is used to identify longitudinally extending cervical cord lesions with surrounding flow voids.[1] To ensure an accurate treatment plan, preoperative angiography should be performed to confirm the location of the shunt.[10] It was reported that intraoperative angiography via the transpopliteal approach is useful in patients with spinal vascular disorders.[17] Delayed treatment may result in poor outcomes in progressive AVFs.[18,19] In order for these patients to have a good prognosis, early diagnosis and surgical intervention are essential.

## Conclusion

We herein describe a rare case of a spinal arteriovenous fistula with localized brainstem edema. Reports of similar cases are needed to further clarify the pathophysiology. Early diagnosis and surgical intervention are crucial for successful treatment outcomes.

## Abbreviations

SDAVF: Spinal dural arteriovenous fistulas
MRI: Magnetic resonance imaging
T2WI: T2-weighted images
CSF: Cerebrospinal fluid
T1WI: T1-weighted images
CT: Computed tomography

## Consent for Publication and Institutional Approval

Informed consent for publication was obtained from the patient. Our institutional review board approved this study (no. 581).

## Conflicts of Interest Disclosure

The authors declare no conflicts of interest (COI) associated with this paper. Authors who are members of the Japan Neurosurgical Society have registered online self-reported COI Disclosure Statement Forms.

## References

1) Takai K, Endo T, Seki T, et al.: Congestive myelopathy due to craniocervical junction arteriovenous fistulas mimicking transverse myelitis: A multicenter study on 27 cases. *J Neurol* 270: 1745-1753, 2023
2) Narvid J, Hetts SW, Larsen D, et al.: Spinal dural arteriovenous fistulae: Clinical features and long-term results. *Neurosurgery* 62: 159-166, discussion 166, 2008
3) Tsuruta W, Matsumaru Y, Iihara K, et al.: Clinical characteristics and endovascular treatment for spinal dural arteriovenous fistula in Japan: Japanese registry of neuroendovascular therapy 2 and 3. *Neurol Med Chir (Tokyo)* 59: 492-497, 2019
4) 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
5) Bouardi N El, Lamrani Y, Haloua M, Alami B, Boubbou M, Maaroufi M: A case report of spinal dural arteriovenous fistula: A threatening cause of paraplegia. *Cureus* 14: e31469, 2022
6) Maimon S, Luckman Y, Strauss I: Spinal dural arteriovenous fistula: A review. *Adv Tech Stand Neurosurg* 43: 111-137, 2016
7) Yokoya S, Oka H, Neurosurg T: Isolated brainstem congestion caused by craniocervical junction arteriovenous fistula. *Turk Neurosurg* 33: 514-518, 2023

8) Sasaki K, Inoue T, Nishijima Y, et al.: Thoracic dural arteriovenous fistula presenting with isolated pseudobulbar palsy mimicking brainstem lesion. *World Neurosurg* 136: 157-160, 2020
9) Cordato DJ, Davies MA, Masters LT, et al.: Positional vomiting due to a thoracic spinal dural arteriovenous fistula. *J Neurosurg* 2: 219-222, 2004
10) Sasaki T, Manabe H, Yasuhara T, Miyoshi Y, Sugiu K, Date I: Cervical spinal dural arteriovenous fistula with rapidly progressive brainstem dysfunction due to venous congestion: A case report. *No Shinkei Geka* 43: 51-56, 2015
11) 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
12) Geibprasert S, Pongpech S, Jiarakongmun P, Krings T: Cervical spine dural arteriovenous fistula presenting with congestive myelopathy of the conus: Case report. *J Neurosurg Spine* 11: 427-431, 2009
13) Shimizu K, Takeda M, Mitsuhara T, et al.: Asymptomatic spinal dural arteriovenous fistula: Case series and systematic review. *J Neurosurg Spine* 31: 733-741, 2019
14) Sato K, Endo T, Niizuma K, et al.: Concurrent dural and perimedullary arteriovenous fistulas at the craniocervical junction: Case series with special reference to angioarchitecture; Clinical article. *J Neurosurg* 118: 451-459, 2013
15) Stringer MD, Restieaux M, Fisher AL, Crosado B: The vertebral venous plexuses: The internal veins are muscular and external veins have valves. *Clin Anat* 25: 609-618, 2012
16) Zhong W, Shahbaz O, Teskey G, et al.: Mechanisms of nausea and vomiting: Current knowledge and recent advances in intracellular emetic signaling systems. *Int J Mol Sci* 22: 5797, 2021
17) Maeda Y, Mitsuhara T, Kume S, et al.: Assessment of intraoperative spinal angiography via the popliteal artery for spinal vascular diseases. *World Neurosurg* 169: 1-8, 2023
18) Lee J, Lim YM, Suh DC, Rhim SC, Kim SJ, Kim KK: Clinical presentation, imaging findings, and prognosis of spinal dural arteriovenous fistula. *J Clin Neurosci* 26: 105-109, 2016
19) Sherif C, Gruber A, Bavinzski G, et al.: Long-term outcome of a multidisciplinary concept of spinal dural arteriovenous fistulae treatment. *Neuroradiology* 50: 67-74, 2008

Corresponding author: Takafumi Mitsuhara, MD., Ph.D.
Department of Neurosurgery, Graduate School of Biomedical and Health Sciences, Hiroshima University, 1-2-3 Kasumi, Minami-ku, Hiroshima, Hiroshima 734-8551, Japan.
*e-mail*: mitsuhara@hiroshima-u.ac.jp