An Isolated Unilateral Pontomedullary Lesion Due to An Intracranial Dural Arteriovenous Fistula Mimicking A Brain Tumor - Case and Review

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Intracranial dural arteriovenous fistula (DAVF) with perimedullary venous drainage may cause brainstem swelling and represent a diagnostic challenge.

A 66-year-old man presented to the emergency room with recurrent vertigo, minimal truncal ataxia with a wide-based gait, and a slightly impaired tandem gait. Brain magnetic resonance imaging (MRI) revealed a hyperintense lesion in the left pontomedullary area on T2-weighted images (T2WIs) with partial gadolinium enhancement, but without increased signals on diffusion-weighted images. Abnormal serpentine flow void vessels surrounding the medulla and upper cervical spinal cord were initially overlooked but discovered later. An angiogram revealed DAVF with feeders from the right occipital artery and the meningeal branch of the right distal vertebral artery with drainage into the anterior medullary venous system and the perimedullary veins. The patient underwent a successful transarterial endovascular embolization and improved gradually. A brain MRI at 3-month follow-up revealed a residual hyperintense signal on the T2WIs in the left lower medulla. Six cases of patients exhibiting DAVF with isolated unilateral brainstem swelling from the literature were reviewed.

Isolated unilateral brainstem swelling due to intracranial DAVF with perimedullary venous drainage is extremely rare and might mimic a tumor on MRI. Abnormal serpentine flow void vessels on the surface of the brainstem or spinal cord are crucial diagnostic clues. (J Nippon Med Sch 2019; 86: 48–54)

Key words: brainstem swelling, dural arteriovenous fistula, perimedullary venous drainage, venous congestion

Introduction

Dural arteriovenous fistulas (DAVFs) are abnormal connections between small arterial and venous systems without intervening capillary beds¹. Intracranial DAVF is rare, and is estimated to account for less than 10% of cerebral vascular malformations². The clinical symptoms of intracranial DAVFs are mainly related to the locations of venous drainage, with the most aggressive presentation being intracranial hemorrhage or neurological deficits³. The two most widely used grading systems, according to the complicated venous drainage patterns, were devised by Cognard and Borden⁴. High-grade DAVFs (Cognard Type III-V with venous cortical or perimedullary drainage) have an estimated incidence of aggressive clinical presentation of 89% and an estimated annual risk of hemorrhage of 10%⁵. Intracranial DAVF with perimedullary drainage (Cognard Type V) may cause brainstem dysfunction or cervical myelopathy due to swelling or ischemia of the corresponding structures. However, early diagnosis of DAVF is challenging. The hallmark of dilated perimedullary veins caused by venous congestion in the DAVF usually presents as serpentine flow void lesions around the brainstem or spinal cord. However, they may be too subtle to be observed or even absent in a magnetic resonance imaging (MRI) study. Delayed diagnosis might cause unpredictable complications.

Herein, we present a rare case of intracranial DAVF with perimedullary drainage that presented as recurrent
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**Case Presentation**

A 66-year-old man, without hypertension, diabetes, or a history of head injury, had an acute onset of dizziness, vertigo, and unsteady gait 2 days prior to visiting the emergency room. He had a history of an operation and radiotherapy for small cell lung cancer 11 years previously. He did not complain of any associated nausea, vomiting, double vision, slurred speech, swallowing disturbance, or limb weakness. He had a similar episode of vertigo with unsteady gait and had visited another hospital approximately 1 month previously. A carotid sonography study at that time indicated a mild atheromatous lesion. His symptoms of dizziness and vertigo improved several days later but he still felt a slightly unsteady sensation when walking. A neurological examination on admission revealed truncal ataxia attenuated on the left side, a very mild wide-based gait, and a slightly impaired tandem gait. Brain computed tomography (CT) at the emergency room did not indicate any abnormality (Fig. 1A, B). A brain MRI study revealed a hyperintense lesion at the left lower pons and medulla oblongata on T2-weighted images (T2WIs) without increased signals on diffusion-weighted images (DWIs), but a slightly high value on the apparent diffusion coefficient (ADC) map (Fig. 1C-F). The lesion exhibited partial gadolinium enhancement on T1-weighted images (Fig. 1G). Magnetic resonance angiography (MRA) did not reveal a prominent atherosclerotic change in the intracranial arteries (Fig. 1H). An acute brainstem infarct was excluded from the findings of the MRI study. However, vasogenic edema from an inflammatory, infectious, or neoplastic process was considered due to the similar MRI patterns. The laboratory studies revealed a slightly elevated fasting glucose and low-density lipoprotein, but normal hemogram and antinuclear antibodies. A tumor marker survey based on the patient’s history of lung cancer, including squamous cell carcinoma antigen, tissue polypeptide antigen, carcinoembryonic antigen, cancer antigen 15-3, carbohydrate antigen 19-9, and prostate specific antigen, revealed normal results. A cerebrospinal fluid analysis demonstrated an elevated total protein level of only 100 mg/dL with normal results for cell counts, cryptococcal antigen, and viral immunological values.

A carotid and transcranial duplex sonographic study
revealed a mild atheromatous lesion in the right carotid bifurcation with normal flow velocities, flow resistances, and flow volumes in the bilateral common carotid arteries, internal carotid arteries, external carotid arteries (ECAs), vertebral arteries (VAs), occipital arteries, and posterior auricular arteries. Neither engorgement nor pulsatile retrograde flow was observed in the bilateral superior ophthalmic veins. A brainstem biopsy was considered for a pathological diagnosis. However, a detailed review of the MRI images with radiologists discovered an abnormal serpentine flow void at the ventral surface of the medulla (Fig. 1C) and along the anterior and posterior surfaces of the upper cervical spinal cord (Fig. 1D). A further angiography study revealed DAVF with a main feeder from the right occipital artery and a minor feeder from the meningeal branch of the right distal VA (Fig. 2 A-E). The fistula drained into the anterior and posterior perimedullary veins with reflux into the pontomesencephalic, left transverse pontine, and left anterolateral pontine veins (arrowheads in C), as well as the anterior medullary venous system (arrowheads in C). Anterior (D) and lateral (E) views in a vertebral artery (VA) angiogram, revealing a DAVF with a feeder from the meningeal branch of the right VA (small arrow) that drains into the anterior and posterior perimedullary veins (arrowheads). Oblique view (F) in a right ECA angiogram, revealing the disappearance of DAVF after embolization therapy with the Onyx-18 solution (large arrow).

The patient underwent a transarterial endovascular embolization using a right femoral artery approach with a 6-French Neuron guiding catheter (Penumbra Inc, San Leandro, California, USA) and a microcatheter (Apollo, ev3-Covidien, Irvine CA, USA), under general anesthesia. The embolization of the AVF was performed via the superselective right occipital artery with injection of Onyx-18™ (3 mL; ev3-Covidien, Irvine CA, USA) in 10 min into the feeder branch (Fig. 2F). After embolization, the patient’s symptoms improved gradually and no further vertigo or ataxia occurred. A brain MRA at 3-month follow-up revealed that the serpentine flow void at the medulla and cervical spinal cord had disappeared and that the edematous change of the left lower pons and medulla had considerably improved with a residual hypointense signal on the T2WI at the left lower medulla (Fig. 3).
Follow-up brain magnetic resonance imaging after 3 months revealed the disappearance of the serpentine flow void lesion with a residual hyperintense signal on the T2-weighted images at the left lower medulla.

**Discussion**

Our patient experienced recurrent acute dizziness and vertigo with an unsteady gait during a 1-month period. No other neurological deficit was reported except for subtle cerebellar signs presenting as minimal truncal ataxia and a slightly impaired tandem gait. Although the brain CT study revealed a normal result, a further brain MRI study revealed an edematous lesion at the left lower pons and medulla oblongata. However, the abnormal serpentine flow void vascular structures around the lower brainstem and upper cervical cord were overlooked initially. No other inflammatory, infectious, or neoplastic process was observed after serial studies. Abnormal serpentine flow void vessels discovered during the subsequent careful review of the MRI images raised the suspicion of DAVF. The cerebral angiography confirmed the final diagnosis of intracranial DAVF and the patient received successful endovascular treatment.

Recurrent acute vertigo with unsteady gait may originate from a peripheral or central source. When no or negligible brainstem or cerebellar signs are observed, these sources are difficult to differentiate, particularly when severe vertigo and vomiting interfere with a proper neurological examination during the acute stage. For our patient, a neurological consultation in the emergency room enabled the detection of these subtle cerebellar signs. Otherwise, this patient may have been discharged from the emergency room based on a normal brain CT study, an absence of prominent stroke risk factors, and the partial relief of symptoms after treatment, similar to his episode 1 month previously.

A brain MRI study provides much more information than a brain CT for the study of acute lesions involving the posterior fossa or brainstem. Common causes of the hyperintense lesions on T2WIs in an MRI study include infarction, inflammation, infection, demyelination, tumor, and vasogenic edema. The most common vascular cause for a hyperintense lesion on T2WIs is an acute cerebral infarct, which also causes an increased signal on DWIs but a decreased signal on the ADC map. An acute cerebral infarct was excluded quickly from the MRI results in this patient due to a normal signal on DWIs and a slightly high value on the ADC map. Decision-making for further surveys of unilateral hyperintense lesions on T2WIs is difficult. Noninvasive laboratory studies were unable to identify the possible pathological correlation. Similar to venous sinus thrombosis, DAVF is a critical vascular cause of brain edema from venous congestion. In contrast to cytotoxic edema in an acute infarct with an increased intracellular fluid and a low ADC value, vasogenic edema is associated with an increase of extracellular fluid and might cause a high ADC value in an MRI study. A tumor-mimicking brainstem edematous lesion has been emphasized to distinguish vasogenic edema of the brainstem from DAVF. Careful interpretation of the MRI images for the abnormal serpentine flow void vascular structures is crucial for the recognition of DAVF. However, studies have reported brainstem swelling from DAVF without adjacent abnormal serpentine flow void vascular structures, either due to small shunting or distant spinal DAVF. In Haryu’s review, the presence of perimedullary flow-related signal voids was detected in only 37% of patients with Cognard Type V DAVF. However, Roelz et al suggested that combining contrast-
enhanced MRI and MRA might increase the detection rate to 85%4. A “black butterfly” sign, due to hemorrhage caused by prolonged venous congestion, on T2*- and susceptibility-weighted MRI in the dorsal aspect of the medulla and the central gray matter of the cervical spinal cord has been reported to facilitate the diagnosis of DAVF5.

Intracranial DAVFs have a wide range of presentations, including pulsatile tinnitus, acute subarachnoid hemorrhage, brainstem dysfunction, myelopathy, radiculopathy, neuralgia, cranial palsy, and seizure. Neurological symptoms correlate with the location and severity of venous drainage6,7. Male patients might have more aggressive symptoms, such as hemorrhage due to more frequent cortical venous drainage. The clinical presentation and classification of DAVF are chiefly based on its venous drainage pattern. According to the Cognard classification, the DAVF in our patient was classified as a Type V fistula, which is characterized by perimedullary venous drainage. This type of DAVF may also be classified as craniocervical junction, infratentorial, or posterior fossa DAVF according to its specific location6–11. Progressive myelopathy or brainstem dysfunction occurs due to an edematous or ischemic change in the cervical spine or brainstem caused by venous congestion with high venous pressure in the pontomesencephalic vein, anterior medullary venous system, and perimedullary veins. Typically, the MRI images of patients with Cognard Type V DAVF reveal centrally located medullary or pontomedullary edema, with various degrees of cervical spinal cord involvement6. Isolated brainstem swelling without spinal cord involvement is uncommon12,13. Asri et al reviewed 58 patients who had intracranial DAVF with perimedullary venous drainage (Cognard Type V) from 1988 until 20114. Swelling changes involving both the brainstem and spinal cord, and involving the spinal cord alone were observed in 52% and 44% of patients, respectively. Only 2 patients had isolated brainstem swelling in the MRI studies. Furthermore, isolated unilateral brainstem swelling without spinal cord involvement is extremely rare and more difficult to distinguish from other pathological conditions. Roelz et al also reviewed 58 reported cases of intracranial DAVF with perimedullary spinal venous drainage from 1992 to 2014 and revealed that only 3 patients (7%) had a unilateral brainstem lesion. We conducted a review and discovered 2 more patients with a unilateral brainstem lesion reported by Probst et al in 1994 and by Duan et al in 201712,13. Including the patients mentioned here, only 6 patients with an isolated unilateral brainstem lesion due to DAVF have been reported in studies in the English language literature12,13,14,15. A summary of these 6 cases is presented in Table 1. Nausea, vomiting, and vertigo, which represent a vestibular dysfunction, were the most common initial symptoms in these 6 patients. However, all the patients except for the present patient had other coexisting brainstem signs such as motor deficits, bulbar dysfunction, or Horner syndrome. The ECA and VA were the most common feeders of the DAVF. All patients received endovascular treatment. Two patients received further surgical treatment. All the patients exhibited clinical improvement, and 2 patients had a complete regression of symptoms. In the present case, most shunting flows drained from the right occipital artery into the anterior perimedullary veins with reflux into the pontomesencephalic, left transverse pontine, and the left anterolateral pontine veins. The unstable shunting pressure with hemodynamic stress resulted in focal venous congestion causing isolated edema of the left lower pons and medulla with recurrent brain stem dysfunction symptoms. The reversible edematous left pontine lesion improved after the embolization of the shunting from the right occipital artery and, thereafter, no clinical symptoms were observed.

Carotid duplex sonography has been recommended as the initial screening tool for diagnosis and follow-up in patients exhibiting symptoms related to intracranial DAVF16. The parameter of resistance index of the ECA (cutoff points: right, 0.72; left, 0.71) yielded an 84% accuracy for predicting DAVF. However, small arteriovenous shunting with slow flows may not be detected by using ultrasound. We did not observe any abnormal flows or resistance indexes in the bilateral ECAs, VAs, or even occipital arteries through carotid duplex sonography in our patient. Digital subtraction angiography remains the gold standard for the diagnosis and evaluation of DAVF. A 6-vessel angiography, including bilateral internal carotid arteries, ECAs, and VAs, with appropriate selective arterial angiography, is necessary for a comprehensive survey of such complicated arteriovenous fistulae. An endovascular approach through a super-selective transarterial embolization of the feeders with Onyx has become the mainstay of therapy. Transvenous embolization, surgery, and stereotactic radiosurgery may be adjunctive treatments to transarterial embolization17.

Most of the reported instances of brainstem edema resolved gradually after successful obliteration of the DAVF. A residual brainstem lesion and neurological deficits might be present in patients with brainstem ischemia.
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due to cytotoxic edema. The early recognition and proper treatment of DAVF is crucial to prevent extensive damage and subsequent hemorrhage. For our patient, a residual high-intensity lesion remained at 3-month follow-up in the left lower medulla, which was apparently caused by a small residual shunt from the meningeal branch of the right VA. However, no clinical symptoms were observed. Regular follow-up MRI studies are necessary to assess the development of new symptoms or the progression of residual DAVF.

In conclusion, isolated unilateral brainstem edema without spinal cord involvement due to intracranial DAVF with perimedullary venous drainage is rare and might mimic a brainstem tumor in an MRI study. Abnormal serpentine flow void vascular structures on the surface of the brainstem or spinal cord are crucial diagnostic clues. Early identification and proper treatment of DAVF usually leads to favorable clinical outcomes.

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Conflict of Interest: The authors declare no conflict of interest.

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![Table 1. Reported cases of isolated unilateral brainstem swelling due to intracranial dural arteriovenous fistula](image-url)


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