

Brainstem Venous Congestion Due to Transverse-sigmoid Sinus Dural Arteriovenous Fistula: Case Report and Literature Review

Keiichi TSUJI,¹ Atsushi TSUJI,¹ Yayoi YOSHIMURA,¹ Hiroto KAWANO,¹
Ryo FUJISAWA,¹ and Kazuhiko NOZAKI¹

¹Department of Neurosurgery, Shiga University of Medical Science, Otsu, Shiga, Japan

Abstract

Brainstem venous congestion due to dural arteriovenous fistula (dAVF) can mimic brainstem glioma and infarction. We report a case of a 56-year-old woman with a transverse-sigmoid sinus (TS) dAVF. On MRI, she presented with brainstem edema that was difficult to distinguish from brainstem glioma and infarction. She was referred to our hospital for mild dysarthria with right hemiparesis and a suspected left pontine glioma. On MRI, contrast enhancement of the lesion was demarcated by the pontine raphe, and the ipsilateral vein of Rosenthal was dilated. Cerebral angiography revealed TS dAVF with an isolated sinus. Transarterial followed by transvenous coil embolization was performed to reduce shunt flow, resulting in symptom improvement and normal findings on MRI and cerebral angiography. Brainstem venous congestion due to TS dAVF is as rare as adult brainstem glioma. Differentiating the above-mentioned three diseases on the basis of diagnostic imaging findings and clinical course is necessary for appropriate and timely treatment.

Keywords: dural arteriovenous fistula, venous congestion, endovascular treatment, brainstem glioma

Introduction

Brainstem edema caused by dural arteriovenous fistula (dAVF)-related venous congestion is difficult to distinguish from brainstem glioma¹⁻⁷⁾ and brainstem infarction.^{8,9)} However, it is important to distinguish between brainstem glioma and infarction and dAVF because these three diseases have different treatment methods. Patients with brainstem glioma may be treated with radiation and chemotherapy without resection or biopsy.¹⁰⁾ Here, we report a case of a patient with transverse-sigmoid sinus dAVF (TS dAVF) with an isolated sinus that mimicked brainstem glioma and infarction and review similar previously reported cases.

Case Presentation

A 56-year-old woman lost consciousness and fell while riding a bicycle, and she was transported to

a local emergency hospital. Mild dysarthria and right hemiparesis had gradually progressed since a month and a brainstem glioma was suspected on the basis of head CT and the clinical course. Her medical history was notable for a conservatively treated traumatic acute subdural hematoma 1 year ago. The patient was transferred to our hospital, and examination on admission showed mild symptoms of recent memory disturbance, dysarthria, and right hemiparesis, as well as right cerebellar ataxia. Gadolinium T1-weighted imaging (Gd-T1WI) showed a contrast-enhancing left pontine mass lesion demarcated by the pontine raphe and a dilated vein of Rosenthal on the left. T2-weighted imaging (T2WI) and FLAIR imaging showed a hyperintense region in the medial left temporal lobe that was considered brain edema (Fig. 1). Vascular malformations were considered more likely than brainstem glioma. Therefore, cerebral angiography was performed, which revealed left TS dAVF with an isolated sinus that was supplied by the left middle meningeal artery (MMA) and both occipital arteries (Fig. 2). Retrograde venous flow from the isolated sinus to the superior petrosal sinus, petrosal vein, vein of Rosenthal, right olfactory vein, and vein of the great horizontal fissure

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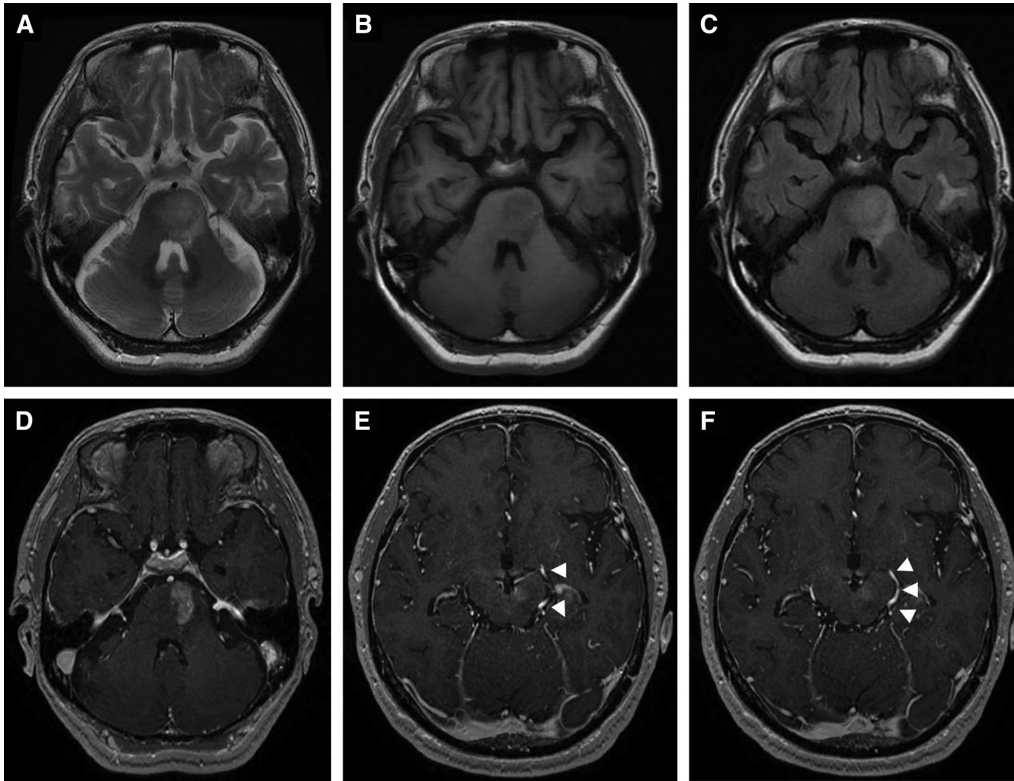


Fig. 1 MRI revealed a left pontine mass exhibiting a high signal intensity on T2-weighted (A) and FLAIR (C) imaging, low intensity on T1-weighted imaging (B), and enhancement demarcated by the pontine raphe on gadolinium T1-weighted imaging (D). T2-weighted and FLAIR imaging showed high signal intensity in the medial left temporal lobe. MRA showed a dilated vein of Rosenthal on the left (arrowheads) (E and F).

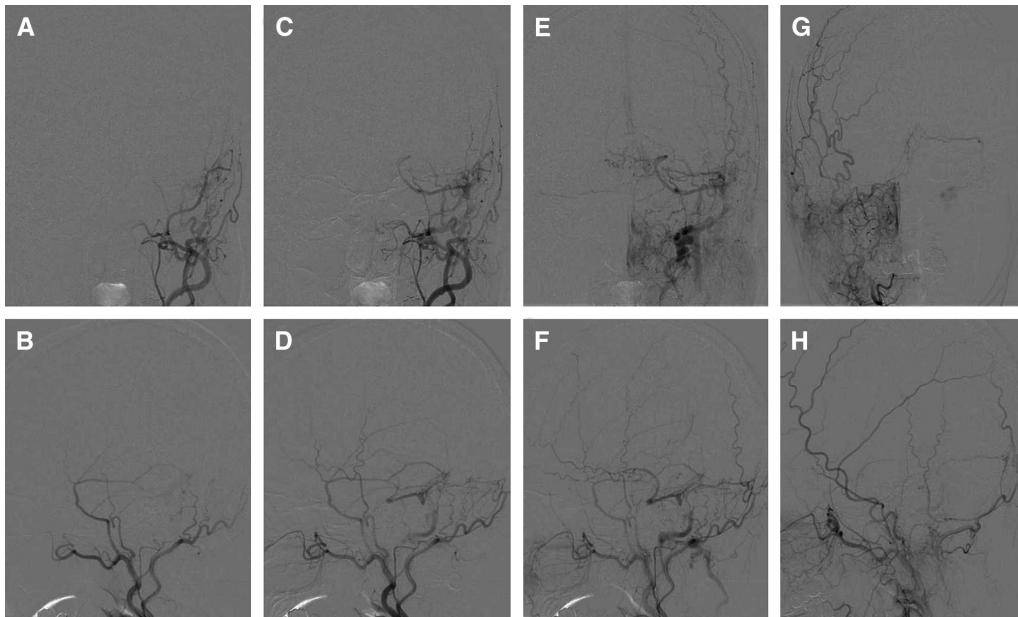


Fig. 2 Anteroposterior (A, C, and E) and lateral (B, D, and F) views of the left external carotid artery show a dural arteriovenous fistula at the transverse-sigmoid junction with an isolated sinus. The posterior branch of the middle meningeal artery and occipital artery were feeding arteries. Retrograde venous flow to the vein of Rosenthal and vein of the great horizontal fissure was confirmed. Anteroposterior (G) and lateral (H) views of the right external carotid artery showed arterial supply from the right occipital artery.

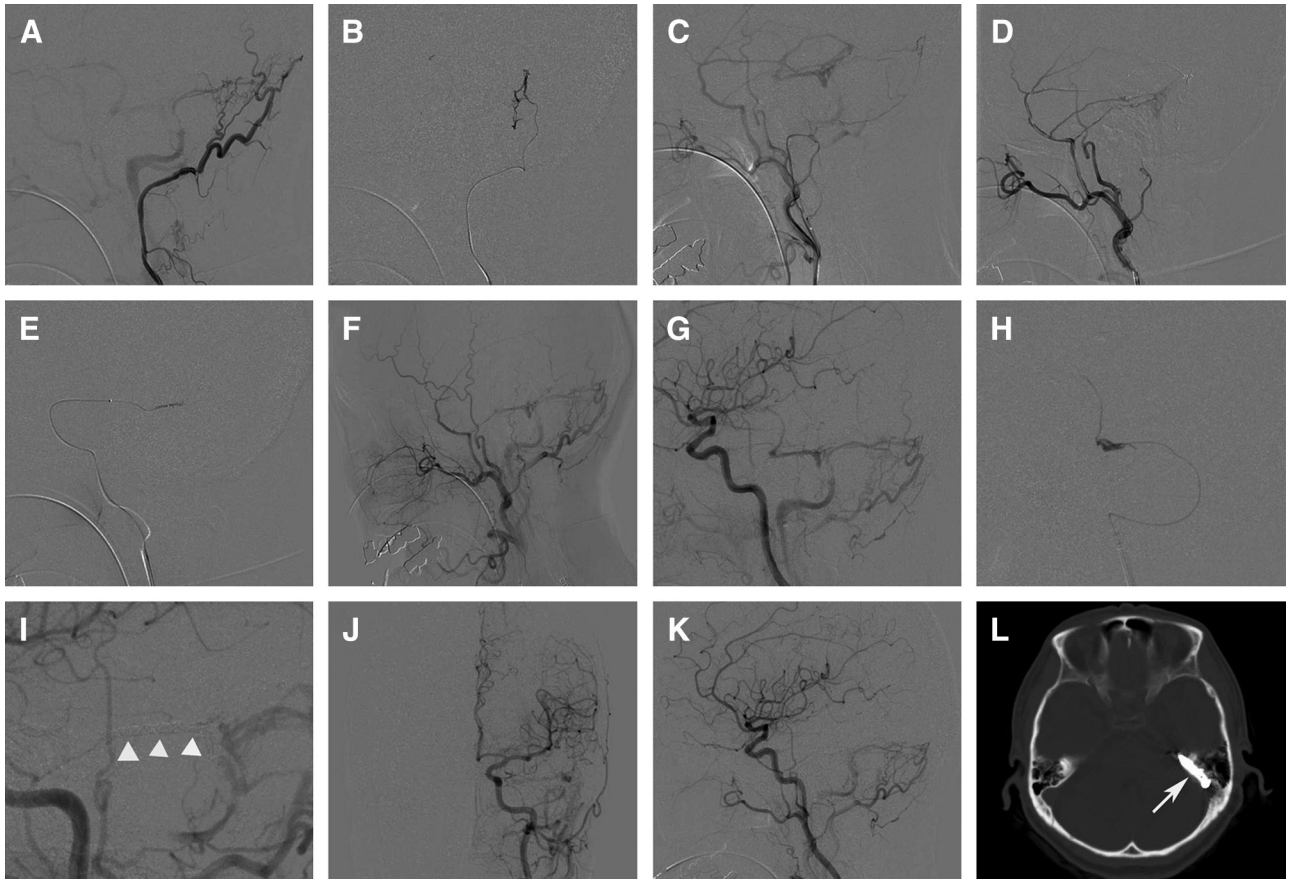


Fig. 3 Embolization of the left occipital (A and B) and left middle meningeal arteries (C–E). Angiography after transarterial embolization (F) and before transvenous embolization (TVE) (G), insertion of microcatheter into the isolated sinus (H), coil embolization of the isolated sinus (arrowheads) (I), angiography after TVE (J and K), and CT showing coils occluding the isolated sinus (arrow) (L).

was present; therefore, the dAVF was classified as Cognard type III.

Transarterial embolization (TAE) was performed under general anesthesia to reduce shunt flow (Fig. 3). A 5-Fr long sheath was inserted in the right femoral artery, after which a 5-Fr guiding catheter (Launcher; Medtronic, Minneapolis, MN, USA) was advanced into the left occipital artery. Then, two microcatheters (Carnelian MARVEL and Carnelian HF; Tokai Medical Products Inc., Kasugai, Aichi, Japan) were coaxially placed in the left posterior meningeal artery using a microguidewire (Traxcess14; Terumo Corp., Tokyo, Japan) and the feeding artery was occluded with detachable coils (Barricade; Balt USA, Irvine, CA, USA) and 33% n-butyl-2-cyanoacrylate (NBCA). Next, the posterior branch of the MMA was occluded using the same catheters and embolic materials. After 3 days, transvenous embolization (TVE) was performed under general anesthesia (Fig. 3). A 4-Fr long sheath was placed in the right radial artery to advance a 4-Fr catheter into the left common carotid artery for

intraoperative angiography. A 6-Fr long sheath was placed in the right femoral vein to advance a Launcher 6-Fr guiding catheter into the left internal jugular vein. After a 4.2-Fr guiding catheter (FUBUKI; ASAHI Intecc, Nagoya, Aichi, Japan) was placed in the left sigmoid sinus for distal access, a microcatheter (SL-10; Stryker, Kalamazoo, MI, USA) was advanced into the isolated sinus with a 0.016-inch microguidewire (Meister; ASAHI Intecc). Fourteen detachable coils were used for embolization from the superior petrosal sinus to the isolated sinus. After the procedure, venous reflux to the intracranial veins disappeared and the patient's neurological symptoms improved. Brain MRI after 3 months showed improvement and shunt flow had completely disappeared on cerebral angiography after 6 months (Fig. 4).

Discussion

DAVF accounts for 10–15% of intracranial arteriovenous malformations.¹¹⁾ Common causes are

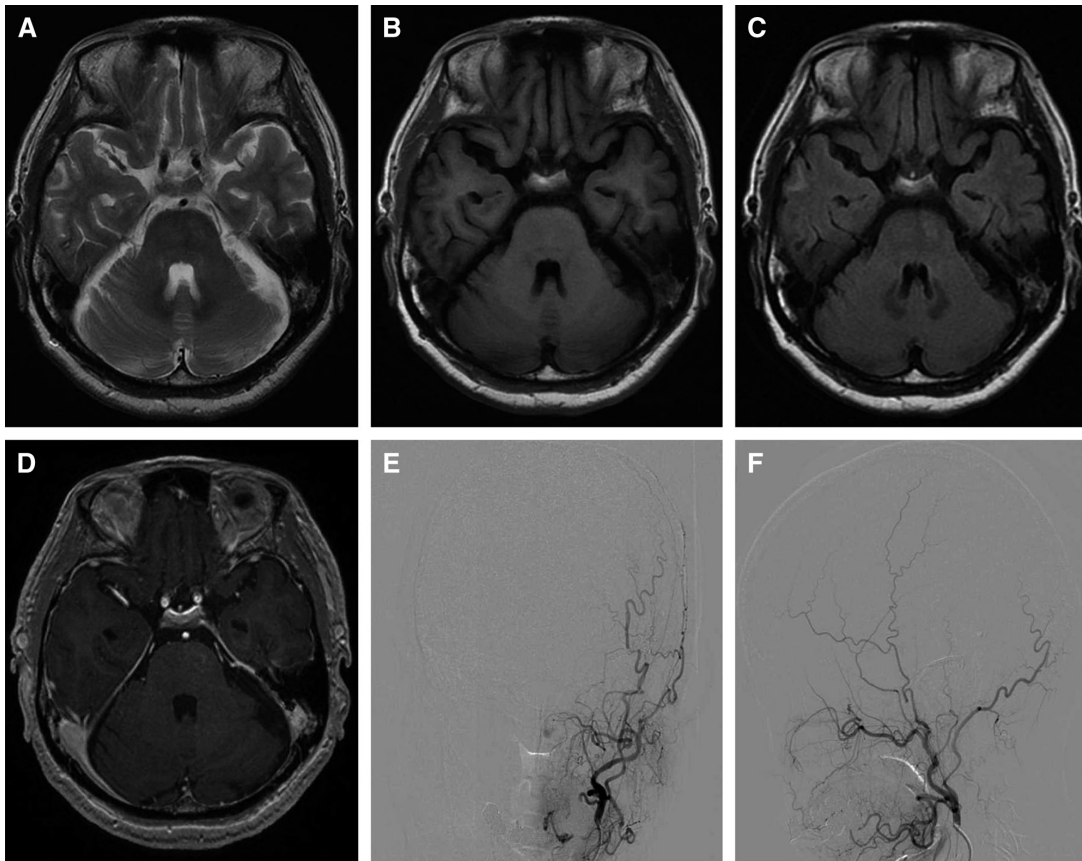


Fig. 4 MRI 3 months after treatment showed normal findings in the left pons (A–D). Cerebral angiography 6 months after treatment showed the disappearance of shunt flow (E and F).

idiopathic, craniotomy, head trauma, and sinus thrombosis.¹²⁾ In Japan, TS is the second most common location for dAVF after the cavernous sinus and the average age of onset is 66 years.¹³⁾ TS dAVFs with an isolated sinus often cause cerebral hemorrhage and acute neurological symptoms.^{14–16)} Our patient presented as an acute case and showed venous congestion in the left pons on MRI because of abnormal venous outflow, which suggested that urgent treatment was necessary. TVE is considered less invasive and more effective in treating dAVFs with an isolated sinus.^{17,18)}

It is important to differentiate between cerebral infarction and brainstem glioma for brainstem venous congestion on head MRI as seen in this case. If sinus thrombosis occurs and intracranial pressure increases, the resulting symptoms resemble those of cerebral infarction may occur in dAVF suddenly.^{8,9)} It is even more important to distinguish between dAVF and brainstem glioma due to the difference in treatment strategy.

Further, dAVF-related brainstem edema mimicking pontine glioma has been reported in 11 patients

(including our case) and is rare (Table 1).^{1–7)} Three patients were male and eight were female, with the average age of 62.5 years (36–80 years). Nine of these cases occurred in the cavernous sinus, one in perimedullary, and one in TS. Endovascular treatment, radiotherapy, sinus packing, or combined treatments were performed and symptoms improved in eight patients. The high signal intensity observed on T2WI is attributed to edema and venous congestion resulting from venous hypertension, whereas the contrast enhancement on Gd-T1WI is attributed to venous congestion and blood–brain barrier breakdown.^{19,20)} In patients showing contrast enhancement in the brainstem, no improvements are observed even after intervention.^{1,3,4)} Therefore, such patients require urgent treatment.

Because biopsy and surgical resection of brainstem gliomas are high-risk procedures,¹⁰⁾ radiation and chemotherapy treatment may be performed without tissue diagnosis. Therefore, the differentiation of brainstem lesions on imaging is important. Brainstem gliomas are relatively more common in children than in adults²¹⁾ and account for less

Table 1 Summary of cases of dural AVF with MRI findings mimicking brainstem glioma

Author	Case	Location of d-AVF	Location of congestion	Drainage route	Symptoms	Treatment	Prognosis
Uchino et al. (1997) ¹⁾	68.F	Cavernous sinus	Pons	Basal v., vein of Labbe	Ophthalmic pain, ophthalmoplegia, pyramidal tract sign, cranial nerve palsy, ataxia	TAE	No change
	74.M	Cavernous sinus	Pons, Rt. cerebellum	Cortical veins of posterior fossa	Chemosis, proptosis, oculomotor palsy, cerebellar ataxia	TAE, radiotherapy	Improved
Takahashi et al. (1999) ²⁾	49.M	Cavernous sinus	Pons	SPS, SOV, IOV	Exophthalmos, chemosis, diplopia	TVE	Improved
	62.F	Cavernous sinus	Pons	Bilateral SOV	Exophthalmos, chemosis, loss of visual acuity	TVE	Improved
Shintani et al. (2000) ³⁾	65.F	Cavernous sinus	Pons	IPS	Chemosis, diplopia, gaze palsy, cerebellar ataxia	None	Dead
Kai et al. (2004) ⁴⁾	56.F	Cavernous sinus	Pons	Petrosal v.	Proptosis, double vision, visual impairment, hemiparesis	Sinus packing (craniotomy)	Remaining symptoms
	70.F	Cavernous sinus	Midbrain	Deep Sylvian v., pontomesencephalic v.	Double vision, chemosis, exophthalmos, mild truncal ataxia	Sinus packing (craniotomy)	Improved
Iwasaki et al. (2006) ⁵⁾	71.F	Cavernous sinus	Upper pons	SPS, anterior pontomesencephalic v.	Abducens nerve palsy	Stereotactic radiosurgery	Improved
Miyagishima et al. (2012) ⁶⁾	80.F	Cavernous sinus	Upper pons	SOV, petrosal v.	Chemosis, exophthalmos, ataxia, dysarthria	TAE, radiotherapy	Improved
Le Guennec et al. (2015) ⁷⁾	36.M	Perimedullary	Medulla oblongata	Spinal perimedullary v.	Headache, right hemifacial and lingual hypoesthesia, nausea, vomiting	TAE	Improved
Present case	56.F	Transverse-sigmoid sinus	Pons	Petrosal v.	Memory loss, dysarthria, hemiparesis, ataxia	TAE, TVE	Improved

AVF: arteriovenous fistula, IOV: inferior orbital vein, IPS: inferior petrosal sinus, SOV: superior orbital vein, SPS: superior petrosal sinus, TAE: transarterial embolization, TVE: transvenous embolization.

than 2% of adult brain tumors.²²⁾ In adults, the average age of diagnosis is 34–40 years, the pons is frequently involved, and diplopia, hemiplegia, and headache are the primary symptoms.^{10,23)} The median symptom duration is 4 months; however, sudden onset of symptoms like stroke may occur when intratumoral hemorrhage occurs.²³⁾ The most common finding on MRI for the tumor is non-enhancing diffusely infiltrative mass (50% of cases), followed by contrast-enhancing localized mass (31% of cases).²³⁾ In contrast, in patients with dAVF, the average age of diagnosis is 65 years,¹³⁾

and symptoms depend on the location of the lesion. Ocular symptoms, such as chemosis, exophthalmos, and hyperemia, are predominant in cavernous sinus dAVF,^{24,25)} whereas pulsatile tinnitus is observed in TS dAVF.²⁶⁾ Although dAVFs generally resemble brainstem gliomas on MRI, the enhancement pattern is demarcated by the pontine raphe.^{2,3)} Furthermore, dAVFs may be associated with supraorbital vein dilatation, the presence of abnormal vascular networks near the cavernous sinus, and the dilatation of veins surrounding the brainstem on MRA.^{2,6,7)}

Our patient's symptoms did not suggest dAVF; therefore, brainstem glioma and cerebral infarction were initially suspected on the basis of imaging findings and the clinical course. In this case, dAVF was considered in the differential diagnosis because of the contrast enhancement pattern and dilated vein of Rosenthal on Gd-T1WI. However, from the perspective of treatment selection, it is important to keep the brainstem glioma in mind in the differential diagnosis of cases such as imaging findings.

Conclusion

TS dAVF can be difficult to distinguish from brainstem glioma and infarction on MRI. Differentiating these three is necessary for appropriate and timely treatment.

Conflicts of Interest Disclosure

All authors report no conflicts of interest about this article.

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Corresponding author: Keiichi Tsuji, MD
Department of Neurosurgery, Shiga University of
Medical Science, Setatsukinowacho, Otsu, Shiga
520-2192, Japan.
e-mail: tsujikei@belle.shiga med.ac.jp