Radiology Case Reports 16 (2021) 3499–3503

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Case report

Early rebleeding of a foramen magnum dural arteriovenous fistula: A case report and review of the literature ☆,☆☆

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ARTICLE INFO

Article history:
Received 8 August 2021
Accepted 13 August 2021

Keywords:
Foramen magnum dural arteriovenous fistula
Craniocervical junction arteriovenous fistula
Early rebleeding
Ascending pharyngeal artery

ABSTRACT

Foramen magnum dural arteriovenous fistula (FM-DAVF) is a subset of craniocervical junction arteriovenous fistulas. We report a rare case of FM-DAVF with early rebleeding and review the literature. A 50-year-old man experienced 3 episodes of intracranial bleeding from a vessel malformation in the acute stage. We identified an FM-DAVF, supplied by multiple feeding arteries (eg, left ascending pharyngeal artery) that drained into the straight sinus and left superior petrosal sinus. The draining vein had venous varices. We performed transarterial feeder embolization and surgical disconnection of the DAVF. Early rebleeding of FM-DAVF is rare. High-risk patients require risk assessment and appropriate treatment as soon as possible in the acute stage.

Introduction

Foramen magnum dural arteriovenous fistula (FM-DAVF) is a subset of craniocervical junction arteriovenous fistula (CCJ-AVF) with the fistulous point at the foramen magnum. Few relevant reports exist regarding FM-DAVF [1]. CCJ-AVF is rare vascular malformation that may result in subarachnoid hemorrhage (SAH), congestive myelopathy, and brainstem dysfunction [2]. Thus, CCJ-AVF has various clinical features

Abbreviations: AVM, arteriovenous malformation; CT, computed tomography; DAVF, dural arteriovenous fistula; DSA, digital subtraction angiography; GKS, gamma knife surgery; ICG, indocyanine green; MRI, magnetic resonance imaging; PICA, posterior inferior cerebellar artery; SAH, subarachnoid hemorrhage.

☆ Acknowledgments: The authors received no funding for this work.
☆☆ Competing interests: The authors have declared that no competing interests exist.

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https://doi.org/10.1016/j.radcr.2021.08.038

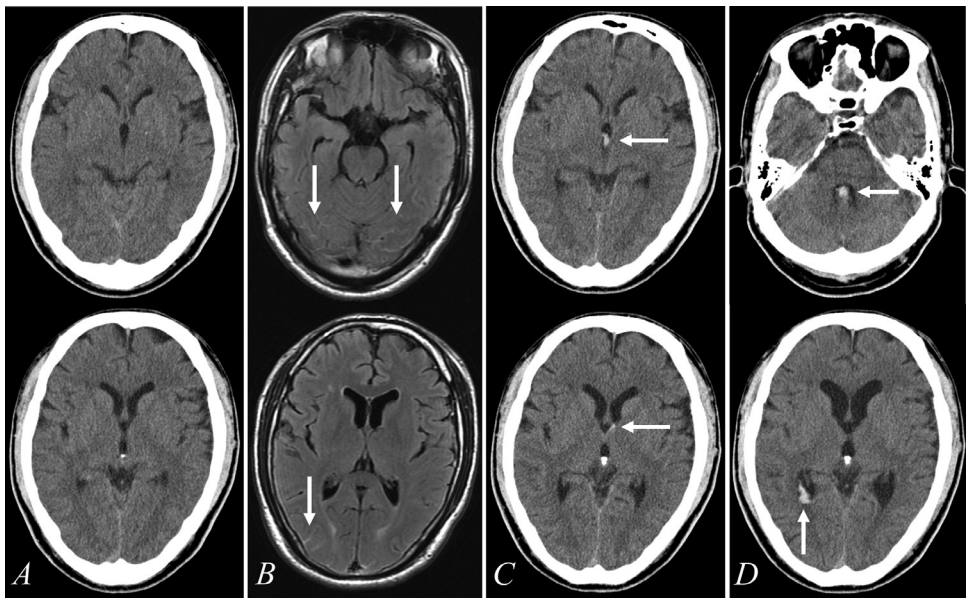

Fig. 1 – Initial computed tomography (CT) displays normal findings (A). The same-day magnetic resonance imaging (MRI) shows a subarachnoid hemorrhage in the bilateral occipital lobe (B, arrows). The next-day CT reveals bleeding, primarily in the third ventricle (C, arrows). 12 days after the initial bleeding event, CT reveals a third intraventricular bleeding event in the fourth ventricle and the lateral ventricle with intracranial ventriculomegaly (D, arrows)

and a spectrum of neuroradiological findings. Diagnosis and appropriate therapy of CCJ-AVF may be delayed because of its low incidence and complex symptomatology [3,4]. However, the rebleeding rate of CCJ-AVFs, including FM-DAVF, is generally very low. Therefore, in patients with SAH, surgery would be performed in the chronic stage [5].

FM-DAVF presenting with rebleeding in the acute stage and a progressive course is rare. To the best of our knowledge, no clinical reports exist on this condition. In this paper, we report a rare case of FM-DAVF manifesting as early rebleeding. We also review the literature and discuss the clinical features of FM-DAVF.

Case presentation

A 50-year-old man, who had a medical history of hypertension and hyperlipidemia, suddenly complained of posterior cervical pain. 6 days after the onset of the initial symptoms, he consulted with his previous physicians and had no apparent neurological deficits. Computed tomography (CT) findings were normal (Fig. 1A), although magnetic resonance imaging revealed a SAH on the bilateral occipital lobes (Fig. 1B, arrows). The next day, his headache worsened. CT revealed bleeding, primarily in the third ventricle (Fig. 1C, arrows). Three-dimensional CT angiography and digital subtraction angiography revealed a vascular malformation. Gamma knife radiosurgery (GKS) (maximum dose, 36 Gy; marginal dose, 18 Gy) was performed to diagnose re-rupture of the posterior fossa arteriovenous malformation (AVM) with arterial supply from the left posterior inferior cerebellar artery. 12 days after the initial bleeding event (ie, 4 days after GKS), severe posterior cervical pain recurred. CT imaging revealed intraventricular rebleeding with ventriculomegaly of the fourth ventricle and the lateral ventricle, which indicated a third bleeding event (Fig. 1D, arrows). The patient was transferred to our hospital for consecutive treatment.

A detailed review of the previous 3-dimensional CT angiography images revealed vascular anomalies fed by meningeal branches around the foramen magnum. A possible DAVF was identified. Re-examination of selective digital subtraction angiography revealed a FM-DAVF fed by the jugular branch of the left occipital artery, the jugular branch and hypoglossal branch of the left ascending pharyngeal artery, the jugular branch and mastoid branch of the right occipital artery, and the posterior meningeal artery from the left vertebral artery. The DAVF drained into a vein of the lateral recess of the fourth ventricle through an arteriovenous shunt on the left lateral margin of the foramen magnum. The draining vein, which was dilated and tortuous with large varices, entered the straight sinus and the left superior petrosal sinus (Figs. 2A-D). No arterial supply existed from the left posterior inferior cerebellar artery or pial feeder.

28 days after the initial bleeding event, he experienced a deterioration in the level of consciousness with a Glasgow Coma Scale score of 10 (E2V3M5). CT imaging revealed severe hydrocephalus due to a radical increase in the size of the varix (Fig. 2E, white arrow and white arrowheads; Fig. 3F, black arrow). Transarterial feeder embolization was immediately performed, after he underwent external ventricular drainage (Fig. 2G).

2 days after the feeder embolization, direct surgical interruption of the draining vein was performed by using a 11-

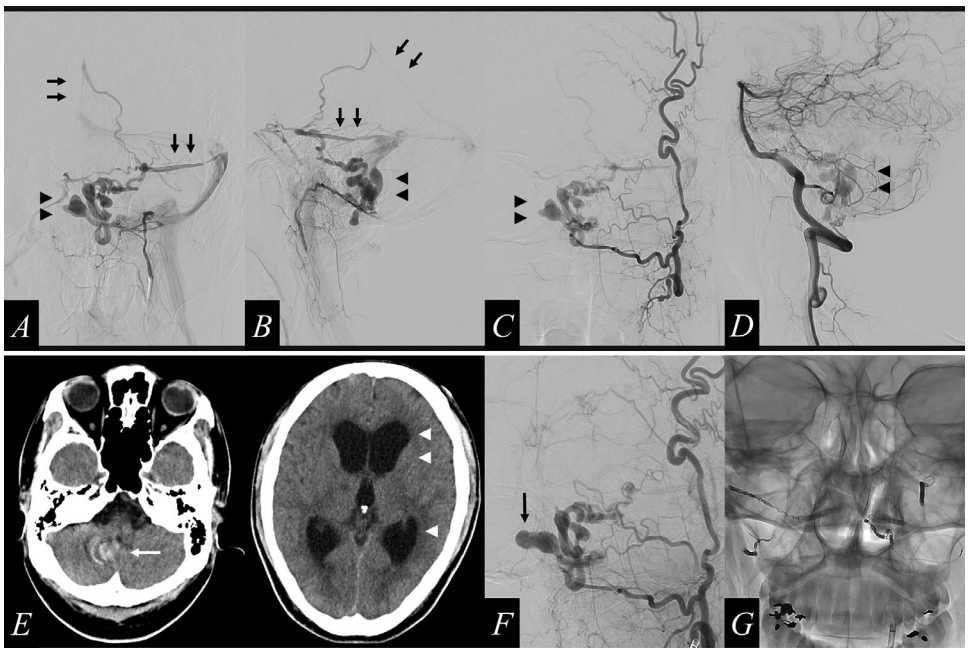

Fig. 2 – Digital subtraction angiography (DSA) shows a dural arteriovenous fistula (DAVF), which is fed by the jugular branch and hypoglossal branch of the left ascending pharyngeal artery (A and B), the jugular branch of the left occipital artery (C), and the posterior meningeal artery from the left vertebral artery (D) that drains into the straight sinus and the left superior petrosal sinus (A, B; arrows) through the vein of the lateral recess of the fourth ventricle, which is dilated and tortuous with large varices (A-D, arrowheads). 28 days after the initial bleeding event, the CT image shows a radical increase in the size of the varix (E, white arrow) and severe ventriculomegaly (E, white arrowheads). DSA imaging shows a radical increase in the size of the varix (F, black arrow), compared to its previous size. We performed transarterial coil embolization on the main feeders (G)

mm straight clip. Complete obliteration was achieved (Fig. 3). After he underwent an additional surgery for hydrocephalus, he was transferred to a rehabilitation facility because of mild weakness of the bilateral lower extremities. He returned home without neurological deficits.

At 6 months postsurgery, follow-up angiographic examinations showed no recurrence of the DAVF. No recurrence of intracranial hemorrhage has occurred during 2 years of follow up.

Discussion

CCJ-AVF occurs in 1%-2% of patients with an intracranial or spinal arteriovenous fistula [6]. CCJ-AVF may result in SAH and congestive myelopathy. A diagnosis of CCJ-AVF may be difficult because of its low incidence and variable clinical features [1,4].

FM-DAVF is a subset of CCJ-AVF. However, FM-DAVF has not been clearly defined. A CCJ-AVF can occur anywhere from the foramen magnum to the high cervical spine. Hiramatsu et al. [6] proposed the disease concept of “radiculomeningeal AVF,” which develops along the C-1 or C-2 nerve roots and mostly includes radiculomeningeal arteries from the vertebral artery as the feeding arteries. By contrast, previous reports [3] reveal that a FM-DAVF is primarily fed by the ascending pharyngeal arteries. This finding is consistent with the fact that a part of the foramen magnum is phylogenetically regarded as a somite, which the ascending pharyngeal artery may supply [7].

We searched relevant published articles regarding FM-DAVF with SAH. We found overall 11 corresponding cases among 5 articles (Table 1) [1,3–5,8]. 5 patients had an arterial supply from the ascending pharyngeal artery. The other cases included the vertebral artery or occipital artery. Zhao et al. [9] report that most FM-DAVFs were supplied by the meningeal branches of the vertebral artery or by the occipital artery and ascending pharyngeal artery. The 3 arteries have a complicated anastomosis with each other at the dura mater around the foramen magnum [10].

In our patient, determining a correct diagnosis and the appropriate treatment took time, owing to misidentifying the venous varix as the nidus of the AVM. The FM-DAVF with its numerous dilated veins and a large varix mimicked an AVM, although the meningeal branches, including the branches of the ascending pharyngeal artery, provided a clue to the diagnosis of the DAVF. Arterial supply from meningeal branches associated with the ascending pharyngeal artery may aid in making a correct diagnosis of FM-DAVF.

Only a few reports exist regarding early rebleeding of a CCJ-AVF. Zhong et al. [11] report that a recurrence of SAH occurred in 3 (8.3%) patients with CCJ-AVF with SAH before surgical treatment and occurred in one of these patients during the

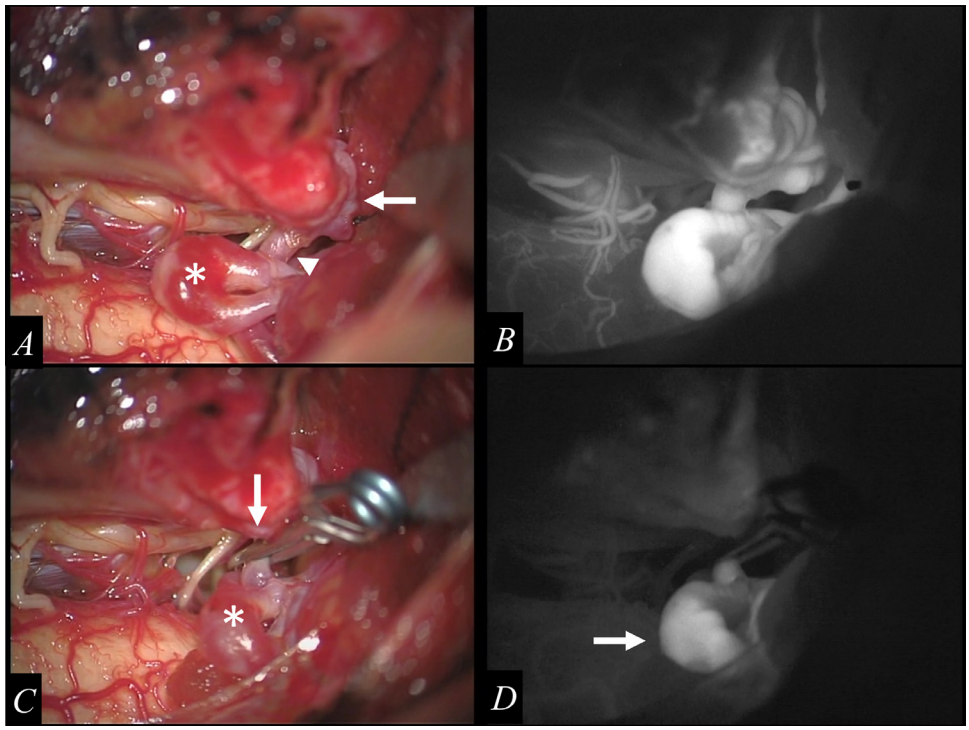

Fig. 3 – The vascular mass suspected of being a "shunted pouch" is confirmed inside of the dura of the left lateral margin of the foramen magnum (A, arrows), which is connected to vein of the lateral recess of the fourth ventricle through a single drainer (A, arrowheads). The inferior portion of the varix is identified (A, asterisk). Intraoperative indocyanine green (ICG) angiography shows retrograde flow to the varix through the single drainer in the early arterial phase (B). The single drainer is obliterated as close as possible to the shunted pouch by using an 11-mm straight clip (C, arrow). The surface of the varix is darker and the variceal wall tension is reduced (C, asterisk). ICG angiography shows stagnation of blood flow in the varix (D, arrow) after the occlusion of the draining vein.

Table 1 – Summary of reports in the literature on FM-DAVF presenting with subarachnoid hemorrhage.

Reference	Age/Sex	Presentation	Feeder	Drainer	Treatment
Kinouchi et al. 1998 [5]	65/M	SAH	VA	MV	Direct surgery
	68/M	SAH	VA	MV	Direct surgery
Guo et al. 2010 [8]	47/M	SAH	VA	MV	Direct surgery
	51/M	SAH	VA	MV	Direct surgery
	35/M	SAH	OA, APA	MV, COS	Direct surgery
	40/M	SAH	OA	MV, StS, COS	Without surgery
Nakamura et al. 2017 [4]	69/F	SAH	VA	MV, IPS	Direct surgery
Motebejane and Choi 2017 [3]	53/M	SAH	APA	MV	IVR with NBCA
	42/M	SAH	APA, VA	MV	IVR with NBCA
	51/M	SAH	APA, VA	MV, JB	IVR with NBCA
Kim et al. 2018 [1]	48/M	SAH	APA	VP, SS	IVR with Onyx

APA, ascending pharyngeal artery; COS, confluence of sinuses; F, female; FM-DAVF, foramen magnum dural arteriovenous fistula; IPS, inferior petrosal sinus; IVR, interventional radiology; JB, jugular bulb; M, male; MV, medullary vein; NBCA, n-butyl cyanoacrylate glue; OA, occipital artery; SAH, subarachnoid hemorrhage; SS, sigmoid sinus; StS, straight sinus; VA, vertebral artery; VP, venous plexus.

acute stage (ie, 14 days after the initial SAH) and occurred in the other 2 patients 28 days later and 4 years later, respectively. However, no report exists of 2 occurrences of rebleeding in the acute stage, as in our patient, and no report exists concerning especially the rebleeding risk of FM-DAVF. Duffau et al. [12] report that intracranial DAVFs with retrograde cortical venous drainage present a high risk of early rebleeding. In addition, Brown et al. [13] report that lesions of petrosal sinus and straight sinus, a venous varix on the draining vein, and lesions draining into leptomeningeal veins increase the risk of intracranial hemorrhage. These features may also be a risk factor for early rebleeding from CCJ-AVFs or FM-DAVFs. GKS was performed because of the misdiagnosis of the DAVF as an AVM in our patient; however, no significant relation-

ship existed between the third bleeding and GKS. In 1 study [14] investigating changes in an animal model of arteriovenous fistula treated with GKS, the researchers concluded that GKS produced morphological, angiographic, and histological changes in the arteriovenous fistula model as early as 6 weeks after treatment.

Table 1 shows that only 1 patient had venous drainage into the straight sinus, although all patients had superior direct drainage into the intracranial venous systems. The aforementioned patient had a high-flow shunt and venous varices, as did our patient. For a FM-DAVF that includes a high-flow shunt and venous varices, venous drainage into the deep cerebral veins may be rare, which is compatible with the low rebleeding rate. However, a significant number of high-risk patients present with an aggressive clinical course. In our patient, the FM-DAVF drained into the petrosal sinus and straight sinus, and it had venous varices on the draining vein with a high-flow shunt. A FM-DAVF that has venous varices and venous drainage into deep cerebral veins is conceivably associated with a higher risk of bleeding, similar to that of an intracranial DAVF involving the aforementioned risk factors of bleeding.

In general, FM-DAVF patients with SAH can be treated surgically in the chronic stage. By contrast, some patients have an aggressive clinical course such as rebleeding or a radical increase in size of the varix. In these situations, administering curative treatments as soon as possible is desirable [10].

Conclusion

FM-DAVF is a rare disease that is sometimes difficult to distinguish from an AVM. Meningeal branches associated with the ascending pharyngeal artery may aid in making a correct diagnosis. The rebleeding rate of FM-DAVF has been considered as very low. However, high-risk patients exist and would require a risk assessment and appropriate treatment as soon as possible in the acute stage.

Patient consent

The patient provided informed consent for treatment and consent for his data to be published in this report. This study was conducted in line with the principles of the Declaration of Helsinki. This is an observational study. The institution review board of Kyoto Prefectural University Graduate School of Medicine (Kyoto, Japan) has confirmed that no ethical approval is required.

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