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Coexistence of a Dural Arteriovenous Fistula and Pial Arteriovenous Malformation Sharing a Common Drainer

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Abstract

In cases of a dural arteriovenous fistula (AVF) with a pial arterial supply, postoperative hemorrhagic complications occur frequently. Six cases in which patients were diagnosed with a coexisting dural AVF and pial arteriovenous malformation (AVM) sharing a common drainer are presented. These cases were initially thought to be dural AVFs with pial arterial supplies, but careful examination of preoperative images showed that a pial AVM coexisted near the dural AVF, and that both shared a common drainer. The coexistence of a pial AVM is difficult to notice during surgery; for this reason, determining the presence of a pial AVM on preoperative imaging is essential to safely treat a dural AVF with a pial arterial supply. The details of each case, specifically, the diagnostic evidence for this condition (coexisting dural AVF and pial AVM sharing a common drainer), as well as imaging findings that should be noted, are presented.

Keywords: arteriovenous malformation, dural arteriovenous fistula, common drainer

Introduction

Among intracranial dural arteriovenous fistulas (AVFs), a dural AVF with a pial arterial supply, supplied not only by the meningeal artery but also by the pial artery, occurs frequently, accounting for 11.3% of all dural AVFs.[1)] This pathology is generally recognized as having the anastomosis between pial arteries and dural arteries, supplying the abnormal shunt in the dura mater. A dural AVF with a pial arterial supply, compared with a dural AVF without a pial arterial supply, has a significantly greater incidence of intracerebral hemorrhagic complications after endovascular treatment (33.3% vs. 2.1%).[2)] Hemorrhagic complications may arise due to insufficient embolization of the shunt or coexistence of a different arteriovenous shunt that could not be identified preoperatively, but the causes are not completely understood.[1,2)] After careful examination of preoperative images, six dural AVFs in which a pial arteriovenous malformation (AVM) coexisted near the dural AVF, sharing a common drainer, were diagnosed (Fig. 1). These cases seemed like a dural AVF with a pial arterial supply on first glance, and were therefore called "coexisting dural AVF and pial AVM sharing a common drainer." Recently, endovascular treatment has been commonly used to treat dural AVFs safely.[3–5)] However, there is a high risk of postoperative hemorrhagic complications if treated without noticing the coexisting pial AVM. Therefore, awareness of this condition is extremely important when determining the treatment strategy for a dural AVF with a pial arterial supply. The details of the six cases, especially the diagnostic evidence and the notable imaging findings, are presented.

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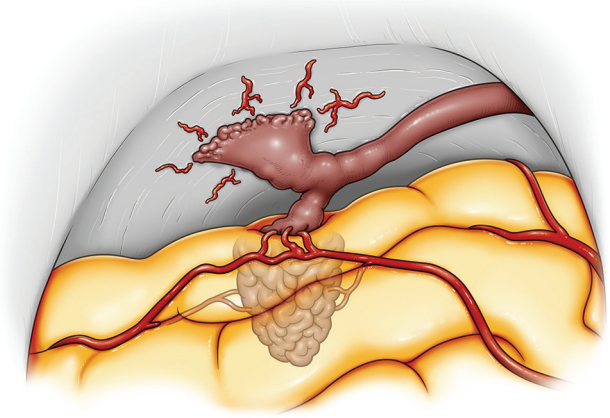

Fig. 1 Schema of the coexistence of a dural AVF and pial AVM sharing a common drainer. AVF: arteriovenous fistula, AVM: arteriovenous malformation.

Case Report

In total, 252 patients with a dural AVF underwent endovascular treatment or craniotomy between January 2010 and June 2020 at our hospital and an affiliated institution; six (2%) were diagnosed with a coexisting dural AVF and pial AVM sharing a common drainer (Figs. 2–4). Table 1 summarizes these patients. The age of the patients ranged from 48 to 75 years, and all patients were men. Three had bleeding while the remaining three had no symptoms. The dural AVF was located in the tentorial sinus (n = 3), convexity (n = 2), and transverse sinus (n = 1). All dural AVFs were Type III, according to the Borden classification. One patient also had transverse-sigmoid thrombosis, although the remaining five patients had no history of other intracranial comorbidities. All patients underwent craniotomy, and two had intracerebral hemorrhage likely due to postoperative bleeding from residual AVM. Informed consent was obtained from the patients for publication of these case reports with accompanying images.

Illustrative cases

Case 1 (Fig. 2): A 52-year-old man with a history of hypertension visited a local physician because of headache. Head computed tomography (CT) and magnetic resonance imaging (MRI) showed a left cerebellar hemorrhage from the left tentorial dural AVF (Fig. 2A), and the patient was referred to our institution for treatment. The tentorial dural AVF supplied mainly by the tentorial artery, middle meningeal artery (MMA), and occipital artery (OA) were observed on cerebral angiography (Fig. 2B and 2C). A single drainer of the dural AVF drained into the straight sinus through the superior vermian vein. On vertebral artery (VA) angiography, development of the pial arteries of the posterior inferior cerebellar artery (PICA) and the superior cerebellar artery (SCA) as feeders were observed. Since they appeared to aggregate in the brain, the coexistence of a pial AVM was suspected (Fig. 2D). To examine the shunt structure in detail, fusion images of the left internal carotid artery (ICA) angiography and left VA angiography were created using the included imaging analysis software syngo X Workplace (SIEMENS, Munich, Germany). The fusion images showed that the pial artery from the PICA and SCA gathered in the brain, formed a nidus, and became a separate drainer from the dural AVF drainer, eventually joining the dural AVF drainer (Fig. 2E and 2F). Based on these findings, the patient was diagnosed with a coexisting dural AVF and pial AVM sharing a single drainer, and craniotomy was selected. The surgery was performed with the patient in the prone position with a midline suboccipital approach. As expected preoperatively, the dural AVF drainer and pial AVM drainer joined in the subarachnoid space (Fig. 2G). The dural AVF drainer was cauterized and cut first, and this expanded the operative field such that the pial AVM could be visualized (Fig. 2I). Corticotomy was added to the brain surface, and the nidus was dissected circumferentially. Subsequently, the common drainer was severed. Communication between the hematoma cavity and pial AVM was observed, and the pial AVM was discovered to be the source of hemorrhage. This case presented with two flow-related aneurysms (an SCA aneurysm and a PICA distal aneurysm), and neck clipping of the PICA distal aneurysm found in the same operative field was performed. The pathological findings of the extracted intracerebral lesion were characteristic of a typical pial AVM (Fig. 2K). The patient was discharged without sequelae.

Case 2 (Fig. 3): A 75-year-old man underwent head and neck MRI due to neck pain, and a right tentorial dural AVF was found coincidentally. He did not have a history of an intracranial lesion and had been taking prednisolone 7.5 mg/day for autoimmune pancreatitis. Cerebral angiography showed that the MMA, OA, and tentorial artery were supplying the dural AVF, and that pial arteries from the middle cerebral artery (MCA), posterior cerebral artery (PCA), and SCA were also involved as feeders of the shunt (Fig. 3A–3D). The drainer of the dural AVF was single and continued to the basal vein of Rosenthal through the posterior temporal basal vein. Magnetic resonance angiography (MRA) showed intracerebral gathering of the pial artery from the PCA (Fig. 3E); however, the coexistence of a pial AVM was not

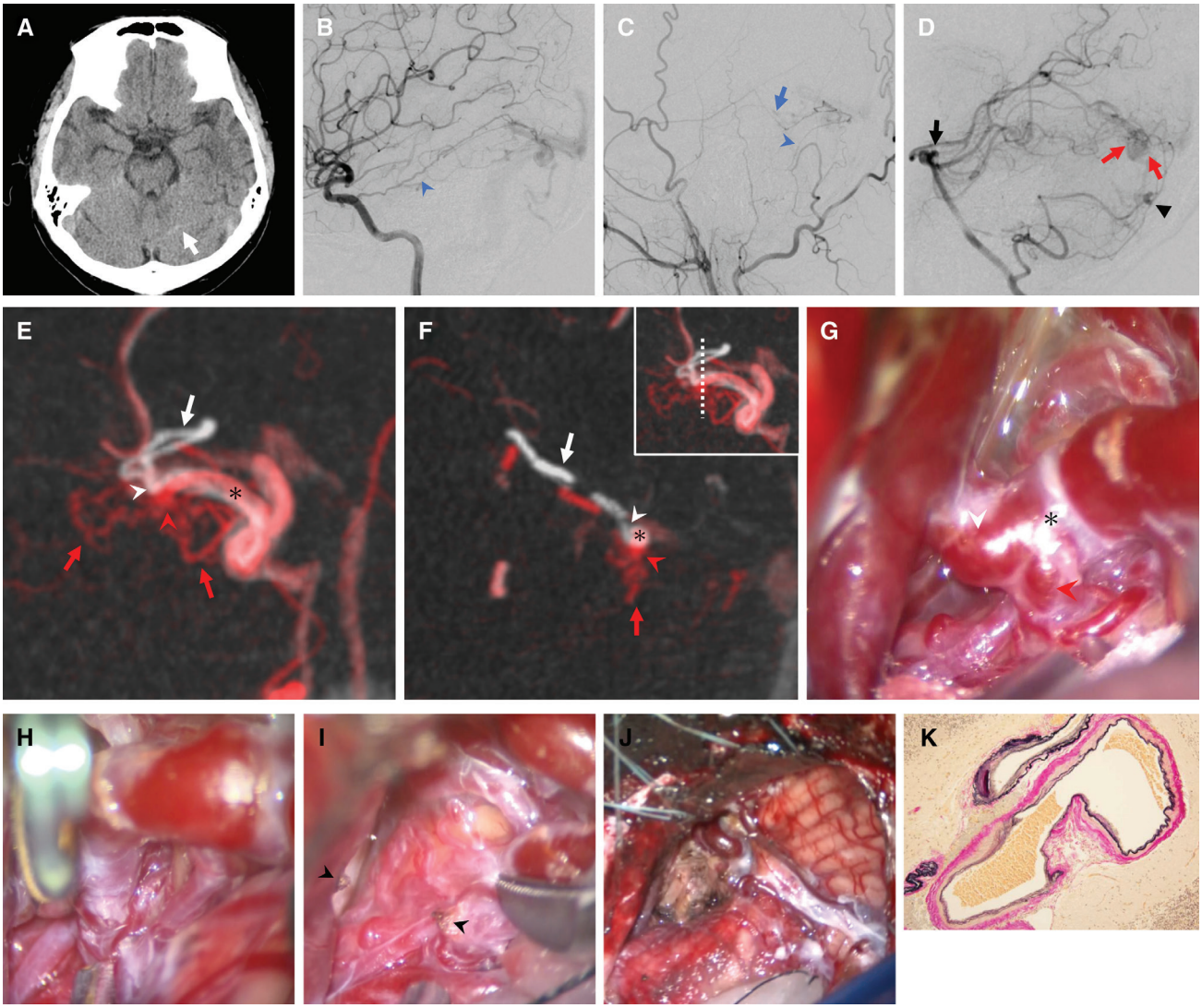

Fig. 2 Case 1, A 52-year-old man undergoing treatment for hypertension. (A) Cerebellar hemorrhage (white arrow) is observed on head CT performed for a headache. (B) Cerebral angiography (lateral view) of the left ICA shows a dural AVF with the tentorial artery (blue arrowhead) as the main feeder and a single drainer that drains into the straight sinus through the superior vermian vein. (C) Cerebral angiography (lateral view) of the left external carotid artery shows that the MMA (blue arrow) and OA (blue arrowhead) are also dural AVF feeders. (D) Cerebral angiography (lateral view) of the left VA shows the development of the left SCA and the left PICA (red arrows). A basilar artery-SCA artery aneurysm (black arrow) and a PICA distal aneurysm (black arrowhead) are observed, which are thought to be flow-related aneurysms. (E and F) Fusion images of the left ICA angiography (white) and left VA angiography (red) created using syngo X Workplace (SIEMENS). (E) shows the long-axis view of the drainer and (F) the short-axis view of the drainer at the dotted line position. In each image, the tentorial artery (white arrow) forms a shunt on the dura and continues to the drainer (white arrowhead). Pial arteries of the PICA and SCA form a nidus in the brain (red arrow) and continue to a different drainer (red arrowhead) from the dural AVF drainer. The two drainers join at the subarachnoid space near the tentorial sinus (asterisk). Thus, this condition is diagnosed as the coexistence of a pial AVM. (G) Surgical findings confirm the junction (asterisk) of the dural AVF drainer (white arrowhead) and the pial AVM drainer (red arrowhead). (H) Because clipping the dural AVF drainer alone does not normalize the red vein, the coexistence of a pial AVM is confirmed. (I) A pial AVM could be visualized after severing the dural AVF drainer. Severed site of the dural AVF drainer (black arrowhead). (J) After pial AVM extraction, the red vein disappears. (K) Pathological findings of the extracted intracerebral lesion (Elastica van Gieson stain, 100× magnification) show aggregation of abnormal dilated blood vessels with thin walls and intervening brain parenchymal tissue, and an AVM is diagnosed. AVF: arteriovenous fistula, AVM: arteriovenous malformation, CT: computed tomography, ICA: internal carotid artery, MMA: middle meningeal artery, OA: occipital artery, PICA: posterior inferior cerebellar artery, SCA: superior cerebellar artery, VA: vertebral artery.

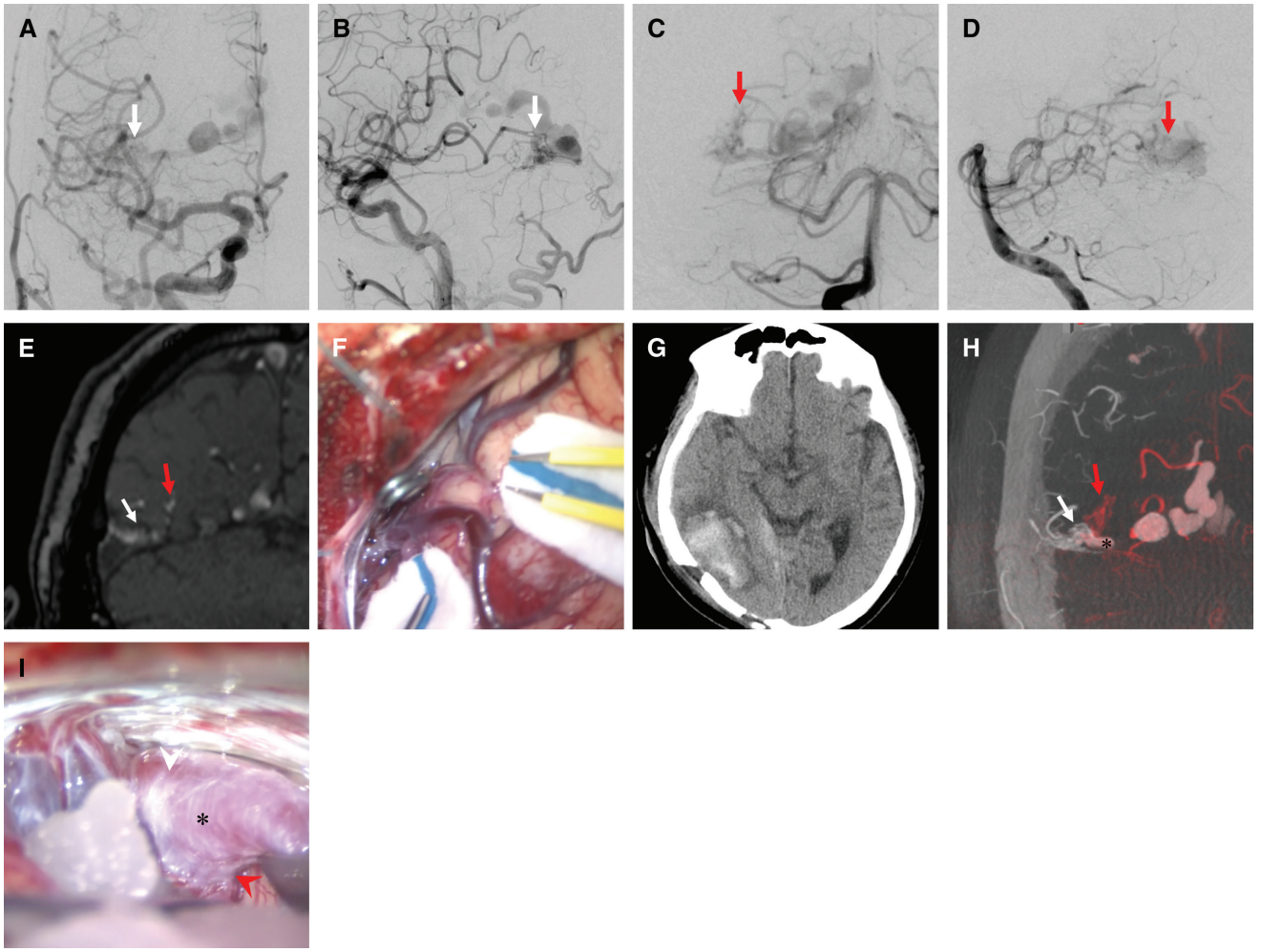

Fig. 3 Case 2, A 75-year-old man with a dural AVF that is found coincidentally. Anteroposterior view (A) and lateral view (B) of the right CCA angiography show a tentorial dural AVF with the MMA and OA as the feeders. The pial artery supply of the MMA (white arrow) is also feeding the dural AVF. Anteroposterior view (C) and lateral view (D) of the right VA angiography show aggregation of the pial artery of the PCA (red arrow). (E) MRA (coronal image) shows the presence of blood vessel aggregation on the dura (white arrow) and in the brain (red arrow). (F) The drainer is clipped with a titanium clip, and the surgery is then finished after confirming the disappearance of the shunt with indocyanine green videoangiography. (G) CT on the day after surgery shows intracerebral hemorrhage with an acute subdural hematoma. (H) Coronal fusion images of the right CCA angiography (white) and right VA angiography (red) created with syngo X Workplace (SIEMENS); preoperative images are re-examined. The meningeal artery and the pial artery of the MCA form the dural AVF (white arrow). The pial artery of the PCA aggregates in the brain and forms a nidus as the pial AVM (red arrow) and both drainers join immediately above the transverse sinus (asterisk). (I) Reviewing the intraoperative image, the junction (asterisk) of the dural AVF drainer (white arrowhead) and the pial AVM drainer (red arrowhead) appears to be present. AVF: arteriovenous fistula, AVM: arteriovenous malformation, CCA: common carotid artery, CT: computed tomography, MCA: middle cerebral artery, MMA: middle meningeal artery, MRA: magnetic resonance angiography, OA: occipital artery, PCA: posterior cerebral artery, VA: vertebral artery.

noticed at this point, and surgery proceeded. With the patient in the prone position, the shunt point was accessed via the supratentorial infra-occipital approach. The area surrounding the drainer showed numerous normal bridging veins, resulting in a narrow operative field and difficulties in fully exposing the drainer for observation. After finishing the surgery with drainer clipping alone (Fig. 3F), an intracerebral hematoma with acute subdural hematoma was observed on head CT the following day (Fig. 3G). The same software as in Case 1 was used to create fusion images of the right common carotid artery angiography and right VA angiography images (Fig. 3H). On careful examination, pial arteries

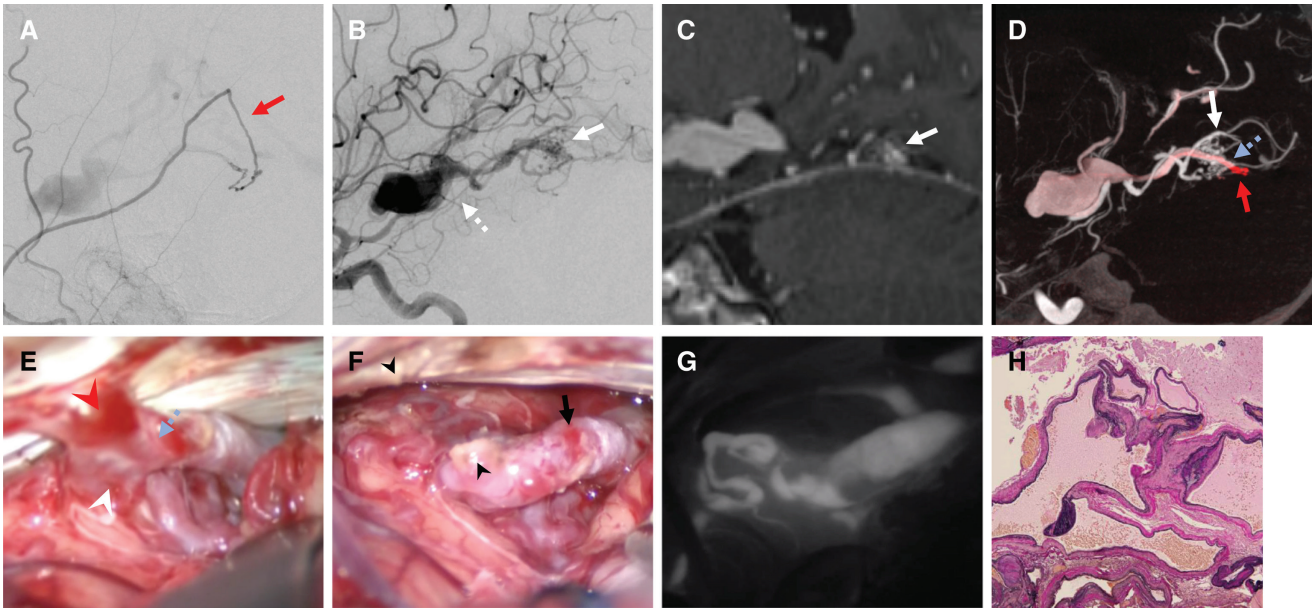

Fig. 4 Case 3, A 48-year-old man with no significant medical history. (A) Right ECA angiography (lateral view) shows a tentorial dural AVF with the MMA (red arrow) as the feeder. A single drainer is present, forms a shunt at the tentorial sinus, and subsequently drains into the straight sinus. (B) Angiography (lateral view) of the right ICA shows that the tentorial artery is also a dural AVF feeder (dashed white arrow). Moreover, the pial artery of the PCA has developed (white arrow). (C) On contrast-enhanced MRI T1 imaging, the pial artery of the PCA gathers in the brain parenchyma and appears to be forming a nidus (white arrow). (D) Fusion images of the right ECA angiography (red) and right ICA angiography (white) are created using syngo X Workplace (SIEMENS). The MMA (red arrow) forms a shunt immediately above the cerebellar tentorium. The pial artery of the PCA forms a nidus (white arrow) and continues to a different drainer from the dural AVF drainer. The junction of the two drainers is shown (blue dashed arrow). Based on these observations, the coexistence of a pial AVM is diagnosed, and craniotomy is selected. (E, F, and G) Images of the surgical finding are shown. (E) As expected preoperatively, the junction (blue dashed arrow) of the dural AVF drainer (red arrowhead) and the pial AVM drainer (white arrowhead) is confirmed. (F) shows the point at which the dural AVF drainer is cut (black arrow head) and the red vein that remains even after the cutting (black arrow). (G) Indocyanine green angiography also visualizes the drainer in the arterial phase, and the coexistence of a pial AVM is confirmed. (H) Pathological findings of the extracted intracerebral lesion (Elastica van Gieson stain, 40× magnification) are shown. Artery-like and vein-like dilated vessels are aggregated, corroborating the presence of an AVM. AVF: arteriovenous fistula, AVM: arteriovenous malformation, ECA: external carotid artery, ICA: internal cerebral artery, MMA: middle meningeal artery, MRI: magnetic resonance imaging, PCA: posterior cerebral artery.

of the MCA and SCA joined the shunt point of the dural AVF, but the pial artery from the PCA formed a nidus in the brain parenchyma. Moreover, reviewing the intraoperative image, the junction of the dural AVF drainer, and the pial AVM drainer appeared to be present (Fig. 3I). Therefore, the hemorrhage was considered to be from the remaining pial AVM. Fortunately, the patient only experienced visual field impairment and was discharged home, albeit requiring a short duration of rehabilitation. Follow-up observation with angiography is ongoing, but a remaining shunt or recurrence has not been observed thus far.

Discussion

The coexistence of a pial AVM was proven by both surgical findings and pathological findings. The coexisting pial AVM is small and extremely close to the shunt point of the dural AVF; thus, at first glance, it appears to be a dural AVF with a pial arterial supply, and this is the most crucial point to note. To our knowledge, this is the first report in which this condition could be clearly diagnosed based on preoperative images.

Since a single drainer is shared in this condition, mistaking the condition as a dural AVF with a pial arterial supply is extremely dangerous. Even when treatment is performed under craniotomy, it is difficult to extensively expose the drainer covered in thick subarachnoid in the narrow operative field and to find the joining point with the pial AVM. Furthermore, occluding the common drainer leads to the disappearance of the shunt on intraoperative indocyanine green videoangiography and

Table 1 Clinical details of six patients diagnosed with coexisting dural AVF and pial AVM sharing common drainage

Case no.	Age (years)/ sex	Clinical presentation	Location			Borden classification	Previous intracranial event	Complication
			Side	Pial AVM	Dural AVF			
1	52/Male	Cerebellar hemorrhage	Left	cerebellum	Tentorial sinus	Type III	None	None
2	75/Male	Asymptomatic	Right	occipital	Tentorial sinus	Type III	None	Intracerebral hemorrhage
3	48/Male	Intraventricular hemorrhage	Right	occipital	Tentorial sinus	Type III	None	None
4	71/Male	Intracerebral hemorrhage	Left	temporal	Transverse sinus	Type III	Transverse-sigmoid Thrombosis	None
5	55/Male	Asymptomatic	Right	occipital	Convexity	Type III	None	Intracerebral hemorrhage
6	75/Male	Asymptomatic	Right	occipital	Convexity	Type III	None	None

AVF: arteriovenous fistula, AVM: arteriovenous malformation.

intraoperative angiography, and this may lead the surgeon to finish the treatment without realizing the potential coexistence of a pial AVM. For these reasons, it is important to determine the coexistence of a pial AVM at the time of preoperative diagnostic imaging to prevent hemorrhagic complications. When pial arterial development at the dural AVF is evident, we consider the possibility of this condition and subsequently determine whether the pial arteries aggregate in the brain, forming a nidus. Blood vessel aggregation is determined by a combination of source images from CT angiography and three-dimensional digital subtraction angiography (3D-DSA), and the location of aggregation (in the brain or in the dura) is determined using contrast-enhanced MRI T1 thin-slice images, constructive interference in steady state (CISS) on plain MRI, and MRA. If a nidus is formed in the brain and the coexistence of an AVM is plausible, we use 3D-DSA to determine the presence of a separate pial AVM drainer from the dural AVF drainer. We find it useful to compare the drainer from the pial feeder and the drainer from the meningeal feeder by fusing the individually taken images selectively.

Similar cases with multiple arteriovenous shunts close to each other that share a single drainer have been previously reported: specifically, two cases with a coexisting dural AVF and pial AVF, and two cases with a coexisting dural AVM and pial AVM.[6–9] To emphasize the presence of two arteriovenous shunts, the term "true mixed pial-dural AVM" was used in one of these cases. "Mixed pial-dural AVM" stems from the three classifications by Newton et al. in 1969: "pure pial AVM" or an arteriovenous shunt with pial arterial supply only; "mixed pial-dural AVM" or an arteriovenous shunt with a pial and meningeal arterial supply; and "pure dural AVM" or an arteriovenous shunt with meningeal arterial supply only.[10] These were classified based on the type of feeder and do not specify the location of shunts, and therefore, often cause confusion.[2,11,12] The important points affecting the treatment of arteriovenous shunt disease are the locations of the shunts, and whether the drainer is shared in cases of multiple shunts. Since an accurate reflection of the condition that does not mislead the medical staff is desirable, it is not appropriate to classify by the type of feeder. We therefore use "coexisting dural AVF and pial AVM sharing a common drainer" to describe this condition. Accordingly, we would like to emphasize the importance of being aware of this condition when treating a dural AVF accompanied by pial arterial development.

It is well known that an increase in venous perfusion pressure due to an arteriovenous shunt

causes local ischemia in the surrounding brain tissue, leading to the formation of a new arteriovenous shunt.[1,8,13] The formation of an arteriovenous shunt following local ischemia may be caused by abnormal expression of various angiogenic factors, such as vascular endothelial growth factor, basic fibroblast growth factor, and alpha transforming growth factor, that act on the capillarovenous endothelial cells and other cellular processes involved in the remodeling of the vascular system.[14,15] Since the AVM in each of our cases was small, it is likely that the dural AVF was the primary pathology and that the AVM was secondary to the dural AVF.

Although the overall prevalence is unknown, the frequency does not seem low. Hemorrhagic complications occur frequently when treating a dural AVF with a pial arterial supply, and it is possible that this condition was confused with them. When development and aggregation of the pial artery are observed, the coexistence of a pial AVM should be considered, and images should be carefully examined. In suspicious cases, craniotomy should be selected to resect the AVM. Case 2 and Case 5 were early cases that led to hemorrhage; however, hemorrhage could have been avoided if this condition had been acknowledged preoperatively in both cases. Therefore, awareness of this condition is extremely important for selecting the treatment strategy for a dural AVF with a pial arterial supply.

Conclusions

A neighboring dural AVF and pial AVM sharing a common drainer initially seems like a dural AVF with a pial arterial supply, but there is a high rate of hemorrhagic complications if treated without awareness of the coexisting pial AVM. Since it is difficult to confirm the coexisting pial AVM intraoperatively, it is important to note the presence of a pial AVM at the time of preoperative imaging diagnosis.

Conflicts of Interest Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

1) Osada T, Krings T: Intracranial dural arteriovenous fistulas with pial arterial supply. *Neurosurgery* 84: 104–115, 2019
2) Wu Q, Zhang XS, Wang HD, et al.: Onyx embolization for tentorial dural arteriovenous fistula with pial arterial supply: case series and analysis of complications. *World Neurosurg* 92: 58–64, 2016
3) Cannizzaro D, Brinjikji W, Rammos S, Murad MH, Lanzino G: Changing clinical and therapeutic trends in tentorial dural arteriovenous fistulas: a systematic review. *AJNR Am J Neuroradiol* 36: 1905–1911, 2015
4) Gandhi D, Chen J, Pearl M, Huang J, Gemmete JJ, Kathuria S: Intracranial dural arteriovenous fistulas: classification, imaging findings, and treatment. *AJNR Am J Neuroradiol* 33: 1007–1013, 2012
5) Liu C, Xu B, Song D, et al.: Clinical approach of using Onyx via transarterial access in treating tentorial dural arteriovenous fistula. *Neurol Res* 36: 983–991, 2014
6) Ozawa T, Miyasaka Y, Tanaka R, Kurata A, Fujii K: Dural-pial arteriovenous malformation after sinus thrombosis. *Stroke* 29: 1721–1724, 1998
7) Elia C, Minasian T, Noufal M, Chhabra V: Pial-dural intracranial arteriovenous fistula with flow-associated aneurysmal rupture-case report with review of literature and proposal on the mechanism of hemorrhage and treatment options. *World Neurosurg* 105: 1040.e1015–1040.e1019, 2017
8) Funakoshi Y, Hatano T, Saka M, et al.: Dural and pial arteriovenous fistulas connected to the same drainer in the middle cranial fossa: a case report. *World Neurosurg* 118: 47–52, 2018
9) Maki Y, Funaki T, Takahashi JC, et al.: "True" mixed pial-dural arteriovenous malformation: a case report. *No Shinkei Geka* 42: 745–750, 2014 (Japanese)
10) Newton TH, Cronqvist S: Involvement of dural arteries in intracranial arteriovenous malformations. *Radiology* 93: 1071–1078, 1969
11) Miyasaka Y, Kurata A, Saegusa H, Yuzawa I, Utsuki S, Ohwada T: Dural-pial arteriovenous malformation with unusual venous drainage. *Neurol Med Chir (Tokyo)* 36: 91–95, 1996
12) Jimbo H, Ikeda Y, Izawa H, Otsuka K, Haraoka J: Mixed pial-dural arteriovenous malformation in the anterior cranial fossa–two case reports. *Neurol Med Chir (Tokyo)* 50: 470–475, 2010
13) Terada T, Higashida RT, Halbach VV, et al.: Development of acquired arteriovenous fistulas in rats due to venous hypertension. *J Neurosurg* 80: 884–889, 1994
14) Shweiki D, Itin A, Soffer D, Keshet E: Vascular endothelial growth factor induced by hypoxia may mediate hypoxia-initiated angiogenesis. *Nature* 359: 843–845, 1992
15) Kilic T, Pamir MN, Kullu S, Eren F, Ozek MM, Black PM: Expression of structural proteins and angiogenic factors in cerebrovascular anomalies. *Neurosurgery* 46: 1179–1191; discussion 1191-1172, 2000

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