INTRACRANIAL pial AVFs are rare cerebrovascular lesions of the brain that have only recently been considered a pathological entity distinct from other vascular malformations. Pial AVFs are composed of ≥ 1 arterial feeding vessels and a single draining vein that usually has high perfusion pressure and generally occur in infants. Cases involving adults are very rare and the developmental mechanisms and natural history of these lesions remain unknown. The authors present a case of multiple pial AVFs in an adult in whom the lesions developed after radiosurgical treatment of dural AVFs. Direct disconnection of pial arterial supplies was performed, and the abnormal shunts were successfully eliminated. The authors report the clinical course of this case and discuss the characteristics of and treatment strategy for multiple pial AVFs, reviewing the published literature.

Case Report

History and Presentation. This 40-year-old woman with chronic headache was admitted to the neurological care unit in our hospital in September 2000. Cerebral angiography showed a partial venous flow defect in the SSS and the corkscrew-like appearance of cortical veins (Fig. 1A). Sinus thrombosis was diagnosed and an anticoagulant was administered. Follow-up angiography demonstrated improved venous circulation (Fig. 1B) and the patient was discharged with no neurological deficit 1 month after admission.

In September 2002, 2 years after the initial admission, she experienced a sudden-onset headache. Fluid-attenuated inversion recovery MR images revealed a thin subarachnoid hemorrhage in the left frontal cortex (Fig. 2A). The early arterial phase of ECA angiography revealed bilateral dural AVFs in the anterior SSS, with arterial supply from the bilateral MMAAs associated with cortical reflux (Fig. 2B). Transvenous embolization was attempted, but venous catheterization was difficult and endovascular treatment was not achieved.
Radiosurgical treatment was applied to these lesions twice—initially using a dose of 25 Gy and then, 2 years later, 20 Gy. Follow-up cerebral angiography 2 years after the last radiosurgery showed a decrease in the bilateral dural AVFs, but the abnormal shunts had not completely disappeared (Fig. 3A). The affected sinus was partially occluded in the proximal portion, and the cortical reflux also had not completely disappeared. Moreover, new development of multiple pial AVFs was observed (Fig. 3B). New arterial supplies had developed from the bilateral cortical branches resulting in new pial AVFs. We decided to perform open surgery to completely disconnect the bilateral MMAs and bilateral cortical arterial supplies into the dural AVFs.

**Operation.** A bilateral frontal craniotomy was performed. Arterial shunting directed toward the SSS and a red vein on the surface of the frontal lobe was observed. Initially, this red vein was thought to be arterialized due to cortical reflux via cortical veins. The feeding arterial branches from the MMAs were coagulated electrically. The reddish and dilated cortical veins flowing into the SSS normalized, but the red vein on the cortical surface remained. This red vein was independent of the dural AVFs and had arterial supply from the pial arteries on the surface of the frontal lobe. There were also red veins independent of the dural AVFs on the surface of the bilateral frontal lobes. They were not symmetrical but were very similar.

Pial arteries around the red vein were dissected and temporarily occluded by vascular clip, and the responsible pial branches were examined. These pial AVFs had multiple arterial shunting points. The pial arterial supplies were disconnected by coagulation, resulting in the recovery of the reddish dilated veins to normal venous flow.

**Postoperative Course.** Postoperative cerebral angiography showed the disappearance of dural AVFs and no abnormal shunting on the right side of the frontal surface, but a tiny remnant of abnormal shunting was evident in the left frontal region (Fig. 5A and B). The patient was discharged without neurological deficits 13 days after the operation.

**Discussion**

Pial AVFs have only recently been considered distinct from AVMs. They consist of ≥ 1 arterial connections to a single venous channel without any intervening nidus or capillary bed. The pathophysiological cause of pial AVFs remains unclear. Congenital pial AVFs usually develop in childhood and are part of syndromes such as Rendu-Osler-Weber or Klippel-Trenaunay-Weber syndrome, suggesting the importance of genetic abnormality. On the other hand, acquired pial AVFs have been reported as a result of ischemia or head trauma. Abnormal angiogenesis caused by cerebral ischemia or contusion might affect the development of pial AVFs. Ratliff and Voorhies reported a case of pial AVF associated with AVM. They discussed whether venous hypertension caused by the drainage pathway of

![Fig. 1. Angiograms obtained before (A) and after (B) the initial anticoagulant therapy (2000). A: Left (left) and right (right) ICA angiograms showing partial occlusion of venous flow in the SSS and corkscrew-like dilated cortical veins, suggesting sinus thrombosis. B: Left (left) and right (right) ICA angiograms obtained after anticoagulant treatment showed improved venous circulation.](image-url)
AVM promotes the synthesis of fistulas. In our present case, the development of pial AVFs proceeded interestingly as typical acquired lesions. Venous hypertension caused by dural AVFs in the SSS might affect cortical venous circulation; however, multiple pial AVFs were observed while the shunting flow of dural AVFs decreased after radiosurgical treatment. Radiation itself or various angiogenic factors caused by the obliteration of coexistent dural AVFs might affect the development of multiple pial AVFs in a similar location of the bilateral frontal lobe. Lai et al. postulated a developmental mechanism for multiple pial AVFs: the vascular modeling and remodeling process might be primarily involved in congenital events at the cellular and structural level, with a trigger such as radiation or mechanical, or infective factors. They noted that ischemia or hypoxia can be an important etiological factor for acquired pial AVFs. Radiosurgical treatment itself might affect congenital vascular vulnerability, and hemodynamic change after the obliteration of dural AVF might cause local ischemia and it might trigger the development of pial AVFs.

Patients with pial AVFs usually present with headache, hemorrhage, seizure, or neurological deficits. Their clinical symptoms are closely related to their age at symptom onset. Enlarged head circumference, cranial erosion, and heart failure have been frequently reported in neonates and infants with AVFs. Turbulence and increased pressure within draining veins may lead to the formation of giant varices. These dilated venous channels can exert a significant mass effect, compressing adjacent structures and impairing the cerebrospinal fluid pathway. Neurological deficits are often caused by the mass effect of the varix or by cerebral venous congestion and ischemia. The natural history and risk of bleeding associated with pial AVFs have not been thoroughly documented in studies of risk factors for bleeding in AVM cases in which multiple factors were considered, such as single venous drainage and small AVM. Single venous drainage and high perfusion pressure are both characteristics of pial AVFs, and the possibility of bleeding in patients with pial AVFs should be considered.

Pial AVFs are treated differently from AVMs, and disconnection of arteriovenous communication should eliminate the abnormality without the necessity for lesion resection. Successful treatment of AVFs by simply disconnecting the shunt either surgically or endovascularly has been reported in several recent reports. Identifying the exact fistula site preoperatively, however, is difficult. In our present case, arterial shunting channels were supplied by small pial arteries on the cortical surface; these arteries were difficult to detect by preoperative angiography, and

Fig. 2. Studies obtained before radiosurgery. A: Fluid attenuated inversion recovery MR images revealing a thin subarachnoid hemorrhage in the left frontal lobe. B: Early arterial phase left (left) and right (right) ECA angiograms revealing bilateral dural AVFs in the anterior SSS, with arterial supply from the bilateral MMAs associated with cortical reflux.
endovascular obliteration was impossible. Direct observation was important and effective for the detection of multiple arterial shunts, but it was not easy to identity all the pial supply.

With careful pretreatment planning by an experienced combined neurosurgical and endovascular team, the decision to treat a patient safely with one technique or combined techniques can be made on the basis of lesion-specific factors, such as location, flow velocity, and angiographic features, as well as patient-specific factors.

**Fig. 3.** Cerebral angiograms obtained 2 years after the last radiosurgery.  
A: Left (left) and right ECA (right) cerebral angiograms showing decreased abnormal shunting flow of dural AVFs.  
B: Left (left) and right ICA angiograms (right) showing that the abnormal shunts did not completely disappear. Multiple pial AVFs were newly observed on the bilateral frontal surface.

**Fig. 4.** Intraoperative images of the left frontal lobe surface.  
A: A red vein on the cortical surface was observed even after reddish and dilated cortical vein flow into the SSS was temporarily occluded by clipping at the entrance to the SSS. The red vein was independent of cortical reflux from dural AVFs and received arterial supply from pial arteries on the surface of the frontal lobe.  
B: The right branched red vein was temporarily occluded by clipping. The left branched red vein normalized, and arterial shunting from pial arteries (arrow) was detected at multiple points along the distal portion of the clipped right branch. The arrow indicates arterial shunting from a pial artery.  
C: The red vein recovered normal venous flow after disconnection of these pial arterial supplies.
Disclaimer

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

References


Fig. 5. A: Postoperative (left) and (right) ECA angiograms demonstrating disappearance of the dural AVFs. B: Postoperative (left) and (right) ICA angiograms demonstrating the absence of abnormal shunting.


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