

Case Report

Intracranial pial arteriovenous fistulas: two case reports

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Abstract: *Objective:* Pial arteriovenous fistulas (pAVFs) are rare neurovascular lesions of the brain. They are characterized by arteriovenous malformation but with absent intervening nidus of vessels, and thus are easily misdiagnosed or never diagnosed. Misdiagnosis and inadequate treatment can lead to recurrent fistulas and cerebral hemorrhage. *Methods:* We report two cases diagnosed as pAVFs. We also performed a brief review concerning the development, etiology, pathology, manifestation, diagnosis, and treatment of pAVFs. *Results:* Case 1, a 44-year-old male, who had suffered from paroxysmal left limbs disability for one year that had aggravated in the last two months. Digital subtraction angiography (DSA) showed a pAVF in the left temporal lobe fed by a branch of the left middle cerebral artery (MCA), and in the late arterial phase, no nidus or dural arteriovenous fistulas were detected (Figure 1E, 1F). The author considered his pAVF was related to transient venous hypertension secondary to mild trauma or recanalization of asymptomatic phlebotrombosis. Moreover, there were no recurrent symptoms after the timely endovascular treatment. In Case 2, a 52-year-old male patient was presented with a 3-day history of headache. DSA showed the right MCA was pushed, a dilated vessel image in right occipitoparietal lobe presented previously, draining via the right sigmoid sinus and the right jugular vein, but no nidus was found (Figure 2B, 2C). And the author considered this pAVF was related to used brain concussion. This patient recovered completely after endovascular treatment. Both cases had been taking Nimotop for 1 month after discharge, and no recurrent cerebral hemorrhage occurred during a follow up of 1 year. *Conclusions:* More attention should be dedicated to cerebral hemorrhage cases with unknown causes, which may well be pAVFs. Endovascular treatment is the first choice for pAVFs, and DSA findings and their interpretation by an experienced radiologist are essential for the correct and timely diagnosis.

Keywords: Intracranial pial arteriovenous fistula (pAVF), digital subtraction angiography (DSA), endovascular treatment

Introduction

Pial arteriovenous fistulas (pAVFs) are rare neurovascular lesions of the brain, accounting for only 1.6% to 4.7% of all brain arteriovenous malformations (AVM) [1]. A limited number of cases have been reported in China and abroad so far, and little is known of their pathogenesis.

They have only recently been considered a distinct pathological entity belonging to AVM [2]. pAVFs are composed of a single venous channel in communication with one or more arterial connections, with no intervening nidus of vessels [3]. As a result, many patients are misdiagnosed or never diagnosed. pAVFs have pial or cortical feeding artery instead of an external

carotid artery, and thus the lesion is located in the pia mater instead of the dura; the supratentorial area is the most common site [3]. The current treatment is associated with “flow disconnection”, which can be accomplished via surgical or endovascular procedures [3, 4]. Herein, we investigate and discuss the diagnosis and treatment of two cases with pAVFs, in which the patients were admitted and treated in the Second Affiliated Hospital of Zhejiang University.

Case report

Case 1, a 44-year-old male, who had suffered from paroxysmal left limbs disability for one year that had aggravated in the last two months. The physical examination on admission revealed no positive signs of this patient.

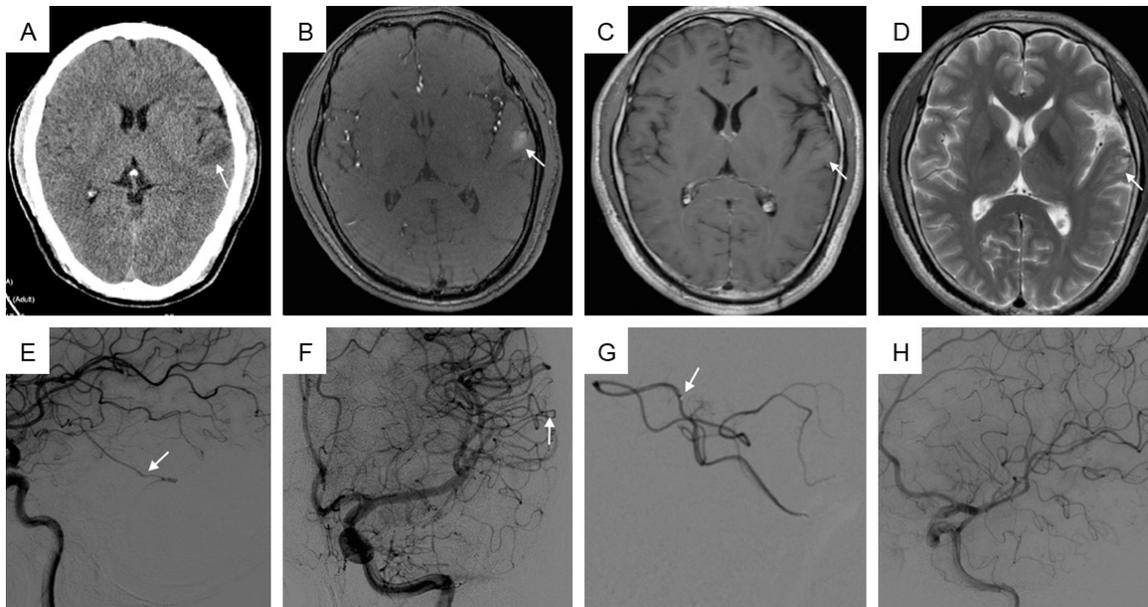


Figure 1. (For Case 1) (A) Cranial CT showing the left temporal low-density area, as change after hemorrhage; (B) Magnetic resonance angiography displays no abnormal vessels; (C and D) MRI exhibits left temporal hematoma; (E and F) Lateral and anteroposterior DSA establish the presence of a fistula fed through a branch of the left MCA, from which drainage veins (arrows) developed into a lateral sinus in the late arterial phase. No nidus was found; (G) Superselective angiography presents the microcatheter (arrow) located in the feeding artery of the fistula in the left temporal lobe, to the proximal part, infusing the colloid; (H) DSA demonstrates the occlusion of the fistula without any residue remaining.

Cerebral Tomography (CT) showed a left temporal low-density area, as change after hemorrhage (**Figure 1A**); Magnetic resonance imaging (MRI) with angiography showed a left temporal hematoma, but with no abnormal vessels (**Figure 1B-D**). DSA showed a pAVF in the left temporal lobe fed by a branch of the left middle cerebral artery (MCA), and in the late arterial phase, no nidus or dural arteriovenous fistulas were detected (**Figure 1E, 1F**). Several days later, endovascular embolization of the pAVF was performed with Onyx18 (**Figure 1G**), which led to an occlusion of the fistula without any residue remaining (**Figure 1H**). The procedure was successful, the patient suffered no complications, and his symptoms of limbs disability disappeared.

In Case 2, a 52-year-old male patient was presented with a 3-day history of headache. He had had “cerebral concussion” 20 years ago. On examination, he presented no positive signs. Cranial CT showed a right temporal hematoma and a right frontal low-density area considering as an encephalomalacia focus (**Figure 2A**). DSA showed the right MCA was pushed, a dilated vessel image in right occi-

toparietal lobe presented previously, draining via the right sigmoid sinus and the right jugular vein, but no nidus was found (**Figure 2B, 2C**). This patient was finally diagnosed with pAVF in the right occipitoparietal lobe. Three weeks later, after the patient was transferred to neurosurgery, endovascular embolization of the AVF was performed with Onyx18. Then, we established that the feeding artery was a branch of the right MCA (**Figure 2D**). There were no abnormal drainage veins, and the flow in the main feeding artery was without any block (**Figure 2E, 2F**). The procedure was successful, and the patient recovered completely. Repeat CT scan one month later showed the right temporal hematoma had already been absorbed (**Figure 2G, 2H**).

Both cases had been taking Nimotop for 1 month after discharge, and no recurrent symptoms or cerebral hemorrhage occurred during a follow up of 1 year.

Discussion

The pAVF is an exceedingly rare disease. In a series of 320 AVMs reported by Halbach *et al.*,

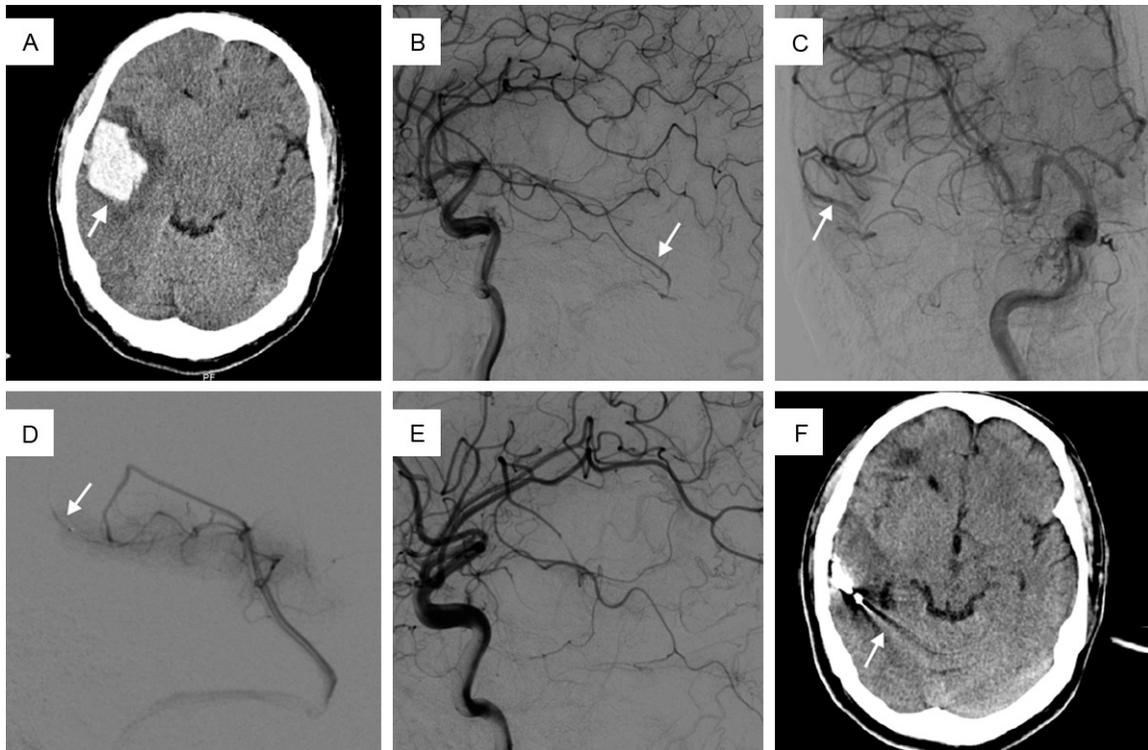


Figure 2. (For Case 2) (A) CT images displaying right temporal high-density area, considering as right temporal hematoma; (B and C) Lateral and anteroposterior DSA showing a dilated vessel image (with arrows) in the right occipitoparietal lobe presented previously, draining via the transverse sinus, the right sigmoid sinus, and the jugular vein. No nidus was found. (D) After the position of the fistula was determined, a microcatheter (arrow) was superselectively delivered into the proximal part of the feeding artery of the right temporal pAVF; then infusion with Onyx was performed. The fistula was embolized: (E) Lateral view showing no residue; (F) Repeat CT scan revealed the right temporal hematoma had already been absorbed.

only five cases (1.6%) were identified [5]. Most pAVFs have more than one feeding artery, named as “multiple-channel” type. Compared with that, the “single-channel” type is much less common. However, both pAVFs we reported were fed by a branch of MCA, and superselective angiography in DSA showed that they were both “single-channel”.

pAVFs may be congenital or acquired [6]. Congenital pAVFs usually present in childhood as part of the Rendu-Osler-Weber disease [7, 8] or Klippel-Trenaunay-Weber syndrome [9]. Although the pathophysiology of congenital pAVFs remain unclear, it is possible that a misstep in the embryologic development of the cerebrovasculature produces these lesions. Alternatively, abnormal angiogenesis and associated vascular growth factors and cytokines may play a role in the development of acquired pAVFs [6]. Current evidence suggests that acquired pAVFs may be related to venous

hypertension, secondary to head trauma, brain surgery, venous thrombosis, etc. [10]. The present cases were of two male patients, in a normal state, with no Rendu-Osler-Weber disease or Klippel-Trenaunay-Weber syndrome. Case 1 had no history of head trauma or brain surgery, whereas Case 2 had a history of “cerebral concussion” (not shown in all his images) that might have led to the development of head trauma or venous hypertension, eventually causing pAVF.

Hemorrhage is the most common symptom of pAVFs. The risk of hemorrhage from pAVFs is not clear. Single-venous drainage, small AVM size, high flow and perfusion pressure, and impaired venous drainage and venous varices may all contribute to the risk of pAVFs hemorrhage [11-14]. In a previous report, conservative management of pAVFs was associated with mortality in five (63%) of eight patients that was caused by hemorrhage in the acute phase or

secondary hemorrhage [15]. The two cases described herein were both male pAVF patients with cerebral hemorrhage. Case 1 presented with neurological deficit, paroxysmal left limbs disability and rigidity; however, the lesion was also in the left side, which was unreasonable, for that, the author considered the symptoms and hemorrhage occurred separately, they were not related, which means the patient might also have had accompanying problems. On the other side, case 2 presented only with headache.

The connection between an arterial feeder directly into a solitary draining vein without any intervening tangle of vessels creates conditions for a rapid high flow. The pathologic features of pAVFs arise principally from its high-flow nature. Associated venous varices are produced by the high, turbulent flow from the arteriovenous shunt and are common in pAVFs [16-22]. The pathologic nature of pAVFs is associated with a high-flow system which needs to be eliminated to remove the symptoms. The disconnection of direct shunting, either by endovascular embolization or surgically, is sufficient to achieve successful treatment. The relationships between the varix and the clinical course of pial AVF have not yet been elucidated. Evidence in the literature shows patients presented with symptoms at a younger age (<15 years old) are more likely to have varices. Furthermore, the absence of a varix does not have a significant correlation with hemorrhage [23]. Using DSA, we did not find any venous varix in the present two cases (**Figure 1E** and **1F**; **Figure 2B** and **2C**). Generally, an experienced radiologist is needed to provide support in the diagnosis of pAVFs without any varix; otherwise, missed diagnosis can occur.

The knowledge of the natural history of these lesions is scarce due to their rare incidence. Only one earlier study established the occurrence of spontaneous thrombosis in a patient with pAVF [24]. Therefore, spontaneous closure of these lesions is not expected. The currently applied treatment aims at "flow disconnection", which can be accomplished via surgical or endovascular means [3, 4]. The treatment of pAVFs and AVMs is different. In AVMs, resection of the entire lesion is necessary because of the multiplicity of communicating shunts. Occlusion of the feeding arteries only of an AVM leaves behind the nidus, which can recruit

new arterial feeders that are often even more difficult to access by transarterial embolization. In addition, in AVMs, the draining veins must be obliterated because they may persist after resection and recruit recanalization with collaterals [3, 25]. Since there is no nidus in pAVFs, surgical clipping for disconnection of the AV shunt should eliminate the abnormality without the necessity for resection of the lesion [25]. Removal of the varix is not attempted unless the malformation bleeds with a resultant hematoma, or the accessible varix produces a mass effect [4]. Nevertheless, the flow disconnection of AV fistulae is sufficient to obliterate the pathological entities, and resection of the lesion is not necessary. DSA can localize the lesion exactly, based on which the optimal therapeutic option can be selected. Although many pAVFs can be treated surgically, the high shunt conductance and deep venous drainage limits the safety and efficacy of surgical treatment [3, 12, 17, 26, 27]. Meyer *et al.* [28] and Bendok *et al.* [29] described the use of deep hypothermic cardiopulmonary bypass to perform surgical resection of such technically difficult lesions. In spite of the emergence of the endovascular treatment as a new therapy option, it is a highly selective approach to eliminate a lesion in contrast to the traditional treatment.

In our cases, the feeding arteries of pAVFs were embolized with Onyx, and no post-surgery complications appeared. The postoperative follow-up revealed no recurrence of cerebral hemorrhage. Therefore, we concluded that this treatment has an adequate short-term efficacy, whereas the long-term efficacy still remains to be determined.

In conclusion, the two cases of pAVFs we reported are rare. At present, pAVF is already dependent from intracranial arteriovenous deformity. Since there is no nidus in pAVFs, cases are easily misdiagnosed or never diagnosed. Therefore, it is critically important that more research attention should be focused on cerebral hemorrhage cases with unknown causes. Furthermore, with increasing the number of cases investigated and reported, fewer patients will be misdiagnosed or never diagnosed. DSA findings and their interpretation by an experienced radiologist are essential for the correct and timely diagnosis.

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Disclosure of conflict of interest

None.

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Reports of intracranial pial arteriovenous fistulas

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