

Case Report

Surgical treatment of the intracranial pial arteriovenous fistula

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Abstract

Background: Pial arteriovenous fistula (PAVF) is a rare vascular condition comprising of one or more arterial vessels that are in direct communication with the draining veins. The condition is also characterized by the absence of a nidus. Due to high blood flows, varicose systems adjacent to the fistula appear. The key characteristic of the arteriovenous direct transition is that it offers a treatment option in which interruption of the blood flow can occur without removing the entire lesion. This study presents two cases of PAVF.

Case Description: The first case is of a 59-year-old male with lesions in the frontal region, fed by a branch of the right anterior cerebral artery and drained by the frontal basal vein to the sphenoparietal sinus. The second case is of a 3-year-old child with a lesion in the right anterior frontal lobe, fed by a branch of the right middle cerebral artery, which drains into the Trolard vein and was associated with large a venous varix.

Conclusion: PAVF is a disease characterized by its rarity, and knowledge of PAVF's clinical presentation is of vital importance in early diagnosis. The treatment of the condition consists of an occlusion of the supply vessel, which can be done by endovascular, microsurgical, or both procedures. Both the cases were successfully treated by microsurgical procedure.

Key Words: Arteriovenous malformation, intracranial, pial arteriovenous fistula

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Quick Response Code:**INTRODUCTION**

Pial arteriovenous fistula (PAVF) is a rare intracranial vascular lesion characterized by having one or more arterial pedicles in direct connection with a single draining vein.^[1] Generally, patients with PAVF present the following initial symptoms: headaches, seizures, hemorrhage, or neurological deficits.^[2-6]

Initially, the PAVF was considered a variation of arteriovenous malformation (AVM), however, it has a

very different clinical appearance as well as differing therapeutic options.^[7,8] This study describes two cases of PAVF, which were surgically treated.

CASE REPORTS**Case 1**

A 59-year-old male reported that he had suffered a complex partial seizure 19 days before admission. The neurological examination was normal.

Computer tomography (CT) showed a lesion in the frontal region that suggested an AVM. The cerebral digital subtraction angiography showed a PAVF fed by a dilated branch of the right anterior cerebral artery, draining from the frontal basal vein to the sphenoparietal sinus and dilation of the frontal basal vein [Figure 1]. There were also small aneurysms in the anterior communicating artery, the pericalosa artery, and two aneurysms in the right internal carotid artery and at the origin of the posterior communicating and anterior choroidal arteries [Figure 1].

The patient was submitted to a frontal basal craniotomy on the left side, contralateral to the draining vein. The fistula and aneurismatic dilation were occluded with clips [Figure 2]. The patient evolved in the immediate postoperative stage with cerebrospinal fluid fistula, which was treated with external ventricular drainage for 4 days. He developed meningitis caused by the multidrug-resistant *Acinetobacter sp.*, which was treated with intrathecal Polymyxin E for 14 days. Postoperative angiographic revealed a complete obliteration of the fistula [Figure 3].

An angiographic exam carried out 6 months after the patient's discharge revealed the persistence of three aneurysms, and a spontaneous regression of the aneurysm on the pericalosa artery. The patient was submitted to a surgical treatment of the aneurysm of the anterior communicating artery and two baby aneurysms on the right internal carotid artery. A postoperative control angiography revealed complete exclusion of these aneurysms.

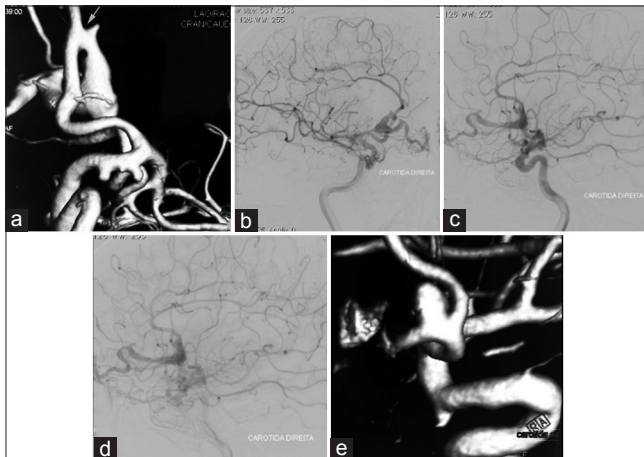


Figure 1: Preoperative angiography of a PAVF with associated aneurysms. (a) 3D angiography showing a PAVF fed by a branch of the anterior cerebral artery with draining from the frontal basal vein (arrow). Aneurysms in the internal carotid artery at the origin of the posterior communicating artery and anterior choroidal arteries and a pericallosal aneurysm. (b) Oblique view of the angiography showing a PAVF. (c) Side view of the angiography in arterial phase showing a PAVF fed by the anterior cerebral artery. (d) Side view of the angiography in venous phase showing dilated frontal basal vein due to a PAVF. (e) 3D angiography showing aneurysm of the anterior communicating artery

Case 2

A 3-year-old child, manifesting a progressive loss of strength on the left side of the body over a 6-month period was hospitalized. Neurological examination revealed mild left spastic hemiparesis.

A CT brain scan indicated a frontal-temporal cortical hypotrophy on the right side and a dilated vascular image in the right anterior frontal region suggesting a cerebral AVM [Figure 4a]. Cerebral angiography showed a PAVF in the anterior frontal region, fed by a branch of the right middle cerebral artery, which drains into the Trolard vein. A large variceal dilatation of the proximal venous and was noted in association with the PAVF [Figure 4b].

The patient was submitted to a frontal temporal sphenoidal craniotomy on the right side and microsurgical dissection of the Sylvian fissure. The variceal dilatation also caused significant mass effect due to very hard and thick wall. Thus, the PAVF was treated by occlusion of the feeding artery. Resection of variceal dilatation was also performed due to mass effect [Figure 5]. The patient recovered without complications at the postoperative stage and was discharged. A postoperative control angiography was not performed as the parents withheld their consent. The child was monitored for 3 years, and manifested a total recovery from hemiparesis and exhibited normal psychomotor development.

DISCUSSION

Many types of cerebrovascular lesions occur in the central nervous system, each one receiving a different name according to its morphological features. If there are abnormal vessels between the two systems (arterial and venous), they are called nidus and are considered an

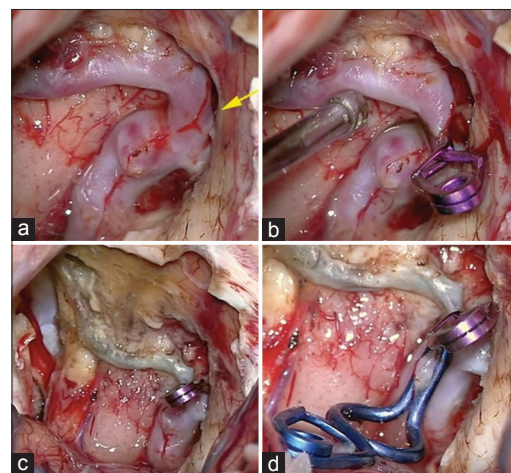


Figure 2: Surgical picture of a PAVF. (a) The fistulous communication between the anterior cerebral artery and frontal basal vein (Yellow Arrow). (b and c) The occlusion of the arteriovenous shunt with an aneurysm clip. (d) The occlusion of the arteriovenous shunt and pericalosa aneurysm

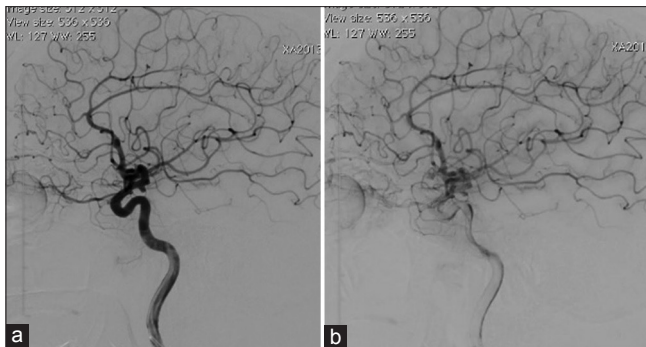


Figure 3: Postoperative angiography showing complete exclusion of a PAVF. (a) Side view of the angiography in arterial phase. (b) Side view of the angiography in cortical phase

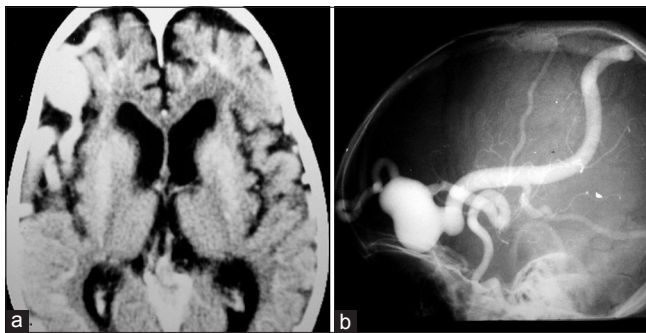


Figure 4: (a) Brain CT showing a right frontal temporal cortical hipotrophy and extensive vascular dilation in the right anterior frontal region. (b) Cerebral angiography showing a PAVF in the anterior frontal region fed by a branch of the right middle cerebral artery and drained by the swollen Trolard vein with large venous aneurysm

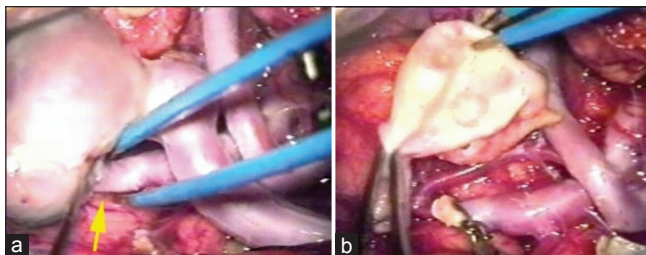


Figure 5: Surgical Picture of a PAVF. (a) Dilatation of the middle cerebral artery in direct communication with venous system (yellow arrow). (b) The surgical clipping of the feeding artery and the resection of the venous dilation

AVM. If there is no nidus the condition is referred to as arteriovenous fistula (AVF), which can be pial or dural. If the blood vessels are not related to the duramater and come from the cortical and/or pial regions, they are called PAVF.^[9] Due to the high blood flow supplied by the arterial network, the formation of varicose regions, adjacent to the area of abnormal communication, may possibly occur.^[10] The present study reported two cases of PAVF treated by microsurgery. In the first case, the PAVF was a direct communication of a branch of the right anterior cerebral artery to the frontal basal vein; and in the second case, the PAVF was secondary to a communication between the

right middle cerebral artery to the Trolard vein and was associated with a large venous varix.

The pathophysiology of PAVF includes congenital, traumatic, and iatrogenic etiologies.^[8] Paramasivam *et al.*^[6] proposed that congenital PAVFs result from the persistence of fistulous connections between pial arteries and veins during the earliest stage of embryological vascular development. Such fistulas are common in transient stages during vascular genesis, and regress with development of a capillary network and with vessel wall maturity. The incidence of PAVF varies in the literature from 1.6% to 7.3% of intracranial vascular lesions in clinical studies.^[6,11,12]

The natural history is unfavorable, especially in children aged less than 2 years, and patients with multiple feeding vessels. The mortality rate can reach 63% in cases with conservative treatment.^[13] Yang *et al.*^[7] reviewed the literature (1977–2009) on PAVFs and found a total of 83 eligible cases for study. The patient's age range from 12 weeks to 65 years (mean age 20.2 years). There were 39 (47%) female and 44 (53%) male patients. The most common clinical presentation was mass effect, seizure, heart failure, or hemorrhage. Mass effect was defined by headache, focal neurological deficit, or increasing head circumference. The clinical presentation mass effect was found in 38 (45.8%) patients, hemorrhage in 27 (32.5%) patients, seizure in 23 (27.7%) patients, and heart failure in 3 (3.6%) patients. Pediatric patients are likely to have seizure as an initial presentation, while adults are more likely to have hemorrhage. Varix was present in 64 (77.1%) patients, and it was found more frequently in pediatric patients (39 patients, 90.6%) than adults (25 patients, 62.5%). The absence of varix had significant correlation with hemorrhage,^[7,14] only 15 of 64 (23.4%) patients with varix presented a bleeding episode, while 12 of 19 (63.1%) patients without varix presented a bleeding event. Surgical treatment was performed in 31 (37.3%) patients, with total obliteration in 30 patients (96.8%). Endovascular treatment was performed in 37 (44.7%) patients, with total obliteration in 37 patients (86.5%). Fourteen (16.8%) patients underwent hybrid endovascular surgical treatment, with total obliteration in 12 patients (85.7%).^[7]

Madsen *et al.*^[12] reviewed the literature on pediatric PAVFs and found a total of 147 eligible cases for study. The mean age of patients was 5.02 years (0 days to 17 years old) and 59.9% of them were male. The most common presenting clinical manifestations were seizure (24.5%), followed by congestive heart failure (19%) and neurological deficit (18.4%). Varix was found in 110 (74.8%) cases. Twenty-three (15.6%) patients were treated with surgical intervention and 113 (76.8%) with endovascular embolization. Of the 108 patients who underwent embolization as their initial

treatment, 74 (68.5%) had angiographic evidence of complete obliteration.^[12]

The treatment of PAVF consists of an occlusion of the supply vessel. The endovascular procedure has been considered less invasive and is adopted as the treatment of choice by many authors.^[6] The embolic agents used preferentially for endovascular therapy are detachable coils and N-butyl-2-cyanoacrylate (NBCA). In single fistula, with single artery-single vein arteriovenous (AV) shunt, the placement of coils is feasible with the advantage of a better controlled positioning of the embolic material, since it can be withdrawn several times before being detached. In complex fistulas, in which there are many AV communications or multiple arterial feeders with single vein outlet, use of NBCA is preferred due to its capability of filling all the AV connections completely and especially the micro AV connections. Onyx embolization is another option for high flow fistulas, because offers the facility to redirect flow during the delivery.^[6,15,16]

Despite the vast majority of patients with PAFV were treated with endovascular approach, the choice of the treatment should be multidisciplinary and based on the experience of the medical team. It is important to evaluate the location and degree of lesion complexity.^[8] We believe that PAVF, with a complex configuration of supply arteries, multiple arterial connections, and high flow in areas of easy surgical access, are cases in which the surgical approach has greater benefits. And in cases with a single feeding artery in eloquent areas, the endovascular approach is preferable.^[8,11] The goals of therapy are to preserve normal neurocognitive development in children and provide protection from hemorrhage and progressive neurologic deficits in children and adults.

CONCLUSIONS

PAVF is a very rare disease. This clinical presentation is of vital importance in early diagnosis. The treatment of this condition consists of an occlusion of the supply vessel and the possible approaches employed are microsurgical,

endovascular, or combined treatment. Both the cases were successfully treated by microsurgical procedure.

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