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ORIGINAL PAPER

Intracranial pial fistulas in pediatric population. Clinical features and treatment modalities

Flavio Requejo[1] · **Roberto Jaimovich**[2] · **Juan Marelli**[1] · **Graciela Zuccaro**[2]

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Abstract
Purpose The purpose of the study is to describe the clinical manifestations and treatment modalities of patients having intracranial pial arteriovenous fistulas (PAVFs).
Methods We retrospectively analyzed the cases of PAVFs from January 2004 to December 2013. Medical charts, diagnostic images, surgical, and endovascular reports were reviewed retrospectively during each of the procedures and follow-up. We recorded patient demographics, clinical presentation, treatment modalities, and outcome.
Results Ten patients with single PAVFs were identified, one of them with multiple holes. The median age was 7.5 years old (20 days to 14 years). Six patients were male (60 % of cases). Four PAVFs were localized in the posterior fossa, and six were supratentorial (60 %). Two patients had intracranial bleeding, three presented seizures, one was studied for chronic headaches, three manifested by growth retardation, one had hydrocephalus, and one had a congestive heart failure (CHF) and vein of Galen aneurysmal malformation (VGAM). The latter did not improve after embolization and died few days later. Endovascular therapy was used in eight, whereas two patients were surgically managed. Total occlusion of the fistula was achieved in all cases.
Conclusions PAVF affects pediatric population at different ages with miscellaneous clinical manifestations. Endovascular treatment is safe and effective when the venous side of the fistula can be occluded.

Keywords Pial arteriovenous fistula · Intracranial shunt · Intracranial hemorrhage · Endovascular treatment

✉ Flavio Requejo
frequejo@intramed.net

[1] Department of Interventional Radiology, Hospital Nacional de Pediatria J P Garrahan, Combate de los Pozos 1881 (1254), Buenos Aires, Argentina

[2] Department of Neurosurgery, Hospital Nacional de Pediatria J P Garrahan, Combate de los Pozos 1881 (1254), Buenos Aires, Argentina

Introduction

Pial arteriovenous fistulas (PAVFs) are a group of infrequent entities affecting predominantly the pediatric population [2–4, 14]. They are a high-flow shunt communicating one or more pial arteries with a single vein without an intervening nidus, located in the subpial space [1, 3, 14].

PAVFs are congenital in nature in most cases, but may be originated by brain ischemia, head trauma, cranial surgery, or after a cortical vein thrombosis [7, 8, 11]. They could be treated by endovascular embolization or open surgery [10, 12].

We describe our experience with patients having PAVF regarding their clinical manifestations and the treatment modalities.

Material and methods

We retrospectively analyzed the cases of PAVF in our hospital, a tertiary pediatric center, from January 2004 to December 2013.

We consider PAVF a pathological entity consisting of one or more direct high-flow arteriovenous connections situated in the subpial space with no intervening capillary bed or a tangle of vessels (nidus). PAVFs are considered single and multiple. Single PAVFs are formed by one (single hole) or more (multiple holes) arterial feeders communicating with a single vein. Multiple PAVFs are anatomically distant from each other;

therefore, distinct veins drain the arterial feeders. Pial arteriovenous malformations (AVM), dural arteriovenous fistulas, and vein of Galen aneurysmal malformations (VGAM) were excluded from the study.

Medical charts, digital subtraction angiography (DSA), cranial tomography (CT) scan, CT angiography and magnetic resonance imaging (MRI), and surgical and endovascular reports were reviewed retrospectively during each of the procedures and follow-up. We recorded the patient demographics, their clinical presentation, treatment modality, and outcome (Table 1). All cases underwent a DSA at least 6 months after treatment. The only exception was the patient no. 9 who died of congestive heart failure (CHF).

The modality of treatment, endovascular or microsurgery, was discussed in every case in a multidisciplinary meeting conformed by vascular neurosurgeons and neuroradiologists.

Results

Results are summarized in Table 1.

From January 2004 to December 2013, ten patients were identified as meeting the inclusion criteria for PAVF in our hospital. They are recorded in Table 1 reporting their age, gender, accompanying vascular disease, treatment modality, clinical manifestations, and outcome.

All cases were single PAVF, nine single hole and one multiple holes (patient no. 10).

In two patients, PAVF was accompanied by other vascular disease. In case no. 8, a frontal AVM fed by branches of the ACA and MCA surrounded the PAVF conformed by a communication between a persistent primitive olfactory artery (PPOA) and a cortical vein (Fig. 1). In case no. 9, the baby has a PAVF fed by a vertebral artery (VA) branch and a VGAM.

The median age of patients was 7.5 years. The youngest patient was a 20-day-old baby boy, and the oldest was a 14-year-old boy.

Six patients were male (60 % of cases). Four PAVFs were localized in the posterior fossa, and six were supratentorial (60 %).

Two patients had intracranial bleeding, three presented seizures, one was studied for chronic headaches, three manifested by growth retardation, one had hydrocephalus, and one was a baby who was born with congenital CHF (and VGAM).

Treatment

Eight patients were treated by endovascular route, two with *N*-2-butyl cyanoacrylate (NBCA) alone, four with coils alone, and two with both coils and NBCA. The remaining two cases were surgically managed.

Table 1 Features of patients having PAVFs

Case	Age	G	Signs and symptoms	Localization	Feeding artery	Treatment	Result	Outcome
1	13 y	F	Acute headache Intracerebral and intraventricular bleeding	Right parietal	Sylvian branch	Embolization NBCA	Total occlusion	No deficit
2	14 y	M	Seizures	Right frontal	Pericallosal artery	Surgical	Total occlusion	Seizure free without medication
3	40 d	M	Intracranial hypertension Hemorrhagic round mass	Right temporal	Posterior cerebral artery branch	Embolization NBCA	Total occlusion	No deficit
4	6 m	F	Hydrocephalus Growth retardation	Left frontal	Branches of anterior cerebral and sylvian artery	Surgical	Total occlusion	No changes
5	5 y	M	Seizures	Left parietal	Pericallosal artery	Embolization NBCA–coils (2 sessions)	Total occlusion	Seizure free with medication
6	12 y	M	Growth retardation	Medulla	PICA	Embolization Coils	Total occlusion	No changes
7	8 y	F	Chronic headache	Cerebellar	Left superior cerebellar artery	Embolization Coils–NBCA	Total occlusion	Asymptomatic
8	13 y	M	Seizures AVM frontal lobe	Right frontal	PPOA	Embolization Coils–NBCA	Total occlusion	Seizure free without drugs
9	20 d	F	CHF VGAM	Medulla	VA	Embolization Coils	Total occlusion	Dead
10	10 y	M	Growth retardation	Cerebellar	VA	Embolization Coils	Total occlusion	No changes

AVM arteriovenous malformation, *d* days, *m* months, *ICH* intracranial hemorrhage, *CHF* congestive heart failure, *MCA* middle cerebral artery, *PA* pericallosal artery, *PCA* posterior cerebral artery, *PPOA* primitive olfactory artery, *VA* vertebral artery, *VGAM* vein of Galen aneurysmal malformation, *G* gender, *y* years

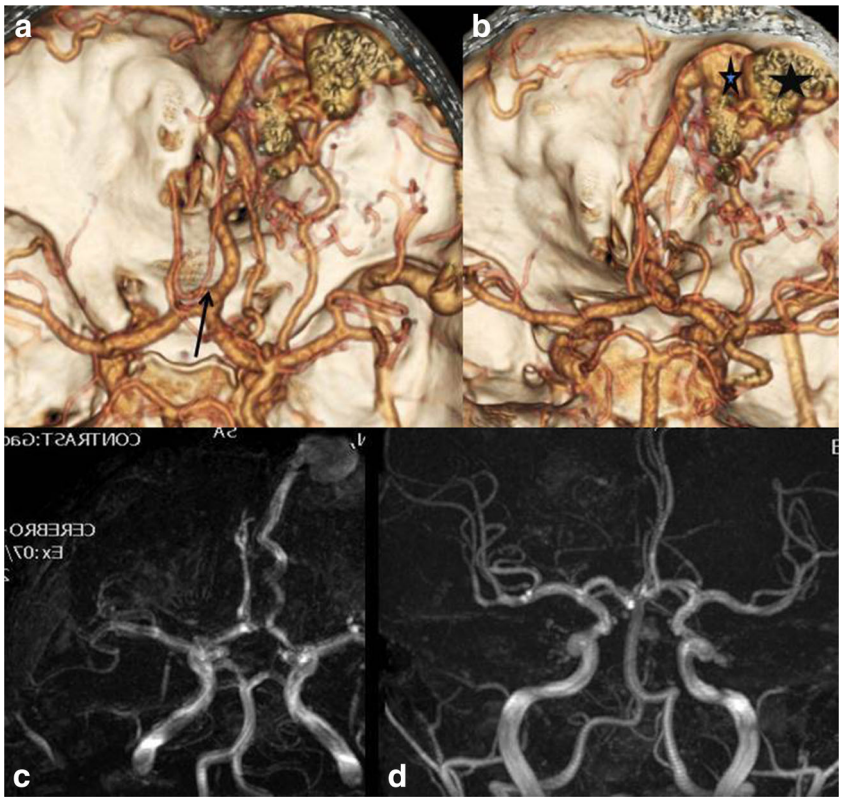

Fig. 1 Case no. 8. PAVF arising from a primitive olfactory artery. **a**, **b** CT angiography reconstructed in work station. An anomalous vessel originating with a common trunk with a smaller A2 (*arrow*) from the A1 segment of the right anterior communicating artery and shunting in a varix (*stars*) can be noted. This is a primitive olfactory artery (POA). Around the varix (not seen), there is an AVM fed by branches of distal ACA and MCA; the draining veins have no communication with the varix. **c** Angiography MRI. **d** Angiography MRI post occlusion of PAVF

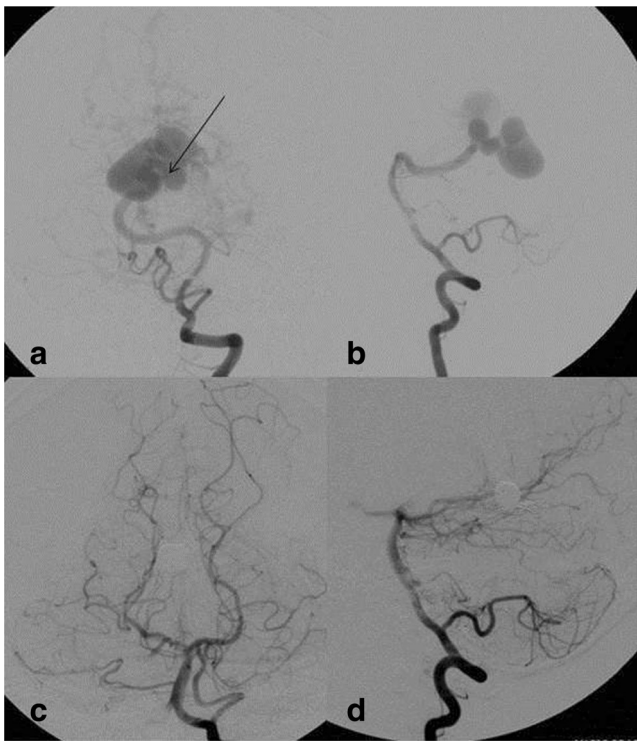

Fig. 2 Case no. 7. DSA showing PAVF arising from the right superior cerebellar artery. **a**, **b** Towne and lateral projection, respectively. The *arrow* points the transition from the artery into the vein. **c**, **d** Follow-up. PAVF was totally occluded using coils and NBCA in the venous side of the shunt. The right superior cerebellar artery is much smaller than before and both PCAs are now visible

Total occlusion of the fistula, confirmed by follow-up DSA, was achieved in all cases.

Discussion

In the last years appeared many articles focusing on the PAVFs [1, 3–16]. One of the largest series of patients was published by Lasjaunias and his coworkers [3, 14]. These authors pointed out the difference between PAVFs and AVMs considering them as two different entities. Because the rarity of this disease, several researchers had to review literature in order to collect a larger number of patients to acquire more reliable information [1, 4, 15].

PAVFs are conformed by a direct arteriovenous connection located in the subpial space [3]. Most of the arteries feeding the PAVF open into a single ectatic vein frequently associated with large to giant venous varix [9]. They are single when one (single hole) or more arteries (multiple holes) communicate with a single vein. PAVFs are considered multiple if two or more arteriovenous fistulas are situated in different anatomical locations having separate veins draining the fistulas [3, 9].

PAVFs account for 4.7 % of pial AVMs [13], reaching 17, 2 % incidence in the pediatric age group [14].

Clinical manifestations

Clinical manifestations are due mainly to the arterialized blood circulating in cortical veins compromising the normal drainage of the brain. The shunt can lead to enlargement of veins, causing in some patients epilepsy and mass effect. The veins can also rupture leading to intracranial hemorrhage (intracerebral, intraventricular, and subarachnoid). If the shunt is big and/or is situated near a dural sinus, PAVFs are prone to produce a rise of the intracranial pressure [4]. In patients before 1 year of age, the venous hypertension can alter the normal circulation of the cerebrospinal fluid (CSF) causing macrocrania and/or hydrocephalus [3, 15]. In the newborn, these shunts can cause CHF [3].

In a series of 52 patients, the male-to-female ratio was 28:13; the most common symptoms were CHF, epilepsy, and macrocrania. Half of patients with seizures had intracranial hemorrhage. Other clinical manifestations were growth retardation, exophthalmos, headache, and epistaxis [14].

Some cases deserve special consideration. The case no. 3 was a 40-day-old baby who presented a hemorrhagic round mass in the temporal lobe causing intracranial hypertension. At surgery, the mass under the clot was actually a vascular structure. The patient was transported to the angiosuite. The angiography revealed a single PAVF formed by a temporal branch of the PCA communicating with a venous varix in the temporal lobe. The shunt was totally occluded with NBCA. This case is similar to the one published by Kraneburg et al. [2]. Case no. 8 was a 13-year-old boy with seizures. CT angiography (Fig. 1) and DSA revealed a PAVF fed by persistent primitive olfactory artery (PPOA) arising from the A1 segment of the right anterior communicating artery (AcoA).

It is evident in our patients and in other series that signs and symptoms are multiple and varied. Diagnostic images like CT, CT angiography, and MRI are very useful to make a correct diagnosis of the disease (Fig. 1). In our institution, we use the DSA as a tool for confirming diagnosis of PAVF and performing the endovascular treatment in the same session.

Treatment

Even with modern endovascular techniques and advancement in microsurgery, PAVFs is still a dangerous illness [4]. Due to the high mortality and bad neurocognitive prognosis, the occlusion of the arteriovenous shunt is necessary [3, 5, 12].

The first choice of treatment in our hospital is the endovascular route. The exploration of the angioarchitecture of the PAVF with microcatheters allows a precise visualization of the arterial and venous part of the fistula, even when the vein is enlarged (varix) as frequently occurs. The dilated, arterialized vein obstructs the surgical field making the

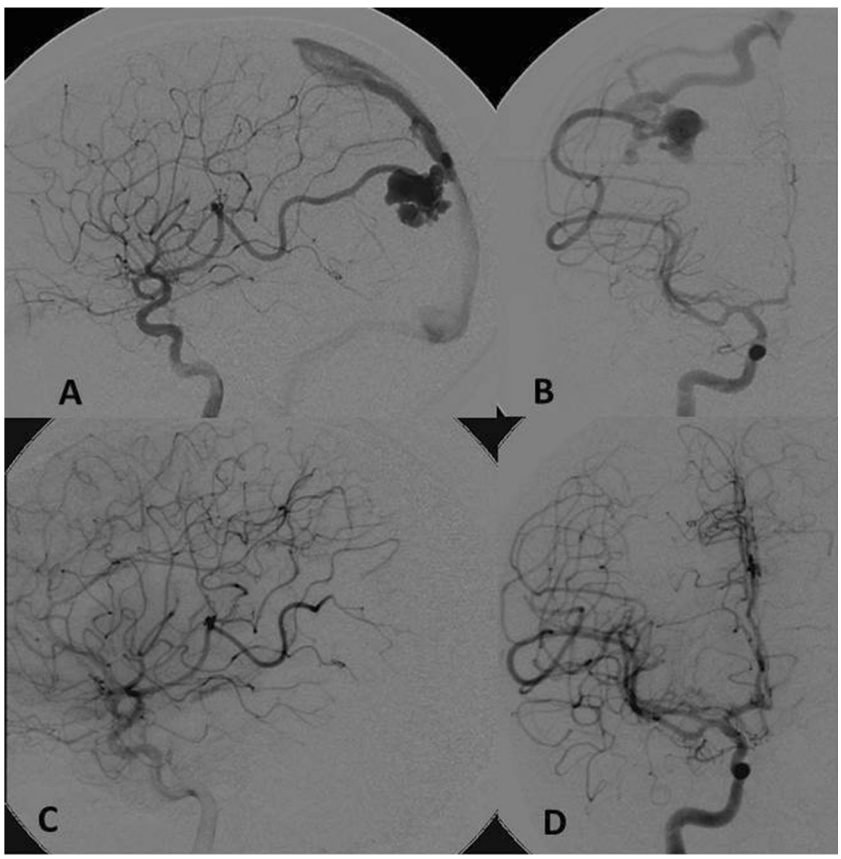

Fig. 3 Case no. 1. **a**, **b** DSA. PAVF arising from the angular artery branch of the MCA. **c**, **d** Totally occluded shunt after embolization with NBCA. Note that the angular artery is now smaller than before when the fistula was patent

procedure more difficult. The knowledge of the angioarchitecture of the shunt is essential to undertake a successful procedure.

The key point to succeed in achieving total occlusion of PAVFs is the closure of the venous portion of the arteriovenous shunt. Our preferred strategy is coiling the venous segment proximal to the fistula first; by doing so, we are sure that all the possible connections are sealed even with the presence of multiple arterial feeders (multiple holes). Thereafter, we put coils and/or glue in the arterial part of the fistula (Fig. 2). The mesh of coils creates a barrier impeding the undue migration of the NBCA [6]. NBCA alone is preferred in cases of small size arteries (Fig. 3).

Surgery is reserved for those cases where embolization is deemed too dangerous because the arterial feeder is a short branch of a cortical artery which could not be occluded as occurred in patients 2 and 4. Sometimes, the varix must be removed because it causes mass effect or seizures. We isolate and clip the feeding artery as close as possible to the shunt, and the venous side is coagulated in order to be sure that even a multiple-hole fistula is closed [12] Fig. 4.

Follow-up is mandatory to look for recanalization, reactive angiogenesis, and de-novo dural AVF development [9]. We have not seen any of these phenomenons in the follow-up of our patients.

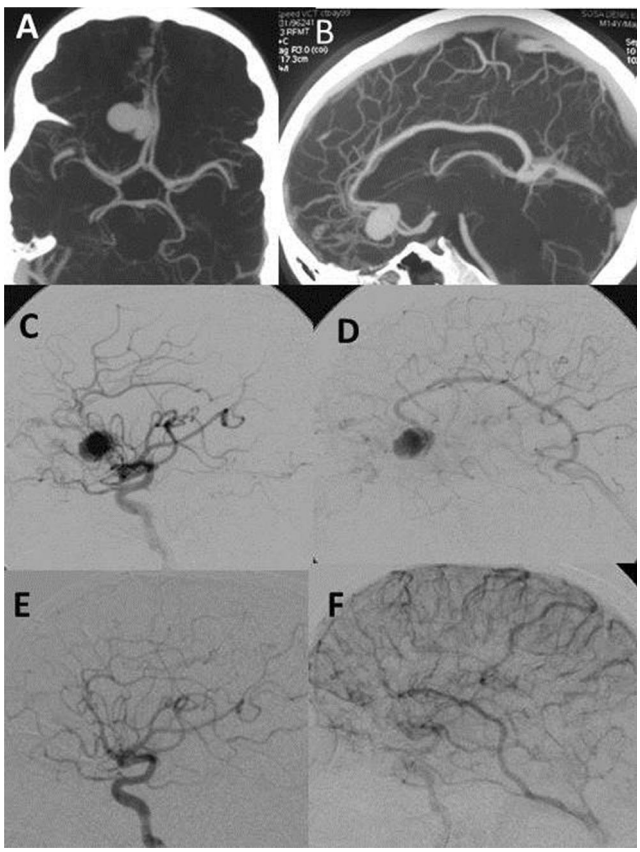

Fig. 4 Case no. 2. **a**, **b** CT angiography. Axial and lateral views. A dilated vein (varix) in the midline of the frontal lobe is seen. The pericallosal artery feeds the PAVF, and the varix drains in a dilated pericallosal vein. **c** Anteroposterior and **d** lateral projection of a DSA depicting the PAVF, the varix, and the pericallosal vein. It was decided to remove the varix which is responsible for the seizures by microsurgery. **d**, **e** Postoperative DSA

Conclusions

PAVF affects pediatric patients of different ages. Even with this short number of patients, it is ostensible that clinical manifestations of this entity are varied. Endovascular treatment is safe and effective when the venous side of the fistula can be occluded. The weakness of the article is the lack of neurocognitive functions evaluation, an important tool to have a reliable assess of treatment.

Conflict of interest There are no conflict of interest.

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