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Original Article

Pial arteriovenous fistulas: two pediatric cases and a literature review

Lei Feng*, Yunzhen Liu*, Jun Liu, Chuanfeng Lv, Chunhai Su

*Department of Neurosurgery, The First People's Hospital of Jining, Jining 272011, Shandong, China. *Equal contributors.*

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Abstract: Pial arteriovenous fistulas (AVFs) of the brain are rare vascular malformations associated with significant risk of hemorrhage and neurological deficits. Depending on their location and high-flow dynamics, these lesions can present treatment challenges for both endovascular and open cerebrovascular surgeons. We present a novel approach to treat intracranial high-flow pial AVFs. A single-channel high-flow pial AVF was diagnosed in two children aged 4 and 12 years, respectively. The patients were treated with coil and/or embolization with balloon-assisted liquid emboli to ensure fistula closure. The pial AVFs were transarterially embolized via feeding arteries. Immediate and complete angiographic obliteration was achieved without procedural complications. At the 5-month follow-up, the simple discontinuation of blood flow resulted in a sustained obliteration of the fistula and the headaches disappeared. In conclusion, arterial feeder embolization represents a potentially effective novel treatment for the rare occurrence of high-flow pial AVFs. The liquid embolism agent was successfully delivered into high-flow lesions after flow arrest using coils and/or a balloon combination, to enable minimally invasive and durable treatment of pial AVFs.

Keywords: Balloon-assisted embolization, coil embolization, pial arteriovenous fistula, vascular disorders, intracranial arteriovenous shunt

Introduction

Intracranial pial arteriovenous fistulas (AVFs) are rare vascular lesions that account for 1-5% of all brain vascular malformations [1-6]. To date, fewer than 150 cases of pial AVFs have been reported. The natural history and developmental mechanisms of these lesions remain unclear [7, 8]; however, AVFs can be congenital or acquired.

Pial AVFs consist of one or more arterial feeding vessels derived from cortical or pial arteries and a single venous channel that typically results in a substantial varix, with no intervening nidus [2, 3, 9, 10]. High shunt and deep venous drainage limit the safety and efficacy of surgical treatment [3, 9, 11], and therefore, endovascular embolization is typically preferred for difficult and surgically inaccessible lesions. Because of the low incidence of pial AVFs, there are limited studies regarding the use of embolization for this condition. The current case report describes two cases with embolization of high-flow pial AVFs and provides an overview of the existing literature and relative merits of endovascular versus surgical options for pial AVF treatment.

Illustrative cases

Informed consent for the operation was obtained from the patients' guardians. The study was approved by the Ethics Committee of the First People's Hospital of Jining.

Case 1

History and examination

A 12-year-old boy presented with progressive headaches and an acute onset of seizures. Computed tomography (CT) studies of the head demonstrated several large calcified masses in the left parietal lobe (**Figure 1**). Magnetic resonance imaging (MRI) and magnetic resonance

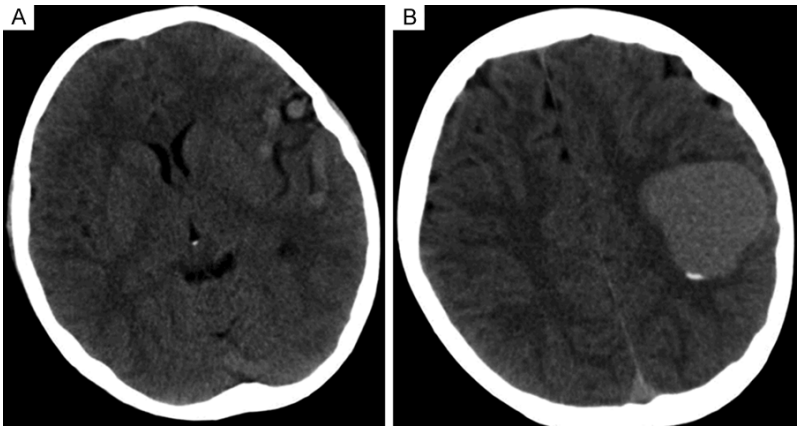

Figure 1. Case 1. Pre-embolization head computed tomography (CT) scans demonstrated large intraparenchymal calcified masses in the left parietal lobe.

angiography (MRA) (**Figure 2**) characterized the lesion as vascular. It demonstrated a single-channel, high-flow pial AVF with a large venous pouch.

Surgical technique

Embolization was performed while the patient was under general anesthesia. After femoral access was established, the remainder of the procedure was performed with full anticoagulation. Cerebral angiograms were obtained first, and they revealed a single-hole pial AVF that arose from the superior division of the left middle cerebral artery (MCA), stealing blood from the posterior circulation through the left posterior communicating artery (**Figure 3**). Venous drainage primarily occurred through the superior vein into the sagittal sinus with varices.

After the angio-architecture of the lesion was delineated, the diagnostic catheter was exchanged for a 6-Fr guiding catheter into the left internal carotid artery. A 4.0- × 10-mm Scepter C occlusion balloon catheter (MicroVention, Tustin, CA, USA) was advanced over a 0.014-in microguide wire and positioned in the feeding artery proximal to the fistulous point. The balloon was subsequently inflated in the feeding artery. An angiogram obtained through the guide catheter indicated that the balloon partially occluded the fistula. Detachable coils (EV3, Irvine, CA, USA) were carefully inserted distal to the exit of the feeding artery to sufficiently slow the arterial input and enable injection of Onyx-18 (EV3, Irvine, CA, USA); a reflux-hold-reinjection technique was used for occlusion of the fistula under roadmap visualization. The working catheter was primed with saline and dimethyl sulfoxide according to the manufacturer's specifications. Approximately 2.5 ml of Onyx-18 was infused into the feeding artery. The balloon was slowly deflated and the Onyx plug was carefully observed for stability. An angiogram obtained through the guiding catheter injection indicated that the fistula had been obliterated (**Figure 3**). The balloon was re-inflated, and the working microcatheter was withdrawn. Final angiography studies obtained after the microcatheter and balloon were removed demonstrated complete closure of the fistula without morbidity or mortality (**Figure 3**).

Postoperative course

Follow-up angiography at 5 months (**Figure 4**) confirmed a durable result. MRI at 5 months (**Figure 5**) indicated no appreciable changes in the mass because of the thrombosed varix with improvement in the patient's ventriculomegaly. The patient remained asymptomatic at the 9-month follow-up.

Case 2

History and examination

A 4-year-old boy developed progressive headaches and lethargy over the course of 3 months. A mass effect was identified on a cranial CT scan. The patient was subsequently transferred to our department.

Surgical technique

The surgical procedure was performed similar to that performed in the patient in Case 1. Cerebral angiograms were obtained, and an arterial pedicle feeding the fistula arising from the left posterior cerebral artery was identified. Venous drainage occurred mainly through the deep vein into the straight sinus with varices. Embolization was performed while the patient was under general anesthesia with full antico-

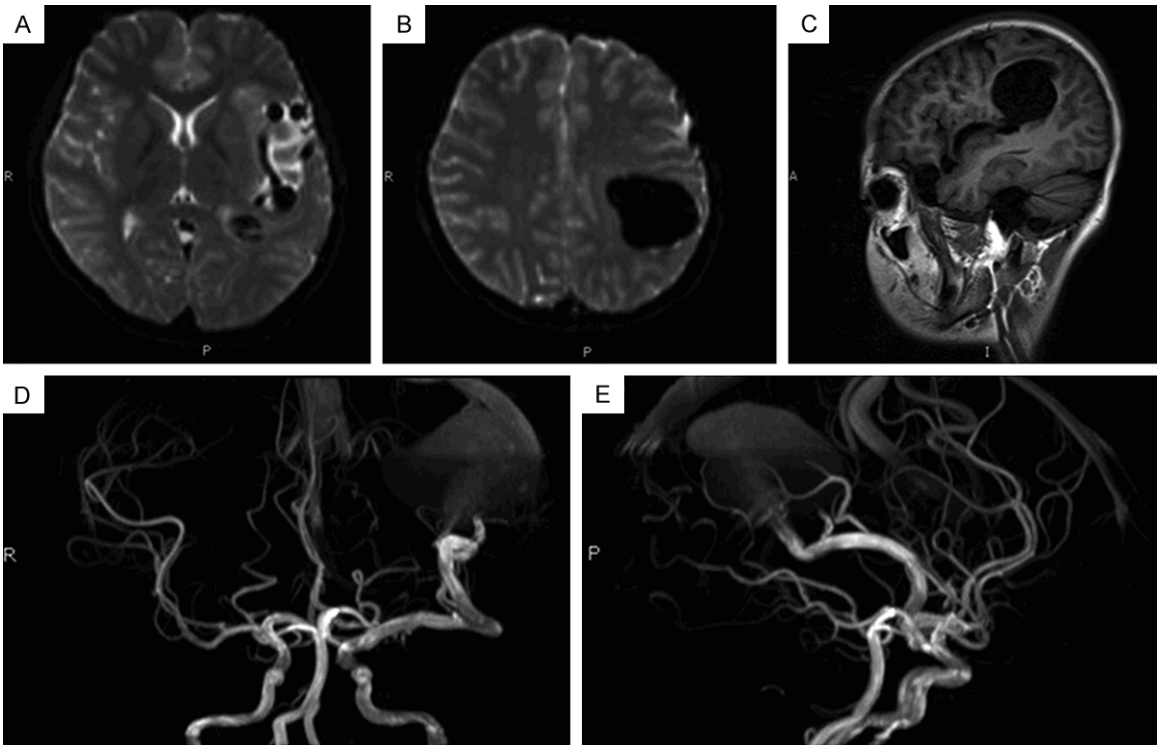

Figure 2. Case 1. Axial diffusion-weighted image (DWI) (A and B) and sagittal T1-weighted image (C) indicate enlarged flow voids in the left parietal lobe and superior vermian cisterns, which flow into a massively enlarged torcular. Maximum intensity projection of the AP (D) and lateral views (E) in magnetic resonance angiography (MRA) demonstrate an enlarged left cerebral arterial superior branch that provided inflow to an arteriovenous fistula (AVF) and converged in the left parietal lobe.

agulation. A 1.7-Fr Echelon-10 microcatheter (EV3, Irvine, CA, USA) was positioned in the feeding artery proximal to the fistulous ostium. Because of the high-flow shunt, it was difficult to deliver the detachable coils precisely and stably to the fistulous point. Oversized coils were placed in the venous pouch to reduce flow and act as a template for the feeding artery embolization. Detachable coils (EV3, Irvine, CA, USA) were carefully placed distal to the exit of the feeding artery to sufficiently slow the arterial input and enable the injection of 30% N-butyl cyanoacrylate (NBCA, Cordis Neurovascular, Miami, FL). Approximately 0.5 ml of 30% NBCA was infused into the feeding artery, after which the Onyx cast was considered stable. An angiogram obtained through a guiding catheter injection demonstrated the obliteration of the fistula (**Figure 6**).

Postoperative course

Because of the uneventful presentation, the patient did not return for follow-up. A telephone follow-up indicated normal development 6 months after treatment.

Discussion

Pial AVFs are rare lesional vascular anomalies and are considered a distinct pathological entity among brain arterovenous malformations (AVMs), which are common congenital diseases. Most arteries that feed the pial AVF open into a single ectatic draining vein without a capillary bed or “nidus” [10]. This anatomical feature creates conditions for rapid high flow, which is associated with substantial venous varices. Their presence is determined by the high flow across the shunt against the venous outflow obstruction.

Most fistulas are typically diagnosed during the first 5 years of life [5, 6]. When managed conservatively, these lesions can cause seizures, headache, hemorrhage, high-output cardiac failure in neonates and infants, macrocephaly, neurological deficits, and symptoms of increased intracranial pressure [4, 8, 10]. Gamma-

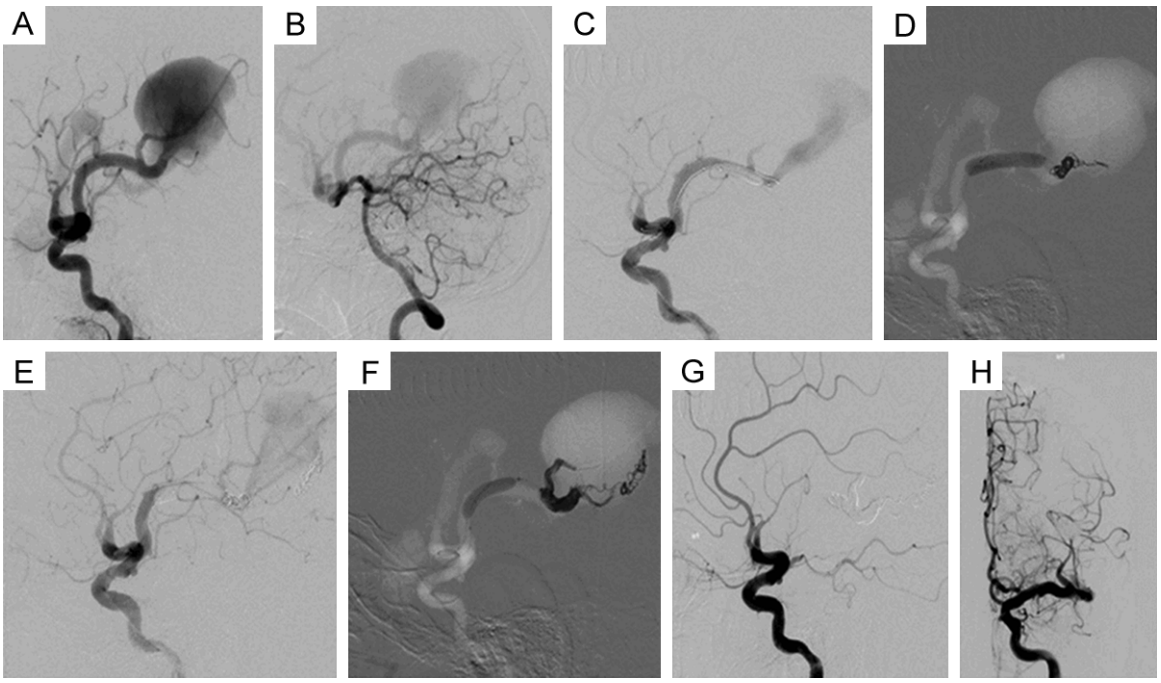

Figure 3. Case 1. Cerebral angiogram in the AP (A) projection of the left carotid artery and the lateral (B) view of the left vertebral artery in the early arterial phase indicate a pAVF with an obvious high-flow shunt and dilated venous sac. The pial AVF stole blood from the posterior circulation through the posterior communicating artery (B). (C) Flow control was attained after balloon inflation. (D-F) The pial AVF underwent successful endovascular embolization with onyx after a balloon was employed and detachable coils attained the blood flow. The Onyx cast is shown between the portion of the balloon and the orphaned coil mass. Postembolization angiogram of the (G) lateral and (H) frontal views of the left carotid artery injection confirmed obliteration of the fistula. No early venous opacification was present.

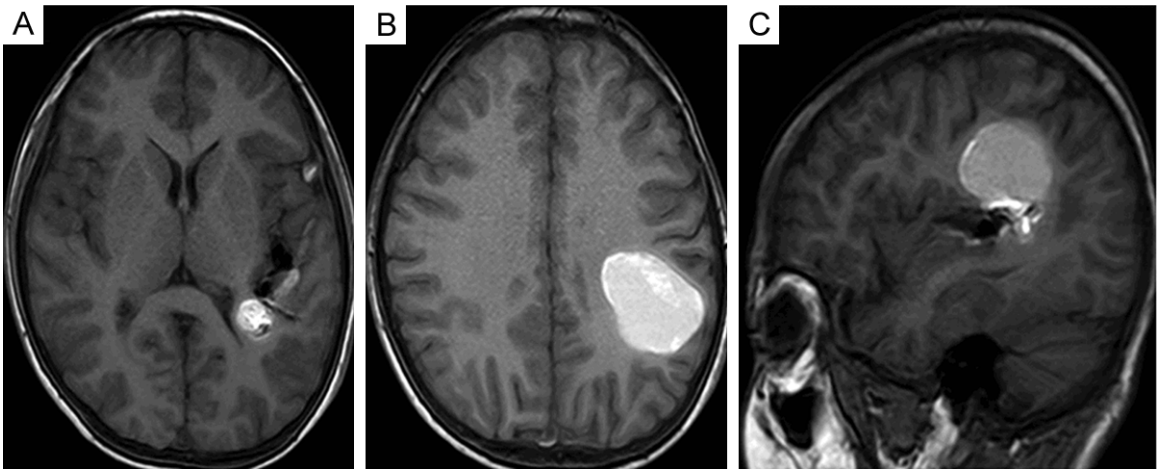

Figure 4. Follow-up magnetic resonance (MR) image (A-C) obtained at 5 months in Case 1, which indicates thrombosis, no progression of the varix, and a stable mass effect.

knife surgery is ineffective for high-flow AVFs [12]. The optimal treatment option for highflow pial AVF is controversial and may include direct surgical disconnection, endovascular embolization, or a combination of surgical and endovascular procedures [8].

A literature review from 1977 to 2009 indicated that the success rate of surgical treatment (96.8%) was comparatively higher than the rate for endovascular treatment (86.5%) [13]. Srinivasan et al. suggested that this increased rate was a result of surgical treatment of select-

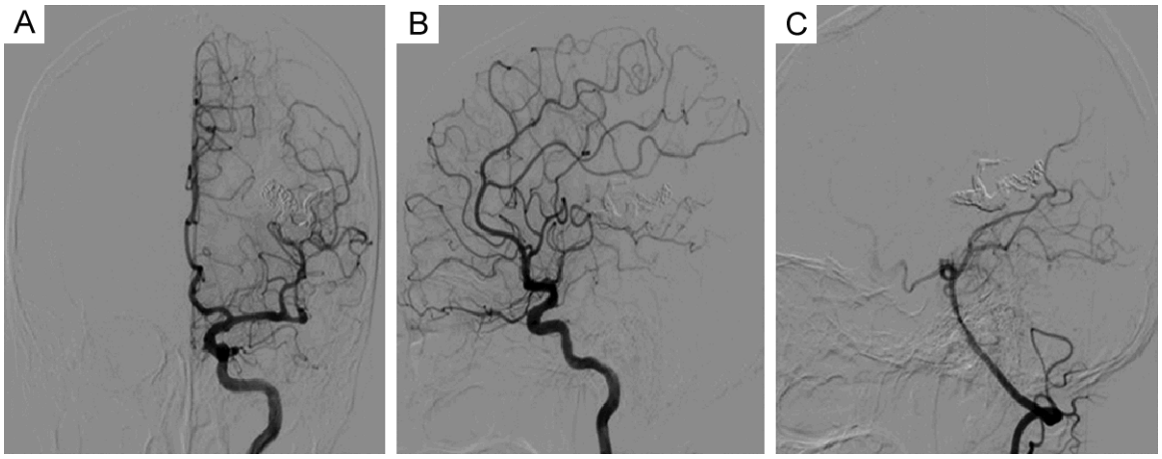

Figure 5. Case 1. Follow-up AP (A) and lateral (B) views of left carotid artery injection; the lateral view (C) of the vertebral artery injection at 5 months confirms obliteration of the fistula.

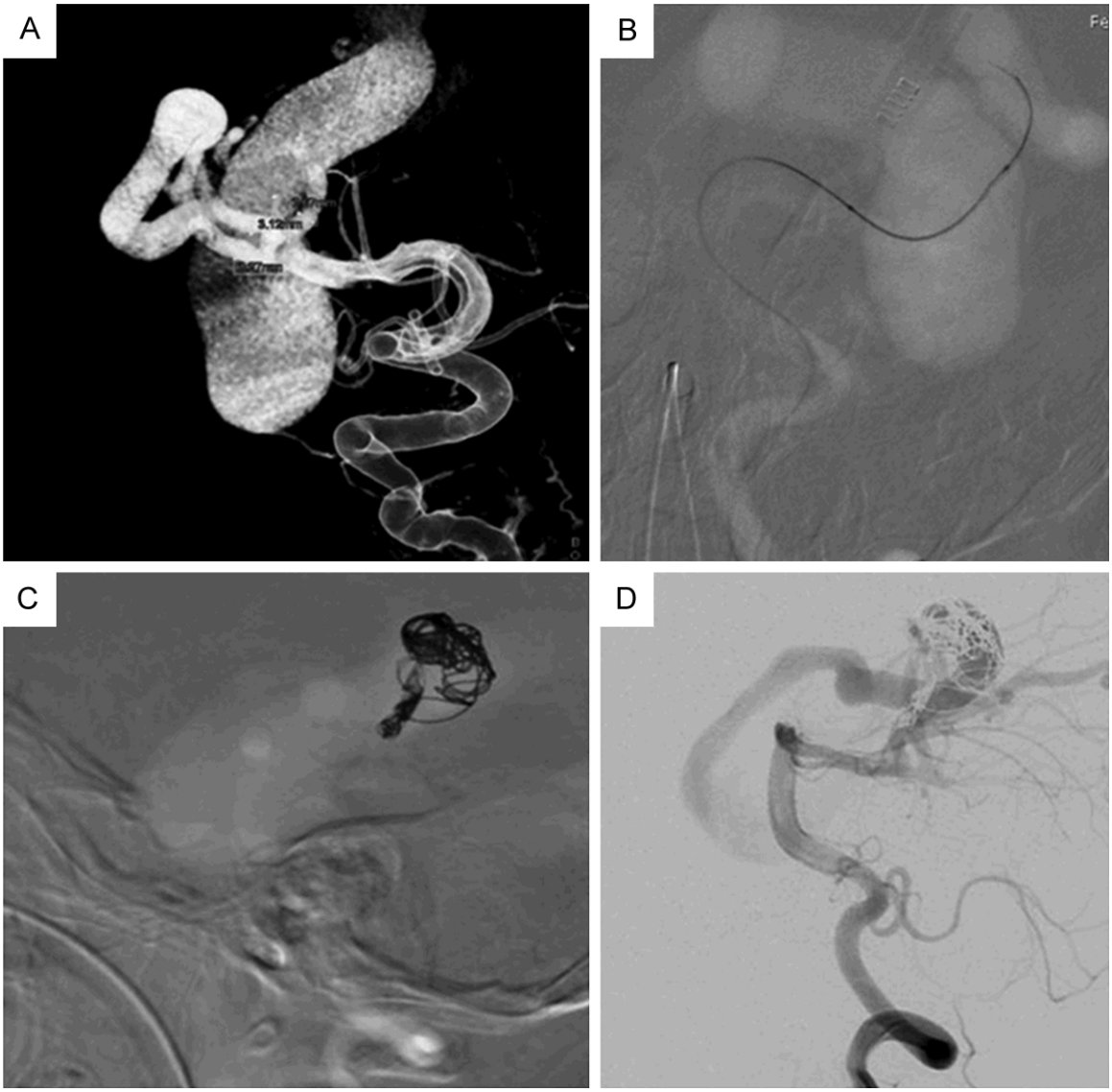

Figure 6. Cerebral angiograms of a 4-year-old boy with a pial arteriovenous fistula treated with transarterial embolization using detachable coils in Case 2. Anteroposterior (A) and lateral (B) left vertebral artery angiograms revealing feeding arteries from the left posterior cerebral artery before endovascular treatment. (C) Lateral view of the fluoroscopic image demonstrating the coil mass. (D) Angiogram of the left vertebral artery post-embolization reveals complete obliteration of the fistula.

ed superficial fistulas, with endovascular approaches reserved for the remaining cases [5]. Furthermore, the complex hemodynamics, deep lesions, eloquent localization, arterialization, and draining vein thickening result in surgically challenging lesions [11, 14, 15]. In addition, some authors have suggested that tiny feeding arteries, which are difficult to access, and the cumulative risk of embolization over multiple sessions, may exceed the risk of surgery, especially when considering the risk of exposure to radiation in young patients [6, 8]. Over the previous two decades, improvements in equipment, advances in endovascular techniques [7, 16], and the less invasive nature of the procedure have made embolization the preferred treatment for most pial AVFs, with surgery reserved for select cases [5, 8].

The goal of endovascular embolization should be obliteration of the fistula with as little embolic material as possible. However, high-flow lesions can make it difficult to precisely deliver the desired embolus to the fistulous point. Embolic material migration into the draining vein may compromise venous outflow of the fistula. It may also cause pulmonary embolism or, most dangerously, embolism of the draining vein, which may result in immediate hemorrhage via blockade of venous outflow [2, 11].

In general, NBCA or Onyx alone is not selected as an embolizer because it is difficult to control their flow and placement post-injection. They are associated with a risk of distal migration of the embolizer to venous drains, which may lead to catastrophic complications, particularly when the pial AVFs only exhibit single venous drains. Detachable coils and a balloon enable occlusion of the AVF so they can be relocated into a stable and safe position proximal to the fistula point or its proximal parent artery, which leads to total occlusion of the fistula. In addition, coils can be successfully implemented to modulate flow through high-flow AVFs and act as a template for the deposition of liquid embolic materials, such as NBCA and Onyx [2]. The liquid embolic material used is based on individual choice and experience. NBCA is delivered in a single injection with limited ability to redirect flow during the course of delivery. Onyx has the advantages of prolonged injection and more precise targeting and delivery to the fistulous point via a reflux-hold-reinjection technique [4].

In our cases, the high-flow shunt prevented delivery of detachable coils precisely and stably to the fistulous point. Our strategy consisted of adjuvant insertion of a balloon or coils into the venous pouch, close to the fistula to slow the flow. It enabled placement of the detachable coils in the feeding artery, the fistula point, to attain flow control. Oversized coils are preferred for placement in the feeding artery to reduce flow and enhance the effect of fistula occlusion. To decrease the coil number and cost, Onyx and NBCA were infused into the residual fistula after subtotal fistula occlusion in the pial AVFs. Slow, patient, and meticulous injection provides a maximally viscous, solid, and controllable cast to avoid antegrade progression of the embolic agent into the venous pouch. Reflux around the distal end of the balloon and the microcatheter should be avoided. If the feeding artery becomes occluded, the risk of Onyx plug migration should be minimal.

In addition, Srinivasan et al. reported that recanalization, reactive angiogenesis with reopening of the shunt, and de-novo dural AVFs were identified in 38% of cases that required additional treatment [17]. A follow-up angiogram was considered mandatory by these authors.

Several limitations of the current study must be considered when interpreting these findings. This study only included two cases, and thus, is not a controlled study. Endovascular management is challenging because of the large size of the afferent and efferent vessels associated with high-flow fistula. A modification of the technique may be necessary to precisely embolize the fistulous location. Proper analysis of the angio-architecture and decisions regarding the

choice and sequence of the embolic materials determine the outcome. Furthermore, the treatment is expensive.

In conclusion, pial AVFs can be safely occluded using endovascular techniques tailored to the specific anatomical configuration of the shunt. In our experience, transarterial endovascular embolization using a liquid embolic agent with a balloon or coils on the feeding artery to attain flow control is the treatment of choice and is associated with low morbidity. High-flow lesions can make it difficult to precisely deliver the desired embolus to the fistulous point.

To the best of our knowledge, this is the first report regarding the successful embolization of high-flow pial AVFs using a highly flexible and navigable scepter occlusion balloon catheter that allows for increased precision. Nevertheless, the adjuvant use of a coil, balloon, or a combination of a balloon and coil to assist endovascular embolization remains a viable treatment option. Decisions regarding the specific technique should be based on the individual patient characteristics and the comfort level of the surgeon.

Disclosure of conflict of interest

None.

Address correspondence to: Lei Feng, Department of Neurosurgery, The First People's Hospital of Jining, No. 6, Healthy Road, Jining 272011, Shandong, China. Tel: +86 537 2978882; Fax: +86 537 2978882; E-mail: fcndoc@126.com

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