Intracranial pial arteriovenous fistulas with single-vein drainage

Report of three cases and review of the literature

YI-CHOU WANG, M.D., HO-FAI WONG, M.D., AND YI-SHIAN YEH, M.D.
Departments of Neurosurgery and Radiology, Chang Gung University and Chang Gung Memorial Hospital, Taoyuan, Taiwan

Intracranial pial arteriovenous fistulas (AVFs) are rare vascular lesions, of which fewer than 90 cases have been reported in the literature. They are composed of one or more arterial feeding vessels and a single draining vein that usually result in a huge varix. Because of the high-flow shunting, a poor prognosis is associated with conservative treatment. Surgical or endovascular treatment also poses various challenges. The authors present the cases of three patients in whom N-butyl-2-cyanoacrylate–assisted embolization was performed. Outcome in all three cases was good. The necessity of staged procedures to obliterate the AVF is also discussed.

KEY WORDS • arteriovenous fistula • embolization • endovascular therapy • venous varix • pediatric neurosurgery

Case Reports

Case 1

History and Presentation. This 17-year-old boy suffered from cerebral palsy and a gait problem since his childhood and progressive hearing loss since 11 years of age. He had experienced headaches as well as slowly progressive left-limb atrophy and weakness in recent months. Cerebral MR imaging demonstrated hydrocephalus and large flow void lesions over the CPA bilaterally with compression of the brainstem (Fig. 1A). An MR angiogram revealed a huge varix over the posterior fossa.

On admission, cerebral angiography revealed a pial AVF arising from a branch of the BA as well as immensely dilated veins draining the fistula at the right CPA (Fig. 1B and C).

Operation. Because of the obstructive hydrocephalus, a shunt was immediately placed. A few days later, the embolization procedure was performed after induction of general anesthesia by using a stream-shaped tracker microcatheter (Target Therapeutic, Fremont, CA) via a No. 5 French femoral sheath and a guiding catheter. After super-selective treatment of the feeding artery with the microcatheter, a mixture of 80% NBCA (Histoacryl; Nycomed Ingenor, Paris, France) and 20% iodized oil (lipiodol; Guerbet AG, Zurich, Switzerland) was injected into the feeding pedicle, resulting in complete closure of the fistula.

Postoperative Course. The patient tolerated the procedure well and the right trunk weakness and headache subsided gradually. Six months later, cerebral angiography revealed stabilization without recanalization of the fistula (Fig. 1D).

Case 2

This 5-year-old girl was examined after falling. Cranial
CT scanning performed in the emergency department revealed a mass lesion over the right CPA. Cerebral MR imaging demonstrated a 1.5-cm mass lesion with flow void signal at the right CPA (Fig. 2A). Cerebral angiography disclosed an AVF arising from the right anterior inferior cerebellar artery and draining into the ecstatic perimesencephalic vein and the transverse sinus (Fig. 2B). Several days later, endovascular occlusion of the AVF was performed using the mixture of NBCA and iodized oil. The patient was symptom free and the AVF was completely occluded without recanalization, as revealed on follow-up cerebral angiography 8 months later (Fig. 2C).

Case 3

This 24-year-old woman without a history of trauma had been experiencing progressive headaches for several months. She presented to the emergency department with severe headache and seizures. A CT scan of the brain revealed intracerebral hemorrhage over the left temporal lobe, and MR imaging demonstrated a flow void lesion at left temporal base (Fig. 3A). Cerebral angiography revealed a high-flow AVF arising from two temporal branches of the posterior cerebral artery and draining into a large venous varix and the left transverse sinus (Fig. 3B and C). Four days later, endovascular embolization of both feeding arteries was conducted using the mixture of NBCA and iodized oil. Follow-up angiography revealed complete occlusion of the AVF and the patient recovered without sequela (Fig. 3D).

Discussion

Nongalenic intracranial AVFs are very rare. They were considered a variant of AVM for decades. Only recently have they been considered a different vascular lesion from AVM.15 They consist of one or more feeding arteries connecting to a single venous drainage without an intervening capillary bed. In a series of 320 AVMs reported by Halbach, et al.,16 only five cases (1.6%) were identified as pial AVFs. In a review of the literature by Hoh, et al.,12 there were a total of 88 cases of pial AVF reported in the literature between 1970 and 2001.

The pathophysiological cause of pial AVFs remains unknown. Congenital pial AVFs usually develop in childhood and are part of a syndrome such as Rendu-Osler-Weber8,8 or Klippel-Trenaunay-Weber19 syndromes. Some authors have also reported AVF and AVM in the same patient.20,25 Whether the AVF and the AVM were caused by the same pathophysiological process has not been illuminated.

Patients with pial AVFs usually present with headache, seizure, hemorrhage, or neurological deficits. Clinical manifestations of a pial AVF are closely related to the age of the patient at onset of symptoms.8,26 Enlarged head circumference, cranial erosion, and heart failure have been frequently reported in neonates and infants.7,25,26 The turbulence and increased pressure within draining veins may lead to the formation of giant varices. These dilated venous channels can exert significant mass effect, producing compression of adjacent structures and impairment of the cerebrospinal fluid pathway, as seen in our Case 1. Neurological deficits are probably caused by mass effect of the varix or by cerebral venous congestion and ischemia.8

The natural history and risk of bleeding associated with pial AVFs have not been thoroughly documented because of the insufficient number of cases. In studies of risk factors for bleeding in cases of AVM in which multiple factors were considered, a single drainage vein was implicated as the cause of the high bleeding rate.1,17 In another study conducted by Spetzler, et al.,23 the possibility of AVM bleeding was directly related to perfusion pressure. Because the single venous drainage and high perfusion pressure are both characteristics of the pial AVFs, the possibility of bleeding in patients with pial AVFs should be considered. In reviewing the literature, we found three patients25,26 who presented with minor symptoms, were treated conservatively, and were event free throughout the study period. Conservative treatment of symptomatic pa-
Intracranial pial arteriovenous fistulas

Patients with pial AVFs, however, has been associated with death in five (63%) of eight patients. Spontaneous closure of the fistula’s tract may occur but is unpredictable. Santosh, et al., have reported one case of spontaneous closure, and Garcia-Monaco, et al., also reported two cases of partial venous thrombosis that resulted in acute neurological deficits. Therefore, active treatment of symptomatic patients with pial AVFs may prevent further morbidity and mortality.

Resection of the lesion to obliterate the fistula and remove the varices has long been considered standard treatment in both AVM and the pial AVF surgery and has been reported by several authors. Successful treatment of the AVF by simply disconnecting the shunt either surgically or endovascularly, however, has been reported in several recent articles. The goal in the treatment of the pial AVF is obliteration of all the feeding arteries as close as possible to the fistula while leaving the venous drainage intact.

Disconnection of the AVF by placement of an aneurysm clip or cauterization of the vessels has been accomplished surgically. The large draining veins, however, can obscure and interfere with the exposure of the fistula. In addition, because of the shunt, arterialization and thickening of the draining veins may occur, making identification of the exact fistula site difficult. Although neuronavigation may be applied in such situations, some lesions may be located in deep or surgically inaccessible regions and the fistula site would not be reachable without the use of circulatory arrest.

Because surgical treatment of these lesions is difficult and often unsuccessful, endovascular therapy involving transarterial embolization of the fistula has become increasingly important. Endovascular embolization of the AVF can be conducted using balloons, silk suture, coils, glue, or polyvinyl alcohol with various rates of success. In 1983, Viñuela, et al., performed the first successful detachable balloon-assisted embolization of an AVF.
Because of the high perfusion pressure, however, application of the embolization material is very difficult, and distal migration is frequently reported. There are several reports detailing the migration of embolization materials such as balloons or coils into the varix, draining veins, or the lung. The anatomical appearance of the feeding arteries is also important. If the AVFs feeding artery and draining vein enlarge progressively from the proximal to distal areas in the absence of localized vascular narrowing at the site of the fistula, then embolization material would be difficult to apply in the distal end of the feeding artery. An adhesive agent, NBCA, has been widely used as an excellent embolization material for decades, although its application technique is challenging. When acrylic glue is used, the time for polymerization of the mixture and its delivery speed should suit the high-flow state of the fistula to avoid embolism-related complications. If the AVF flow is too rapid, a balloon may be inflated proximally in the artery’s pedicle to halt or decrease the flow and facilitate the polymerization of the NBCA (a technique originally described by White). In using NBCA in our three cases, the AVFs were successfully embolized without any complications.

Although successful treatment may result in the elimination of the shunt, the obliteration of longstanding high-flow fistulas may predispose to hemodynamic compromise in which the chronic hypoperfused and vasodilated surrounding brain is incapable of an autoregulatory response to the resulting increase in local perfusion pressure, the so-called normal perfusion pressure breakthrough phenomenon. It is true that bleeding or edema immediately after successful disconnection of an arteriovenous shunt has been reported by several observers. Therefore, the choice of a staged obliteration is often made when several feeding arteries coexist. In a study conducted by Giller, et al., however, two patients with multiple feeding arteries attached to a pial AVF underwent transcranial Doppler and single-photon emission CT scanning before and after treatment; no evidence of impaired autoregulatory response was found to support the existence of normal perfusion pressure breakthrough phenomenon. One possible explanation for postembolization hemorrhage is that there is always the risk of premature embolization of the venous channel instead of the arterial pedicle, which compromises the venous outflow of the fistula. The high perfusion pressure resulting from remaining feeding arteries may result in venous hypertension or a break in the venous wall, with subsequent hemorrhage. Therefore, instead of staged obliteration of the feeding arteries in our Case 3, they were occluded sequentially in one stage and no complication occurred.

Pial AVFs are rare and must be distinguished angiographically from an AVM, because the natural history and the therapeutic goal of these two disease processes are different. Because of the high mortality and morbidity rates associated with pial AVFs, total and definitive occlusion of a pial lesion is mandatory when possible. With careful planning and accurate insertion of the embolization material in the distal end of the feeding arteries, single-stage obliteration of the lesion can be performed safely. As the successful rate of percutaneous procedures increases, endovascular embolization should be considered a potential alternative to surgical intervention in patients with pial AVFs.

References

21. Santos H, Teasdale E, Molyneux A: Spontaneous closure of an
Intracranial pial arteriovenous fistulas


Manuscript received March 5, 2003.
Accepted in final form June 24, 2003.
Address reprint requests to: Ho-Fai Wong, M.D., Department of Neurosurgery, Chang Gung Memorial Hospital, 5 Fu-Shing Street, 333, Kweishan, Taoyuan, Taiwan. email: wang2@adm.cgmh.org.tw.

---

*J. Neurosurg: Pediatrics / Volume 100 / February, 2004*