

Case Report

Intracranial Pial Arteriovenous Fistulas

Ji Yeoun Lee, M.D.¹, Young-Je Son, M.D.², Jeong Eun Kim, M.D.¹*Department of Neurosurgery¹, Seoul National University Hospital, Seoul, Korea**Department of Neurosurgery², Seoul National University Boramae Hospital, Seoul, Korea*

Intracranial pial arteriovenous fistula (AVF) is a rare cerebrovascular lesion that has only recently been recognized as a distinct pathological entity. A 41-year-old woman (Patient 1) presented with the sudden development of an altered mental state. Brain CT showed an acute subdural hematoma. A red sylvian vein was found intraoperatively. A pial AVF was revealed on postoperative angiography, and surgical disconnection of the AVF was performed. A 10-year-old boy (Patient 2) presented with a 10-day history of paraparesis and urinary incontinence. Brain, spinal MRI and angiography revealed an intracranial pial AVF and a spinal perimedullary AVF. Endovascular embolization was performed for both lesions. The AVFs were completely obliterated in both patients. On follow-up, patient 1 reported having no difficulty in performing activities of daily living. Patient 2 is currently able to walk without assistance and voids into a diaper. Intracranial pial AVF is a rare disease entity that can be treated with surgical disconnection or endovascular embolization. It is important for the appropriate treatment strategy to be selected on the basis of patient-specific and lesion-specific factors in order to achieve good outcomes.

KEY WORDS : Intracranial · Pial · Arteriovenous fistula .

INTRODUCTION

Intracranial pial arteriovenous fistula (AVF) is a rare cerebrovascular malformation. According to a series reported by Halbach⁵⁾, pial AVF accounts for 1.6% of all intracranial vascular malformations. Intracranial pial AVFs have a single or multiple arterial connections to a single venous channel. They differ from brain arteriovenous malformations in that they lack a true nidus. They differ from dural AVFs in that they derive their arterial supply from pial or cortical arteries and are not located within the dura mater⁶⁾.

Because pial AVF has a poor natural history, the clinical suspicion of pial AVF, followed by prompt appropriate treatment is important⁸⁾. Here, we present two cases of pial AVFs treated surgically and endovascularly.

CASE REPORT

Case 1

A 41-year-old woman was brought to our hospital because she had been suffering from general weakness for two

weeks. She had been treated with conservative therapy at an oriental medicine clinic. One hour before admission to our hospital, she was found unconscious on the floor. The initial neurological examination revealed that she was in a comatose state (Glasgow Coma Scale was E1M3V1). Right hemiparesis was noted, and her left pupil was fully dilated without light reflex. There was no history of head trauma, or head injury. Brain computed tomography (CT) revealed the presence of an acute subdural hematoma in the left frontoparietal convexity with significant mass effect. There was no evidence of scalp swelling or skull fracture.

Emergency craniectomy was performed, and a subdural hematoma was removed. A red sylvian vein was observed on the brain surface exposed after the removal of the hematoma. The sylvian vein was larger in diameter than usual, and arterialization was suspected because the mixture of arterial and venous blood in the vessel was grossly visible. When the red sylvian vein was traced as far back as possible, we found that it was connected to the cortical red vein (Fig. 1). However, no other abnormal lesions were found in the exposed brain surface. Angioanatomical evaluation was performed under the suspicion of an arteriovenous shunt. Diagnostic conventional cerebral angiography revealed an arteriovenous fistula supplied by the left posterior inferior temporal artery. The fistula drained into the left sylvian vein (Fig. 2A). A left posterior communicating artery aneurysm was also found.

• Received : May 6, 2008 • Accepted : July 24, 2008

• Address for reprints : Young-Je Son, M.D.

Department of Neurosurgery, Seoul National University Boramae Hospital, 425 Sindaebang 2-dong, Dongjak-gu, Seoul 156-707, Korea

Tel : +82-2-840-2681, Fax : +82-2-831-0714

E-mail : pettprince1024@hanmail.net

Reoperation for treatment of the two lesions was performed. The posterior communicating artery aneurysm was treated by direct surgical clipping, and there was no evidence of a previous aneurysmal rupture. Intraoperative hemorrhage occurred while approaching the fistulous point with the guidance of navigation. The bleeding point was located just distal to the red vein of the fistulous point. The fistula was disconnected with the aid of bipolar cautery, and the color of the sylvian vein quickly changed to blue.

On postoperative cerebral angiography, the posterior communicating artery aneurysm was well obliterated, and

complete disconnection of the fistula was demonstrated (Fig. 2B). The 18-month postoperative cerebral angiography showed no interval change. The patient recovered slowly, and after four months of rehabilitation, she was able to return home with no focal neurological deficit, and she has been capable of doing household chores ever since.

Case 2

A 10-year-old boy with nine-day history of paraparesis and urinary incontinence was transferred for neurosurgical management. He began to experience paresthesia in both legs 10 days ago, and free-voiding followed

one day later, along with weakness in both legs. On neurological examination, he showed bilateral weakness in grade III in knee flexion and extension, grade II in ankle dorsi/plantar flexion, and grade I in big toe dorsi/plantar flexion. Sixty percent sensory decrease on his legs was noted. No saddle anesthesia was present, and motor power in the upper extremities was within normal range. Any previous history of seizure or cardiac disease was denied.



Fig. 1. Intraoperative photographs. A : The left sylvian vein appears thicker than usual (arrows) and is connected to a cortical vein (circle). B : Enlargement of the circle in figure A, showing a connection between the sylvian vein and a cortical vein.

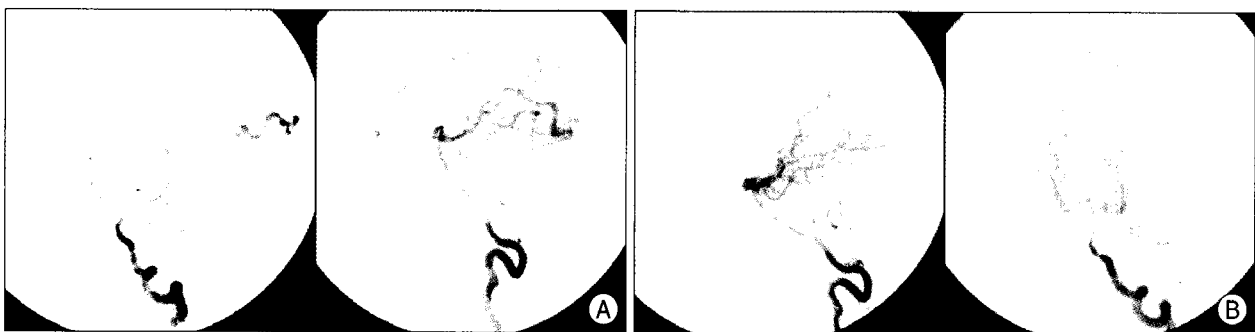


Fig. 2. A : Left vertebral artery angiography, anteroposterior (left) and lateral (right) view from the late arterial phase to the venous phase showing that the arteriovenous fistula is fed by the left inferior temporal artery and drains into the left sylvian vein. B : Postoperative angiography demonstrating the obliteration of the arteriovenous fistula.

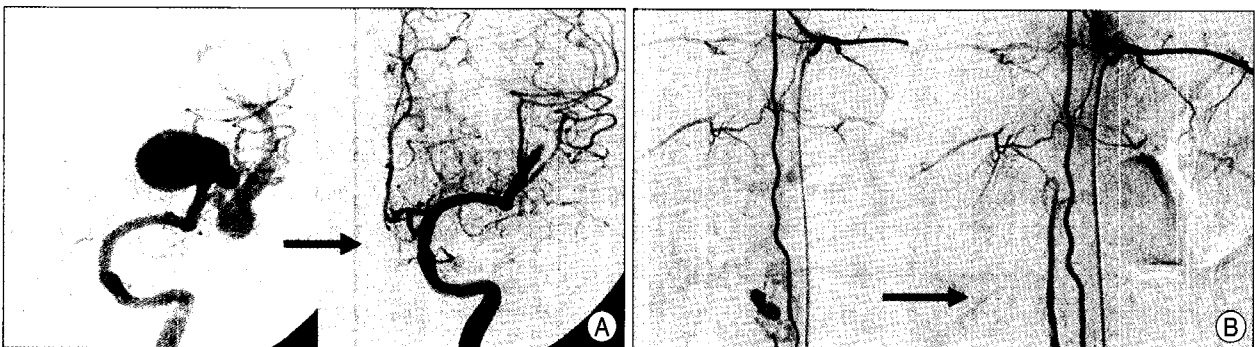


Fig. 3. Pre- and post-embolization images. A : Left : Left internal carotid artery angiogram, anteroposterior view revealing a large AVF supplied from the superior division of the left middle cerebral artery. Right : Post-embolization angiography showing complete obliteration of the arteriovenous fistula. B : Left : Spinal angiography showing left T10 segmental artery feeding an arteriovenous shunt. Right : Post-embolization angiography demonstrating no residual fistula.

On the basis of magnetic resonance imaging (MRI) and angiography of both the brain and spine, he was diagnosed with an intracranial pial AVF (Fig. 3A) involving the superior division of the left middle cerebral artery as well as a spinal perimedullary AVF at the L1 level with feeders from the left T10, right L1, and L2 segmental arteries (Fig. 3B). Endovascular embolization was used to treat both lesions. Postembolization angiography demonstrated complete occlusion of the fistulas.

The patient was discharged with no newly developed deficits, and his symptoms have been slowly improving. On the 18-month follow-up, he was able to walk without assistance, but he was unable to run. He still voids into diapers but occasionally feels the need to void.

DISCUSSION

Intracranial pial AVFs are rare cerebrovascular lesions, with less than 100 reported cases since 1970¹⁴. They have only recently been recognized as a distinct pathological entity from other intracranial cerebrovascular malformations⁷. Intracranial pial AVFs are comprised of a single venous channel in communication with one or more arterial connections from pial or cortical arteries, and they lack a true intervening nidus, unlike cerebral arteriovenous malformations.

Pial AVFs can be congenital or result from a traumatic injury. Little is known about their pathophysiological mechanisms. Abnormal angiogenesis may play a role in the formation of pial AVFs, and it is also possible that an embryological misstep could produce these lesions⁶.

Case 2 had two separate AVFs; one was intracranial and the other was spinal perimedullary. This finding may be another clue to determining the pathogenic mechanism responsible for these lesions. In other words, the coexistence of a pial AVF with another AVF may be an indication of congenital dysplastic elements leading to the formation of such lesions. There have been several case reports of the presence of an aneurysm in the feeder artery in which the pathogenesis could potentially be explained by the high rate of flow in the artery^{2,11}. However, the aneurysm in case 1 was found in a location remote from the feeder.

Pial AVFs can present as hemorrhage, seizure, neurological deficit, cardiac failure in neonates and infants, headache, bruit, symptoms of increased intracranial pressure in infants, giant varices presenting as a palpable mass, skull erosion and macrocephaly^{1-5,12,13}. Congenital pial AVFs usually present during childhood along with syndromes such as Rendu-Osler-Weber disease and Klippel-Trenaunay-Weber syndrome^{5,10}. Case 1 presented with spontaneous

acute SDH, and it was not easy to suspect a vascular malformation in that case. The intraoperative appearance of the arterialized sylvian vein was the only clue. Patient 2's presenting symptoms of lower extremity weakness and voiding difficulty were due to the spinal intramedullary AVF. The intracranial pial AVF was diagnosed incidentally.

Because pial AVFs are very rare, their natural history is not known yet. One study reported that five (63%) of eight patients managed conservatively expired due to acute or subsequent fatal bleeding⁹. On the other hand, the clinical results were good in other studies in which the patients were well treated surgically or endovascularly⁶.

Consensus regarding the best treatment strategy for these lesions had not been established until Hoh et al.⁶ demonstrated that the disconnection of the shunt by surgery or endovascular intervention was sufficient and that resection of the entire vascular malformation was unnecessary. This can be understood by the pathophysiologic mechanism responsible for the abnormality of the AVF, namely, the high-flow nature of the communication between an arterial feeder and a single draining vein without an intervening tangle of vessels. The venous varices commonly associated with AVFs are produced by the high, turbulent flow caused by arteriovenous shunting. Shunt disconnection can be accomplished surgically or endovascularly⁶. The first patient was treated surgically due to the superficial location and difficulty of endovascular access. Endovascular obliteration with glue and coil was performed simultaneously for both the intracranial and spinal AVFs. Certain angiogeometric configurations are unfavorable for endovascular obliteration. These unfavorable factors include multiple arterial connections, drainage from a normal cortical vein into the venous channel of the pial AVF, and embolization that is too proximal, which results in new arterial recruitment and fistula recurrence^{5,6}. However, surgically inaccessible lesions can be treated more safely by endovascular embolization. Until now, the treatment failure rate has been known to be 40% in endovascular embolization and 7.8% in surgical disconnection⁶. However, a more controlled delivery of embolic materials is now feasible with the development of newer endovascular techniques and technologies. Experienced neurosurgical and neuroendovascular teams can carefully determine the safest and most effective treatment modality on the basis of patient-specific and angiospecific factors.

CONCLUSION

Pial AVF is a rare intracranial vascular malformation. Because pial AVFs have a poor natural history and relatively

good clinical outcome with prompt treatment, it is important to establish the clinical diagnosis. Neurosurgeons should be attentive to the fact that the presentation of these lesions is diverse. Once the diagnosis has been made, a neurosurgical and neuroendovascular team should determine the most appropriate treatment modality.

References

1. Aoki N, Sakai T, Oikawa A : Intracranial arteriovenous fistula manifesting as progressive neurological deterioration in an infant : case report. *Neurosurgery* 28 : 619-622; discussion 622-613, 1991
2. Carrillo R, Carreira LM, Prada J, Rosas C, Egas G : Giant aneurysm arising from a single arteriovenous fistula in a child. Case report. *J Neurosurg* 60 : 1085-1088, 1984
3. Drake CG : Cerebral arteriovenous malformations: considerations for and experience with surgical treatment in 166 cases. *Clin Neurosurg* 26 : 145-208, 1979
4. Garcia-Monaco R, De Victor D, Mann C, Hannedouche A, Terbrugge K, Lasjaunias P : Congestive cardiac manifestations from cerebrocranial arteriovenous shunts. Endovascular management in 30 children. *Childs Nerv Syst* 7 : 48-52, 1991
5. Halbach VV, Higashida RT, Hieshima GB, Hardin CW, Dowd CF, Barnwell SL : Transarterial occlusion of solitary intracerebral arteriovenous fistulas. *AJNR Am J Neuroradiol* 10 : 747-752, 1989
6. Hoh BL, Putman CM, Budzik RF, Ogilvy CS : Surgical and endovascular flow disconnection of intracranial pial single-channel arteriovenous fistulae. *Neurosurgery* 49 : 1351-1363; discussion 1363-1354, 2001
7. Lasjaunias P, Manelfe C, Chiu M : Angiographic architecture of intracranial vascular malformations and fistulas--pretherapeutic aspects. *Neurosurg Rev* 9 : 253-263, 1986
8. Morimoto T, Yamada T, Hashimoto H, Tokunaga H, Tsunoda S, Sakaki T : Direct approach to intracranial vertebral arteriovenous fistula. Case report. *Acta Neurochir (Wien)* 137 : 98-101, 1995
9. Nelson K NY, Lasjaunias P, Berenstein A : Endovascular embolization of congenital intracranial pial arteriovenous fistulas. *Neuroimaging Clin N Am* 2 : 309-317, 1992
10. Oyesiku NM, Gahm NH, Goldman RL : Cerebral arteriovenous fistula in the Klippel-Trenaunay-Weber syndrome. *Dev Med Child Neurol* 30 : 245-248, 1988
11. Ratliff J, Voorhies RM : Arteriovenous fistula with associated aneurysms coexisting with dural arteriovenous malformation of the anterior inferior falx. Case report and review of the literature. *J Neurosurg* 91 : 303-307, 1999
12. Talamonti G, Versari PP, D'Aliberti G, Villa F, Fontana RA, Collice M : Complex arteriovenous fistula of the brain in an infant. Case report. *J Neurosurg Sci* 41 : 337-341, 1997
13. Tomlinson FH, Rufenacht DA, Sundt TM Jr, Nichols DA, Fode NC : Arteriovenous fistulas of the brain and the spinal cord. *J Neurosurg* 79 : 16-27, 1993
14. Yamashita K, Ohe N, Yoshimura S, Iwama T : Intracranial pial arteriovenous fistula. *Neurol Med Chir (Tokyo)* 47 : 550-554, 2007