Case Report

Intracerebral pial arteriovenous fistula with large venous varix: A rare case report

Souvagya Panigrahi, Sudhansu S. Mishra, Srikant Das, Deepak K. Parida

Department of Neurosurgery, S.C.B. Medical College and Hospital, Cuttack, Odisha, India
E-mail: *Souvagya Panigrahi - souvagya.ms@gmail.com; Sudhansu S. Mishra - drsmishra@gmail.com; Srikant Das - srikant@gmail.com; Deepak K. Parida - drdparida@gmail.com
*Corresponding author

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Abstract

Background: Intracerebral pial arteriovenous fistulas (AVFs) are rare vascular lesions of the brain. These lesions are composed of one or more direct arterial connection to a single venous channel without true intervening nidus and usually have associated venous varix or giant venous aneurysms. Intracerebral varices are occasionally associated with high-flow AVF, and usually treated by interrupting the feeding arteries leaving the varices intact.

Case Description: We report a rare case of a 24-year-old male with a single-channel pial AVF of the left cerebral hemisphere, which was fed by the left anterior cerebral artery (ACA) and was associated with large venous varix and continuous varicose venous dilatation. This superficially located varix was over 6 cm in diameter posing significant mass effect and had calcified walls. Direct surgical flow disconnection followed by removal of large varix resulted in complete disappearance of pial AVF without complication.

Conclusion: Though endovascular occlusion of feeding arteries offers a simple and safe option, direct surgical removal should be considered in rare cases of intracerebral superficially located large AVF with calcified wall and mass effect.

Key Words: Direct surgical removal, Intracerebral pial arteriovenous fistulas, large venous varix, left anterior cerebral artery

INTRODUCTION

Intracerebral pial arteriovenous fistulas (AVFs) are rare vascular lesions of the brain and account for 1.6% of all brain vascular malformations. They have been recently recognized as distinct vascular anomalies, different from brain arteriovenous malformations (AVMs). These lesions differ from AVMs as they are direct artery to vein connections, have no nidus, and are composed of one or more direct arterial feeders with a single draining vein. Many of these have associated venous varix or giant venous aneurysm. They differ from dural AVFs as they acquire feeders from pial or cortical arteries and are not located within the leaflets of the dura. Usually, only disconnection of the feeding artery is sufficient to decompress the lesion because the varices might shrink and immediately lose the mass effect. However, if the varices have a thick and hard calcified wall, only disconnection of the feeding artery might not reduce the volume.

Here we report a rare case of intracerebral giant venous varix with calcified walls and mass effect, secondary...
to a single-channel pial AVF. Because of the superficial location, mass effect and thick calcified wall, direct surgical removal of the AVF mass was performed and the varix removed without complication.

**CASE REPORT**

A 24-year-old right-handed male, without history of head trauma or alcohol abuse, was admitted to our department with 6-year history of repeated episodes of generalized tonic-clonic seizure. In spite of anticonvulsant therapy, the attack became more frequent. There was no history suggestive of a prior subarachnoid hemorrhage or transient ischemic events. The patient had no mucocutaneous telangiectasia or episodes of recurrent epistaxis. None of his family members including siblings and parents suffered from such type of illness. On examination, he was alert and behaving normally with no focal neurological deficit. There were no signs of meningism. General physical examination showed normal results. Routine laboratory tests were within normal limits and inter-ictal electroencephalogram revealed no abnormality.

On computed tomography (CT) scan, a large well circumscribed round walled strong and uniformly enhancing hyper dense space occupying lesion with peripheral wall calcification was found in left frontal lobe [Figure 1]. The lesion was causing significant mass effect with midline shift. A left frontal well defined flow void of size 60 × 66 mm communicating with the superior sagittal sinus (SSS) through dilated cortical vein with feeder supply from left ACA was observed in the cranial magnetic resonance imaging (MRI) with angiogram [Figure 2]. Possibility of pial AVF with large venous varix was considered. 128 Multi slice cerebral CT angiography (CTA) revealed a large left frontal AVF supplied by the distal left ACA draining through a dilated cortical vein into the SSS [Figure 3]. A large variceal dilatation of the proximal venous end measuring nearly 6.5 cm in diameter with wall calcification was noted in association with the AVF.

The patient underwent a left frontal craniotomy crossing midline. On reflecting the dura to opposite side, a globular soft pinkish pulsatile varix (≈ 6 × 6 cm) was encountered superficially in left frontal lobe [Figure 4a and b] close to the anterior falx with dilated draining vein reaching up to the anterior part of the SSS. A fistulous connection between the mass and left ACA was noted, which confirmed the presence of AVF. As the mass was posing significant mass effect with very hard and thick wall, simple occlusion of feeding artery did not result into disappearance of the AVF. Hence total excision of the AVF mass along with the venous varix was performed [Figure 4c] after

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**Figure 1:** Computed tomography scan of brain showing a left frontal large well circumscribed round smooth walled strong and uniformly enhancing hyper dense space occupying lesion with peripheral wall calcification. The lesion demonstrates no obvious perilesional edema with significant mass effect with midline shift.

**Figure 2:** (a) T2 axial, (b) T2 coronal, (c) T1 sagittal and, (d) MR angiogram sequence of cerebral magnetic resonance imaging reveals a left frontal well defined flow void of size 60 × 66 mm communicating with the superior sagittal sinus through dilated cortical vein with feeder supply from left anterior cerebral artery.

**Figure 3:** 128 Multi slice cerebral computed tomographic angiography sagittal (a) and coronal (b) view demonstrates a large left frontal arteriovenous fistula supplied by the distal left anterior cerebral artery draining through a dilated cortical vein into the superior sagittal sinus. A large variceal dilatation of the proximal venous end measuring nearly 6.5 cm in diameter with wall calcification is shown in association with the arteriovenous fistula.

**Figure 4:** (a) T2 axial, (b) T2 coronal, (c) T1 sagittal and, (d) MR angiogram sequence of cerebral magnetic resonance imaging reveals a left frontal well defined flow void of size 60 × 66 mm communicating with the superior sagittal sinus through dilated cortical vein with feeder supply from left anterior cerebral artery.
ligating the primary feeding artery. The patient had an unremarkable postoperative course with no new focal neurological deficit. A CT scan on second postoperative day demonstrated a large filling defect in the area of the varix excision [Figure 5a and b]. Postoperative CTA done at the time of discharge showed complete obliteration of the AVF and reconstitution of the distal ACA [Figure 5c and d]. He has been followed up for 6 months and has been seizure-free since then on a daily dosage of 300 mg of phenytoin.

**DISCUSSION**

Intracranial pial single-channel AVFs are rare cerebrovascular lesions, accounting for 1.6% of all lesions.\(^1\) They have recently been considered distinct from AVMs.\(^3\) They consist of one or more arterial connection to a single venous channel without true intervening nidus, unlike cerebral AVMs.\(^2\) The abnormality from AVFs arises from its high-flow nature. A fistulous communication between feeding artery and single draining vein can produce venous dilatation, varix and even aneurysms by turbulent high flow.\(^2,4,9\)

Pial AVFs can result from trauma or may be congenital.\(^1,8\) Congenital AVFs are usually diagnosed during infancy or early childhood, although many patients in the literature are young adults, like this case.\(^2\) The pathophysiologic mechanisms giving rise to these lesions are still not clear.\(^1\) Hoh, et al.\(^2\) postulated that a misstep in embryological development of the cerebrovasculature could produce these lesions. Alternatively, abnormal angiogenesis and associated vascular growth factors and cytokine may play a role.

Pial AVFs come to clinical attention with seizures, hemorrhage, headache, focal neurological deficit, symptoms of increased intracranial pressure, and intracranial bruit.\(^2,7\) In neonates and infants, they may present with high output cardiac failure, increased head circumference, and bruit.\(^6\)

Pial AVFs can be diagnosed with cerebral CTA, especially 3D angiograms. Three dimensional angiograms can delineate complex angiography well because of its inherent capability of obtaining reconstruction images at any angles. In this case, the feeder from the left ACA was visualized clearly on 128 multi slice CTA.

Treatment strategies of pial AVFs are different than that of cerebral AVMs. Simple disconnection of arteriovenous shunting is considered enough in most cases, either by microsurgery or endovascular embolization without resection of entire vascular malformation.\(^2\) This strategy is based on the characteristics of pial AVFs, which are high-flow nature of communication between an arterial feeder and a single draining vein without an intervening tangle of vessels. After establishment of arteriovenous connection, associated venous varices can be produced.
by high, turbulent flow caused by arteriovenous shunting. Thus, removal of varix was not necessary unless the malformation accompanied hematoma with mass effect. Obliteration of the fistula by an endovascular route, avoiding the risks associated with craniotomy, should always be considered especially when the lesion is deep seated or the risk of neurological deficit with surgery is high.

In our case, single channel pial AVF fed directly by left ACA with associated large venous varix was demonstrated in CTA. Because of superficial location, significant mass effect and thick calcified wall, direct surgical removal of the AVF mass was performed after disconnection of single feeder from large venous varix, which was the most proximal part of dilated cortical vein [Figure 6].

CONCLUSION

Intracerebral pial AVFs with large venous varix are rare vascular malformations that can be successfully managed surgically with good outcome. Although endovascular occlusion of feeding arteries offers a simple and safe option, direct surgical removal should be considered in rare cases of superficially located intracerebral large pial AVF with calcified wall and mass effect.

REFERENCES

1. Halbach VV, Higashida RT, Hieshima GB, Hardin CW, Dowd CF, Barnwell SL. Transarterial occlusion of solitary intracerebral arteriovenous fistulas. AJNR Am J Neuroradiol 1989;10:747-52.
2. Hoh BL, Putman CM, Budzik RF, Ogilvy CS. Surgical and endovascular flow disconnection of intracranial pial single-channel arteriovenous fistulae. Neurosurgery 2001;49:1351-64.
3. Lasjaunias P, Manelfe C, Chiu M. Angiographic architecture of intracranial vascular malformations and fistulas–pretherapeutic aspects. Neurosurg Rev 1986;9:253-63.
4. Lawton MT, Jacobowitz R, Spezzler RF. Redefined role of angiogenesis in the pathogenesis of dural arteriovenous malformations. J Neurosurg 1999;87:267-74.
5. Lee JY, Son YJ, Kim JE. Intracranial pial arteriovenous fistulas. J Korean Neurosurg Soc 2008;44:101-4.
6. Limaye US, Siddhartha W, Shrivastav M, Anand S, Ghatge S. Endovascular management of intracranial pial arterio-venous fistulas. Neurol India 2004;52:87-90.
7. Lv X, Li Y, Jiang C, Wu Z. Endovascular treatment of brain arteriovenous fistulas. AJNR Am J Neuroradiol 2009;30:851-6.
8. Santosh C, Teasdale E, Molyneux A. Spontaneous closure of an intracranial middle cerebral arteriovenous fistula. Neuroradiology 1991;33:65-6.
9. Vives-Bauza C, Drake CG, Fox AJ, Pelz DM. Giant intracranial varices secondary to high-flow arteriovenous fistulae. J Neurosurg 1987;66:198-203.