




# Endovascular Management of Intracranial Pial Arteriovenous Fistula with Giant Venous Varix: Case Report and Review of Literature.

Nishtha Yadav<sup>1</sup> Ambuj Kumar<sup>2</sup> Ketan Hedao<sup>2</sup> Sonjjay Pande<sup>1</sup>

<sup>1</sup> Department of Radiology, Super Speciality Hospital, Netaji Subhash Chandra Bose Medical College, Jabalpur, Madhya Pradesh, India

<sup>2</sup> Department of Neurosurgery, Super Speciality Hospital, Netaji Subhash Chandra Bose Medical College, Jabalpur, Madhya Pradesh, India

**Address for correspondence** Nishtha Yadav, MBBS, MD, DM, Department of Radiology, Super Speciality Hospital, Netaji Subhash Chandra Bose Medical College Jabalpur, Madhya Pradesh, 482003, India (e-mail: nishthayadav@yahoo.com).

J Clin Interv Radiol ISVIR

## Abstract:

**Background:** Intracranial pial arteriovenous fistulas (AVFs) are rare intracranial vascular malformation and their treatment is challenging because of high-flow pial supply of the lesion. We report a case of intracranial pial AVF with associated giant venous varices managed successfully by embolization.

**Case Description:** A 25-year-old male presented with sudden onset headache and altered sensorium. Computed tomographic scan was suggestive of frontal lobar hematoma that had dissected into the third ventricle with associated intraventricular hemorrhage. Magnetic resonance imaging revealed presence of large vascular channels with the presence of multiple flow voids in bilateral frontal location. Digital subtraction angiography revealed pial fistula with arterial supply from hypertrophied left anterior cerebral artery. Treatment was done using combined coil and onyx embolization after careful analysis of angioarchitecture of the lesion using three-dimensional rotational angiogram cross-sectional reconstructions. Postprocedure angiogram revealed closure of fistula and giant varices.

**Conclusion:** Pial AVFs with giant venous varix in adults are rare vascular malformation and understanding the angioarchitecture with exact identification of fistulous point is important to plan for surgical/endovascular management strategies. Combined strategy of embolization using coils and onyx is a safe option in such cases with giant varices.

## Keywords

- ▶ pial arteriovenous fistula
- ▶ giant venous varices
- ▶ embolization
- ▶ coils
- ▶ onyx
- ▶ vascular malformation

## Introduction

Intracranial pial arteriovenous fistulas (AVFs) are rare intracranial vascular malformation and account for 1.6% of all brain vascular malformations.<sup>1</sup> They are supplied by intra-

cranial pial vessels and do not have intervening nidus, with direct fistulous connection between the supplying pial artery and draining vein. The treatment of these lesions may be challenging because of high-flow pial supply of the lesion. Treatment options include microsurgical or endovascular

received

August 2, 2020

accepted after revision

June 9, 2021

DOI <https://doi.org/>

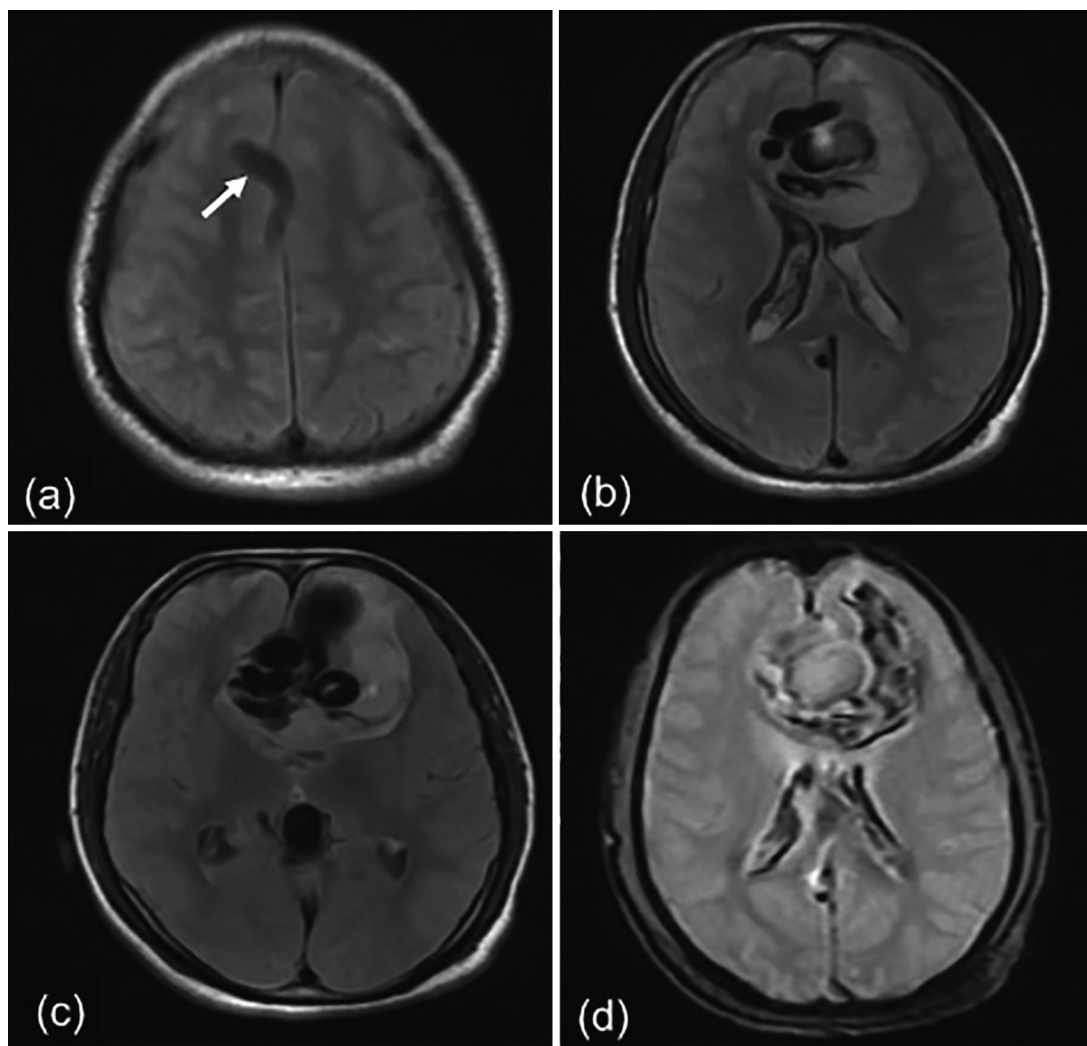
10.1055/s-0042-1750434.

ISSN 2457-0214.

© 2022. Indian Society of Vascular and Interventional Radiology. All rights reserved.

This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India



**Fig. 1** (A) Fluid-attenuated inversion recovery (FLAIR) axial image shows sulcal FLAIR hyperintensities (due to subarachnoid hemorrhage) with dilated vein. FLAIR axial images at level of lateral ventricles (B) and basal ganglia (C) show hypointense lesion in frontal interhemispheric region with flow voids with hyperintensity in bilateral frontal lobes; sulcal FLAIR hyperintensities are also noted. (D) Gradient recalled echo image shows blooming in frontal lobe (due to hematoma) and bilateral lateral ventricles (secondary to intraventricular extension of hemorrhage).

methods; however, in some cases, combination of both techniques is required. We report a case of intracranial pial AVF with associated giant venous varices managed successfully by embolization.

### Case Description

A 25-year-old male presented with sudden onset headache and altered sensorium. Computed tomographic (CT) scan was done that revealed subarachnoid hemorrhage in suprasellar cistern with lobar hematoma in bilateral frontal lobes. The lobar hematoma had dissected into the third ventricle with associated intraventricular hemorrhage. Hyperdense serpiginous lesions were noted surrounding the lesion. Magnetic resonance imaging (MRI) revealed presence of large vascular channels with presence of multiple flow voids in bilateral frontal location (►Fig. 1). Digital subtraction angiography (DSA) revealed a pial fistula with arterial supply from hypertrophied left anterior cerebral artery (ACA). Three-dimensional (3D) rotational angiogram cross-sectional images were

reviewed that confirmed presence of single hole fistula. The fistulous sac was irregular and measured approximately 12 × 11 mm with the presence of narrowing distally. Multiple draining venous channels were noted from the fistulous sac with presence of dilated venous varices. Drainage was noted into the deep venous system and further into straight sinus (►Fig. 2). It was decided to embolize the fistula.

Procedure was done under general anesthesia. A 7F long sheath was positioned in left common carotid artery and a 6F guide catheter was taken and positioned in left internal carotid artery petrous segment. Microcatheter was taken over 0.014 microwire and the microcatheter was positioned in the fistula sac. Multiple coils (EV3, Irvine, California, United States) were deployed in the aneurysm sac and in the feeding left ACA vessel. Total eight coils were deployed and check injection showed marked slowing of flow in fistula. It was decided to further proceed with liquid embolic embolization. About 0.3 mL onyx (EV3) was injected under blank roadmap in an attempt to close fistula. Final check angiogram revealed complete closure of fistula (►Fig. 2(E-I)).

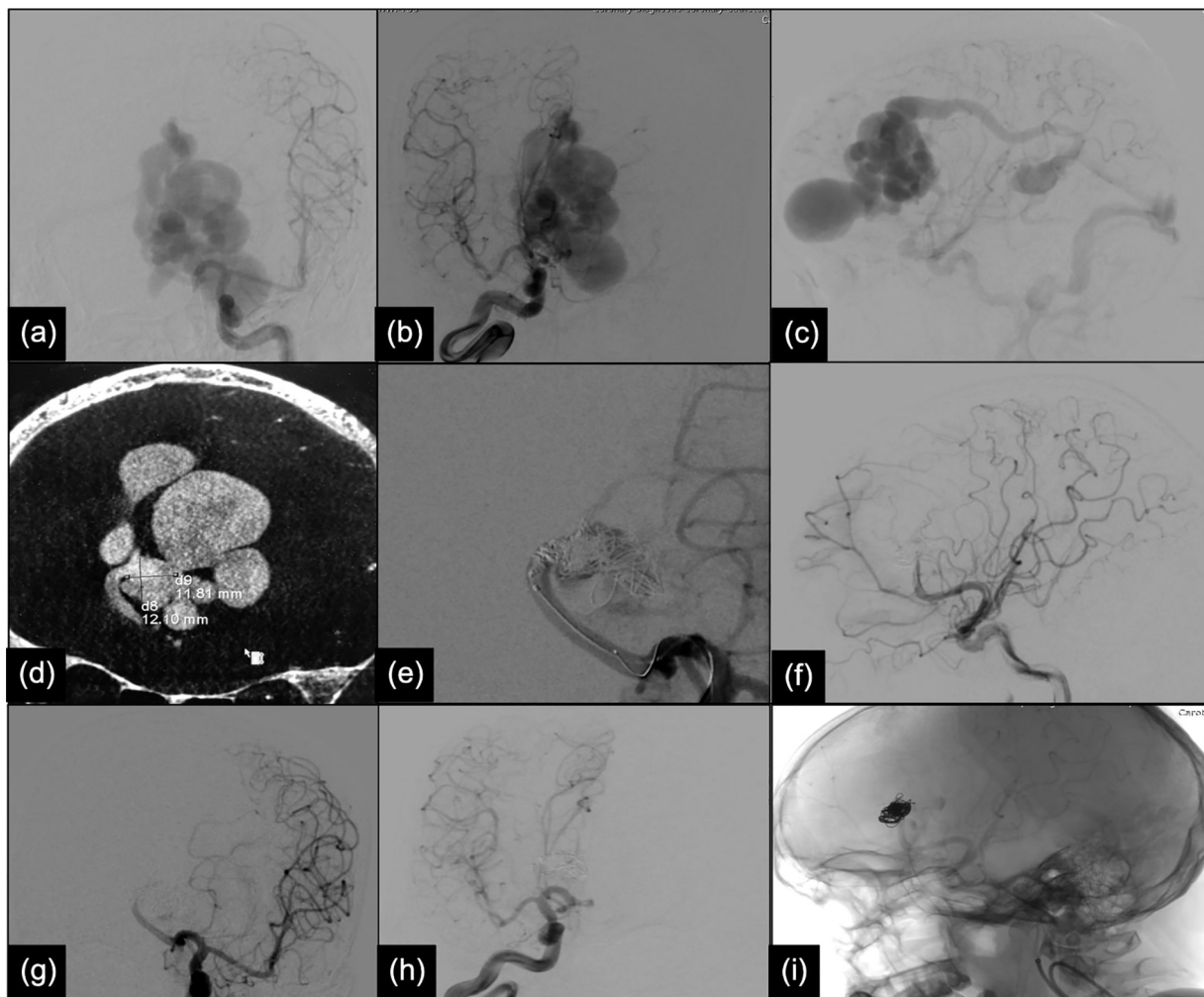
Postprocedure patient was extubated on table and his neurological status was same as preprocedure. Mannitol and steroids were given to prevent excessive edema from thrombosis of venous channels. Postprocedure MRI revealed thrombosed sac in bilateral frontal location with mild vasogenic edema in left frontal lobe. Patient was discharged with modified Rankin Scale (mRS) score 0 on seventh postoperative day. On 1-year follow-up, the patient was asymptomatic (with mRS score 0). Pre-embolization, Post-embolization and 1 Year Follow up MRI images are given in **Fig. 3**.

## Discussion

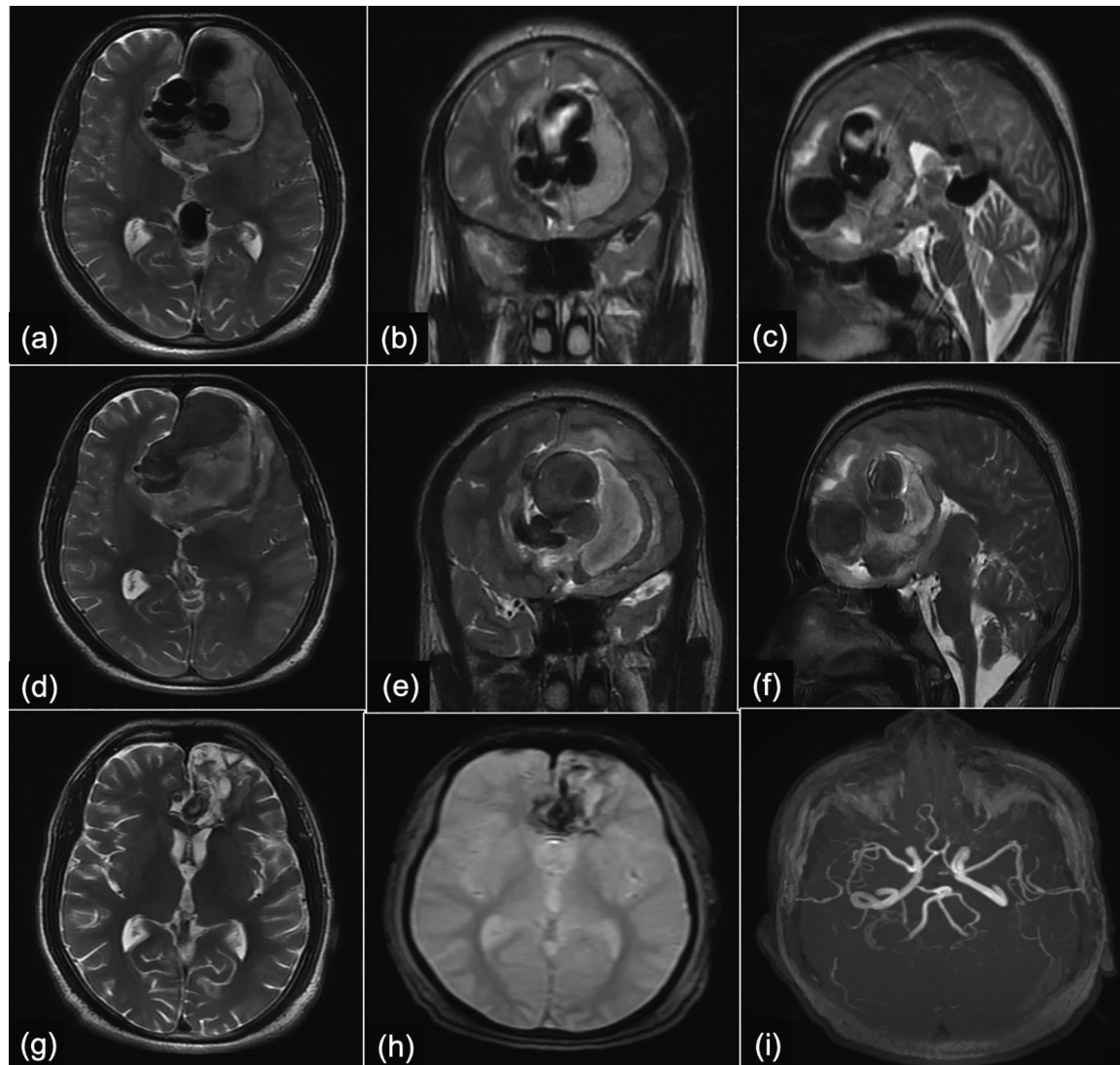
Pial AVFs may be congenital or posttraumatic in nature. They may present in childhood; however, many cases are reported in young adult as in our case.<sup>2,3</sup> They may present with headache, mass effect, seizures, or focal neurological deficit.

Ruptured pial fistula leading to intraparenchymal hemorrhage with intraventricular hemorrhage and subarachnoid hemorrhage is rare.<sup>4-6</sup>

As per literature review, pial fistulas in pediatric patients (< 15 years old) are likely to have seizure as an initial presentation, while adult patients (>15 years old) are more likely to have hemorrhage.<sup>7</sup> Presence of varix was noted in 77.1% of patients and was more commonly seen in pediatric patients.<sup>7</sup> Diagnosis is usually made on DSA, which helps in assessing the angioarchitecture of the lesion, in terms of single hole versus multiple holes, feeding vessels, draining veins, presence of venous varix, and stenosis of draining vein. 3D rotational angiography is very helpful not only in assessing these characteristics but also in planning for endovascular management. Identification and measurement of fistulous sac are essential to plan the endovascular therapy while considering coil embolization. **Fig. 1-3**



**Fig. 2** (A) Left internal carotid artery (ICA) injection anteroposterior (AP) view shows early draining dilated engorged veins in midline with arterial connection to left anterior cerebral artery (ACA) that is dilated. No intervening nidus is noted. (B) Right ICA injection AP view shows opacification of pial arteriovenous fistula filling through left ACA via ACom. Right ACA is displaced toward right of midline due to mass effect. (C) Left ICA injection lateral view delayed phase shows dilated veins with venous varices. (D) Dyna computed tomography coronal reconstruction shows presence of fistulous site with venous sac (12 × 11 mm) as measured; this venous sac was the target of loose coil packing for embolization. (E) Embolization image showing coils in the fistula sac and in the feeding artery, 0.3 mL onyx was injected after placement of coils at the site marked with arrow (F, G). Left ICA angiogram after coil + onyx embolization showing occlusion of fistula with normal filling of vessels. (H) Right ICA injection after embolization showing occlusion of fistula with normal filling of vessels. (I) Native image showing coil and onyx cast in situ.



**Fig. 3** Preembolization T2-weighted axial, coronal, and sagittal images (A–C) compared with corresponding T2-weighted images post-embolization (D–F), showing thrombosis of venous sac and varices after embolization. One-year follow-up images (G–I) are shown. T2-weighted axial image (G) shows thrombosed fistula with collapsed veins and gliosis in left frontal lobe. Gradient recalled echo axial image (H) shows blooming in thrombosed fistula. Maximum intensity projection time of flight magnetic resonance angiography image (I) shows occlusion of fistula in follow-up imaging.

Treatment strategies include surgical dissociation of lesion or endovascular embolization.<sup>6</sup> Microsurgical approaches are particularly useful for pial fistula cases in which embolization is deemed dangerous in cases where the arterial feeder is additionally supplying brain parenchyma. Other advantage of surgical method is that it simultaneously deals with the mass effect caused by venous varices. Jouibari et al reported two cases of pial AVF with giant varices managed by surgical disconnection of fistula as well as removal of variceal sac.<sup>8</sup>

However, due to medial frontal location of fistula in our case with deep seated sac, surgical dissection may lead to additional neurological morbidity.

Endovascular methods include n-butyl cyanoacrylate embolization, coiling, onyx embolization, or a combination of these.<sup>6</sup> Glue embolization is effective and permanent but requires expertise especially in such high-flow fistulas.

Intraprocedural hypotension should be given while injecting glue in such cases. Embolization using onyx can be more controlled; however, it usually requires placement of a proximal balloon to enable settling of onyx at fistula site. While using these liquid embolic agents, care must be taken to prevent liquid embolic migration into venous pouches or distally in pulmonary circulation. Using liquid embolic agents as the sole method of embolization is therefore risky and requires high level of expertise. Venous occlusion without occlusion of fistulous site can be disastrous and can lead to massive bleed. Coiling is helpful in cases where a fistulous sac can be identified that will enable placement of coils in the sac. Loose packing of the sac can be done with coils followed by tight packing of fistula site and feeding artery. Embolization can be completed with coiling alone; however, it may cost a lot due to large number of coils required for tight packing of fistula.<sup>9</sup> To reduce the cost of large number of

coils, combined embolization using coils and onyx or glue may be considered. The mesh of coils causes reduction in flow and acts as a template for settlement of liquid embolic agent and prevents their distal migration due to high flow.<sup>10</sup> This combined strategy of using coils and onyx was used successfully in our case to achieve complete embolization. Disadvantages of coil embolization include increased cost and metallic artifact on CT that can obscure small amount of early bleeding. In expert hands transarterial glue embolization for high-flow pial fistulas remains the gold standard.

## Conclusion

Pial AVFs with giant venous varix in adults are rare vascular malformation and understanding the angio-architecture with exact identification of fistulous point is important to plan for surgical/endovascular management strategies. Combined strategy of embolization using coils and onyx is a safe option in such cases with giant varices.

### Conflict of Interest

None declared.

## 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(04):747-752
- 2 Hoh BL, Putman CM, Budzik RF, Ogilvy CS. Surgical and endovascular flow disconnection of intracranial pial single-channel arteriovenous fistulae. *Neurosurgery* 2001;49(06):1351-1363, discussion 1363-1364
- 3 Oh H-J, Yoon S-M, Kim S-H, Shim J-J. A case of pial arteriovenous fistula with giant venous aneurysm and multiple varices treated with coil embolization. *J Korean Neurosurg Soc* 2011;50(03):248-251
- 4 Coubes P, Humbertclaude V, Rodesch G, Lasjaunias P, Echenne B, Frerebeau P. Total endovascular occlusion of a giant direct arteriovenous fistula in the posterior fossa in a case of Rendu-Osler-Weber disease. *Childs Nerv Syst* 1996;12(12):785-788
- 5 Lin N, Brouillard AM, Snyder KV, Levy EI, Siddiqui AH. Non-galenic arteriovenous fistulas in adults: transarterial embolization and literature review. *J Neurointerv Surg* 2015;7(11):835-840
- 6 Yu J, Shi L, Lv X, Wu Z, Yang H. Intracranial non-galenic pial arteriovenous fistula: a review of the literature. *Interv Neuroradiol* 2016;22(05):557-568
- 7 Yang W-H, Lu M-S, Cheng Y-K, Wang T-C. Pial arteriovenous fistula: a review of literature. *Br J Neurosurg* 2011;25(05):580-585
- 8 Jouibari MF, Zadeh MZ, Khadivi M, Khoshnevisan A, Moazzeni K, Abdollahzade S. Pial arteriovenous fistula with giant varices: report of two cases with good surgical outcome. *J Cerebrovasc Endovasc Neurosurg* 2014;16(02):98-103
- 9 Bongetta D, Lefe E, Pugliese R, Cattalani A, Gaetani P, Thyron FZ. Endovascular embolization of pial arteriovenous fistula fed from P1 segment of posterior cerebral artery in 12 years old girl: case report and review of literature. *Neuroradiol J* 2015;28(03):268-273
- 10 Feng L, Liu Y, Liu J, Lv C, Su C. Pial arteriovenous fistulas: two pediatric cases and a literature review. *Int J Clin Exp Med* 2016;9(05):7855-7862