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Case Report

Pial arteriovenous fistula associated with vein of Galen dilatation in adult: A case report and MRI findings ☆,☆☆

Duc Tan Vo, MD, PhD[a,b], *Tram Bich Thi Ha, MD*[a,b,*], *Tu Ngoc Ho, MD*[a], *Linh Hong Thi Nguyen, MD*[a], *Hoa Viet Nguyen, MD*[c]

[a] *Department of Diagnostic Imaging, University Medical Center, Ho Chi Minh City, Vietnam*
[b] *Department of Radiology, University of Medicine and Pharmacy, Ho Chi Minh City, Vietnam*
[c] *Department of Neurosurgery, University Medical Center, Ho Chi Minh City, UMC, Vietnam*

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ABSTRACT

Pial arteriovenous fistula (PAVF) is a rare intracranial vascular lesion where direct communication exists between one or more pial arteries and a cerebral vein, without an intervening nidus and located in the subpial meningeal space. When the drainage of PAVF involves a dilated, but already formed vein of Galen (VOG), it should be distinguished from other vascular lesions located in this area, because their angio-architecture, natural history and treatment options are different. A 33-year-old female was admitted to our hospital with a history of new-onset generalized tonic-clonic seizures. Clinical examination showed no neurological deficit. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) depicted an arteriovenous fistula that was fed by the pial branches from left posterior cerebral artery and drained into the medial atrial vein before joining the VOG confluence and causing VOG dilatation. No nidus between the feeding arteries and draining vein, dural feeding arteries, or anatomical variations commonly seen with true vein of Galen aneurysmal malformations (VOGM) were found. These finding suggested a diagnosis of a PAVF associated with vein of Galen dilatation, which was confirmed by digital subtraction angiography. The patient was treated with transarterial glue embolization in 1 section, resulting in nearly complete occlusion of the fistula. Conventional MRI and MRA are noninvasive modalities that can provide valuable information regarding the anatomic localization of the fistula point, the feeding arteries, the venous sac, and their relationship with surrounding structures. These techniques are helpful for accurate diagnosis and treatment planning.

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* Corresponding author.
E-mail address: tram.htb1@umc.edu.vn (T.B.T. Ha).
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Introduction

Pial arteriovenous fistula (PAVF) is a rare intracranial vascular lesion where direct communication exists between one or more pial arteries and a cerebral vein, without an intervening nidus and located in the subpial meningeal space. It accounts for about 1.6% of all intracranial vascular malformations and is predominantly seen in children. The exact pathophysiology and the natural history of PAVF remain unclear due to its rarity [1,2]. When the drainage of PAVF involves a dilated, but already formed vein of Galen (VOG), it should be distinguished from other vascular lesions located in this area, including true vein of Galen aneurysmal malformations (VOGM), dural arteriovenous fistulas (DAVF) in Galen region, and arteriovenous malformations (AVM) with VOG drainage and dilatation. Since the angio-architecture, natural history and treatment options for PAVF are different, careful imaging analysis is required to obtain an accurate diagnosis and better treatment outcome.

In recent years, conventional magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) are increasingly being used for assessing intracranial vascular lesions because of their non-invasive nature and ability to delineate soft tissues effectively. Moreover, cross-sectional imaging can allow precise anatomic localization of the fistula point, feeding arteries, venous sac, and relationship with surrounding structures. This information is valuable for both diagnosis and treatment planning [3,4]. Here we report a case of a PAVF associated with intraventricular giant venous varices and vein of Galen dilatation in a 33-year-old female presenting with new-onset seizures. The patient was treated with transarterial glue embolization in 1 section, resulting in nearly complete occlusion of the fistula. The imaging characteristics of PAVF were demonstrated and discussed, focusing on the role of MRI and MRA techniques.

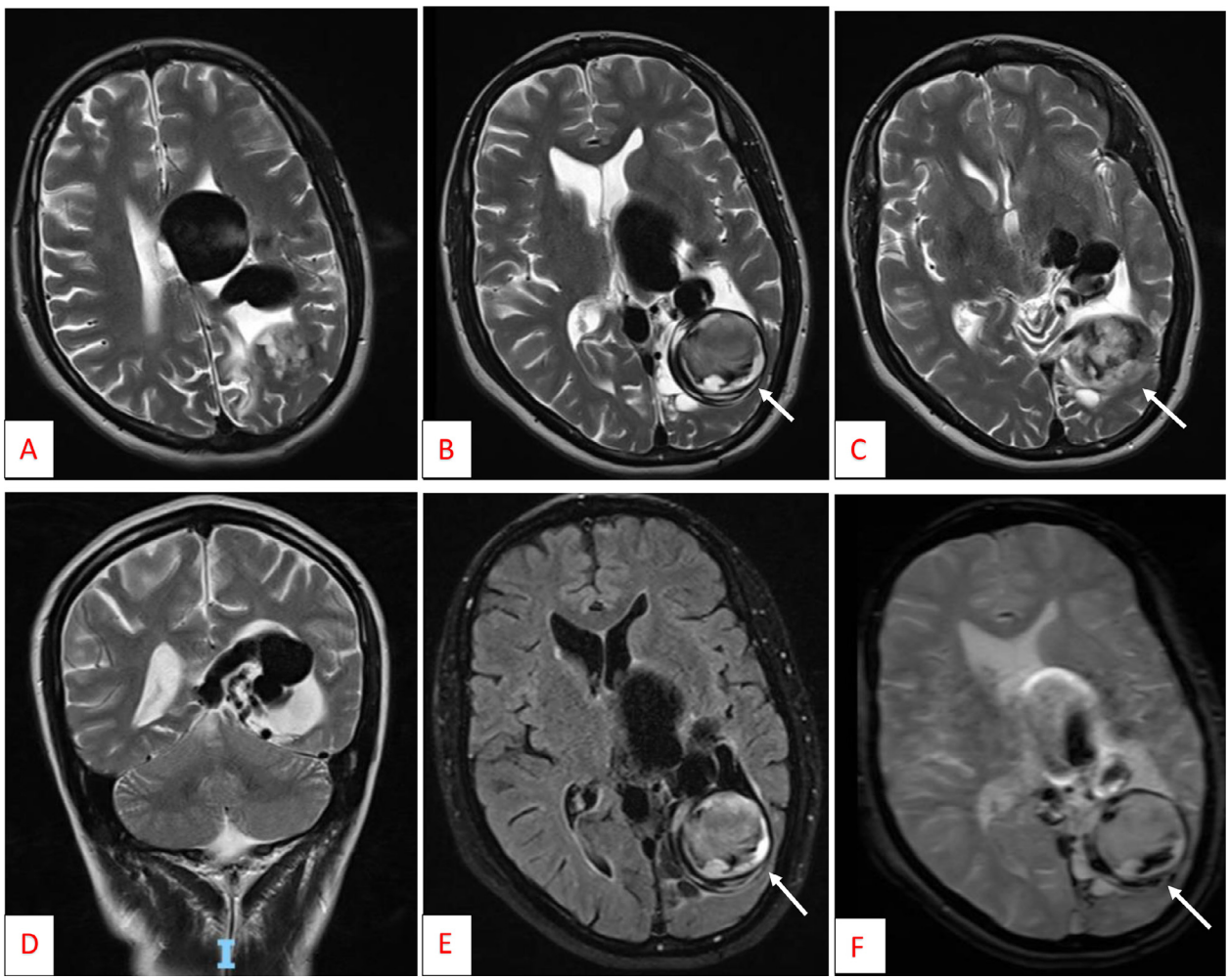

Fig. 1 – Magnetic resonance imaging findings. Axial T2 weighted (A–C), coronal T2 weighted (D) and axial FLAIR images (E) illustrate several large flow voids in the body and atrium of the left lateral ventricle as well as in the ambient cistern and quadrigeminal cisterns, corresponding to multiple venous varices. T2*-weighted MRI image (F) shows partial thrombosis in one of the giant venous varices in the left lateral ventricle (white arrow).

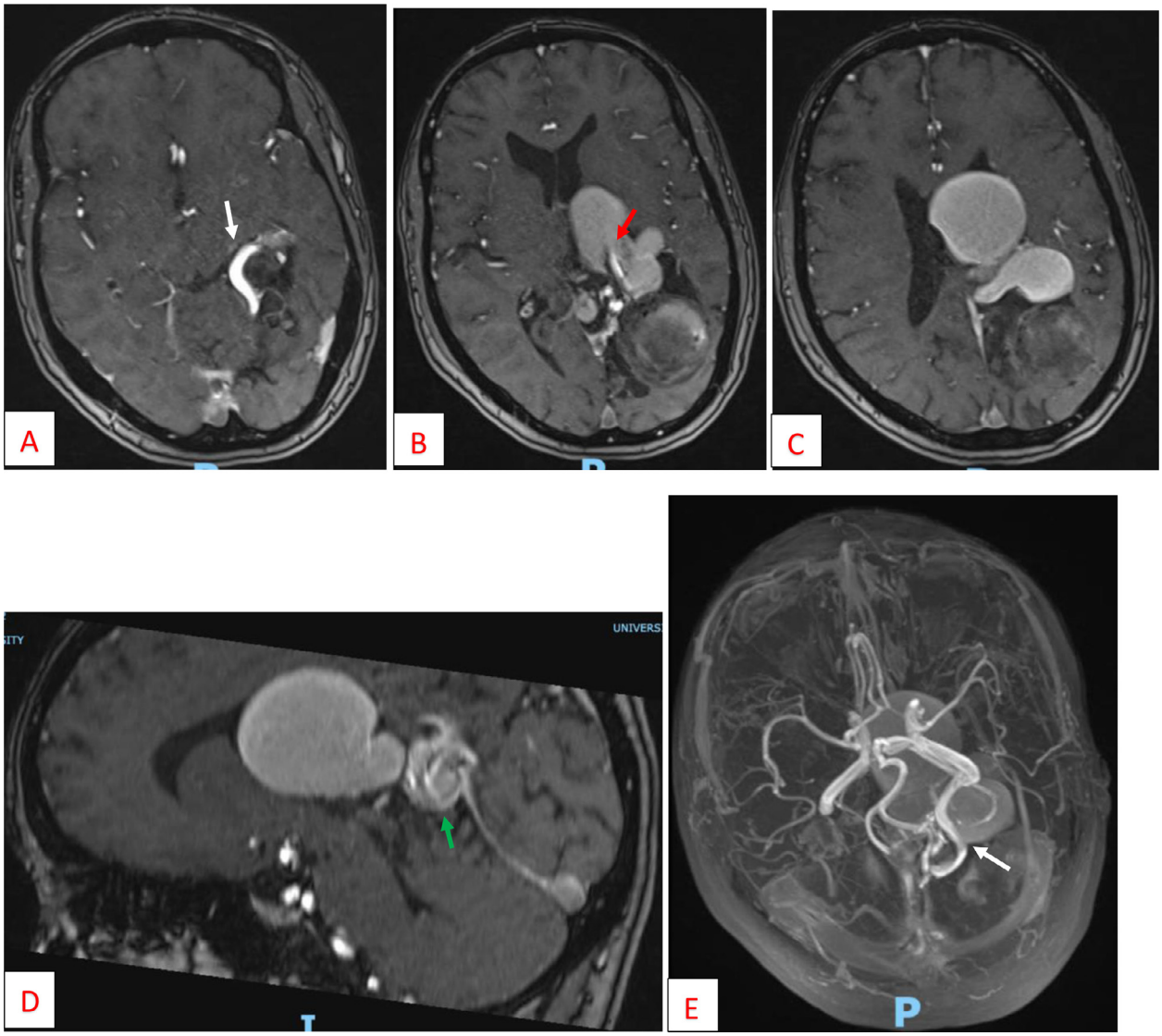

Fig. 2 – Time-of-flight (TOF) magnetic resonance angiography (MRA) findings. TOF images in axial plane (A–C) and sagittal plane (D) demonstrate an arteriovenous shunt with venous flow-related enhancement. The fistula is fed by the pial branches from left posterior cerebral artery (white arrow) and drains into the medial atrial vein before joining the vein of Galen (VOG) confluence and causing VOG dilatation (green arrow). Several giant intraventricular venous varices are noted. The shunt point (red arrow) is identified by the abrupt change in caliber and signal intensity between the feeding arteries and the draining vein. Reconstructed 3D TOF MRA image (E) shows the angio-architecture of the pial arteriovenous fistula.

Case report

A 34-year-old female was admitted to University Medical Center at Ho Chi Minh city because of a 3-month history of repeated episodes of generalized tonic-clonic seizures. There was no history of head trauma or transient ischemic events. On admission, she was alert and oriented with no focal neurological deficit. General physical examination revealed no abnormality.

On T2-weighted images, several large flow voids were found in the body and atrium of the left lateral ventricle as well as in the ambient cistern and quadrigeminal cisterns (Fig. 1). Time-of-flight (TOF) (Fig. 2) and contrast-enhanced (CE) MRA (Fig. 3) demonstrated an arteriovenous shunt with venous flow-related enhancement. The fistula was fed by the pial branches from left posterior cerebral artery (PCA) and drained into the medial atrial vein before joining the VOG confluence and causing VOG dilatation. Several giant intraventricular venous varices were noted. Asymmetric dilatation of the left PCA that fed the fistula was seen at the level of the circle of Willis. The shunt point was identified by the abrupt change in caliber and signal intensity between the feeding arteries and the draining vein. T2*-weighted images showed partial thrombosis in one of the giant venous varices in the left lateral ventricle. The drainage of the deep venous system near the lesion was assessed by 2D-TOF and CE magnetic resonance venography (MRV), which showed the connection of right superior

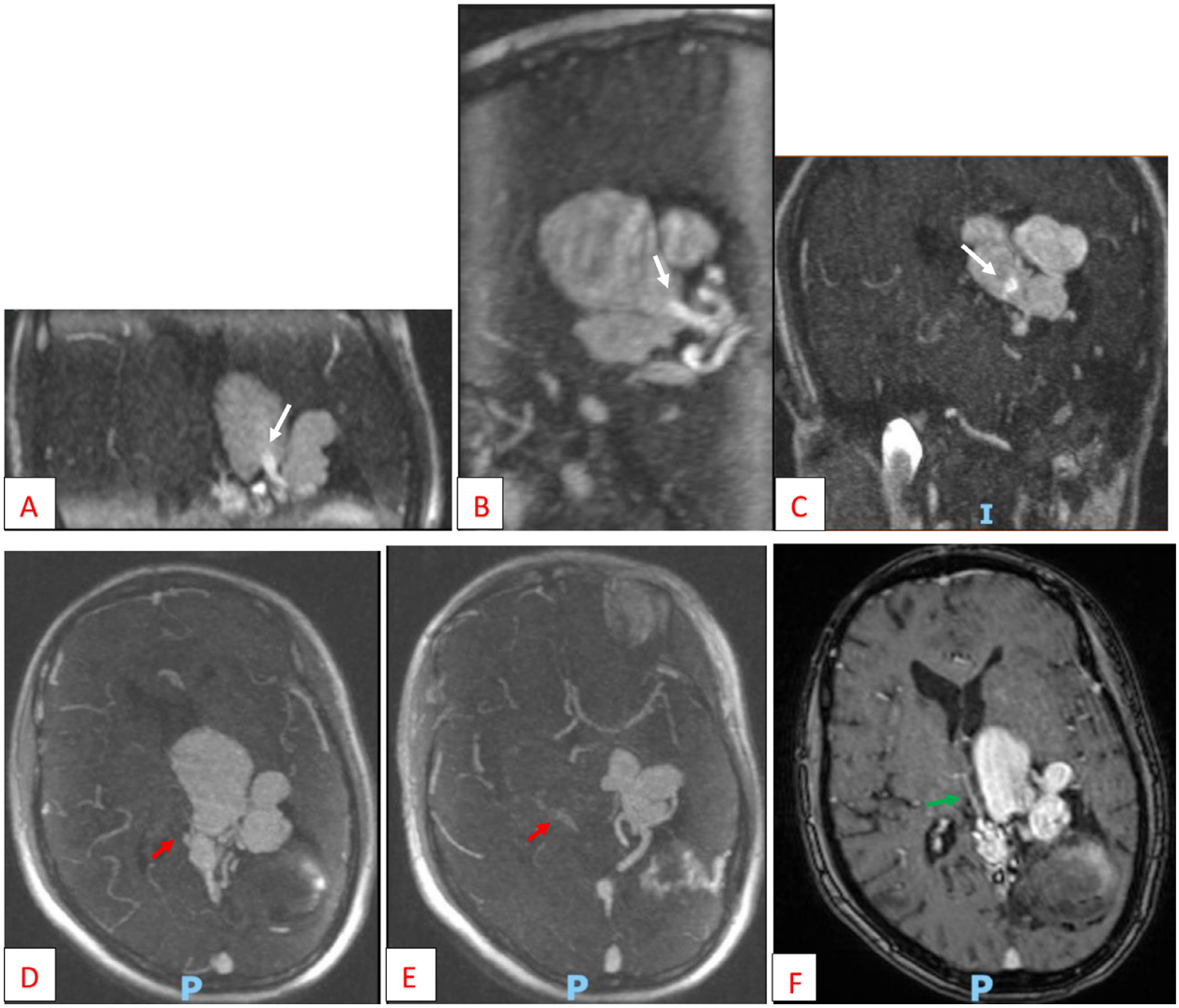

Fig. 3 – Contrast-enhanced magnetic resonance angiography (CE-MRA) findings. CE-MRA images in axial (A), sagittal (B) and coronal planes (C) demonstrate the shunt point (white arrow) by the abrupt change in caliber and signal intensity between the feeding arteries and the draining vein. Contrast-enhanced magnetic resonance venography (CE-MRV) images (D and E) and postcontrast T1-weighted axial image (F) show the connection of right superior thalamic vein (green arrow) and right basal vein of Rosenthal (red arrow) with the dilated vein of Galen.

thalamic vein and right basal vein of Rosenthal with the dilated VOG (Fig. 3). No nidus between the feeding arteries and draining vein, dural feeding arteries, or anatomical variations including presence of occipital and marginal sinuses, absence of straight sinus with persistent falcine sinus, and persistent limbic arterial arch were found. These MRI and MRA findings suggested a diagnosis of a PAVF associated with vein of Galen dilatation, which was confirmed by digital subtraction angiography (DSA) (Fig. 4).

The patient underwent transarterial embolization under general anesthesia. The microcatheter tip was placed just proximal to the fistulous site through a feeding branch of PCA and NBCA mixed with Lipiodol was used. Control angiogram showed near-complete occlusion of the PAVF (Fig. 4). No procedural or postprocedural complications were found. The patient had an unremarkable postoperative course and was discharged in stable condition 3 days after the intervention.

Discussion

In this case report, we described a PAVF in a 33-year-old patient with intraventricular giant venous varices and VOG dilatation. In clinical practice, several differential diagnoses should be considered in case of an arteriovenous shunt involving the VOG region. PAVF associated with VOG dilatation consists of abnormal direct connections between pial arteries that would normally supply the brain and a venous channel, without any intervening network. The fistula is located in the subpial meningeal space, drains into a normally developed VOG with dilation due to venous overload, and usually does not communicate directly with the VOG [1,2]. Brain AVMs can drain into the VOG system and lead to VOG dilatation. However, their feeding arteries and draining veins are connected via a cluster of abnormal networks of vessels

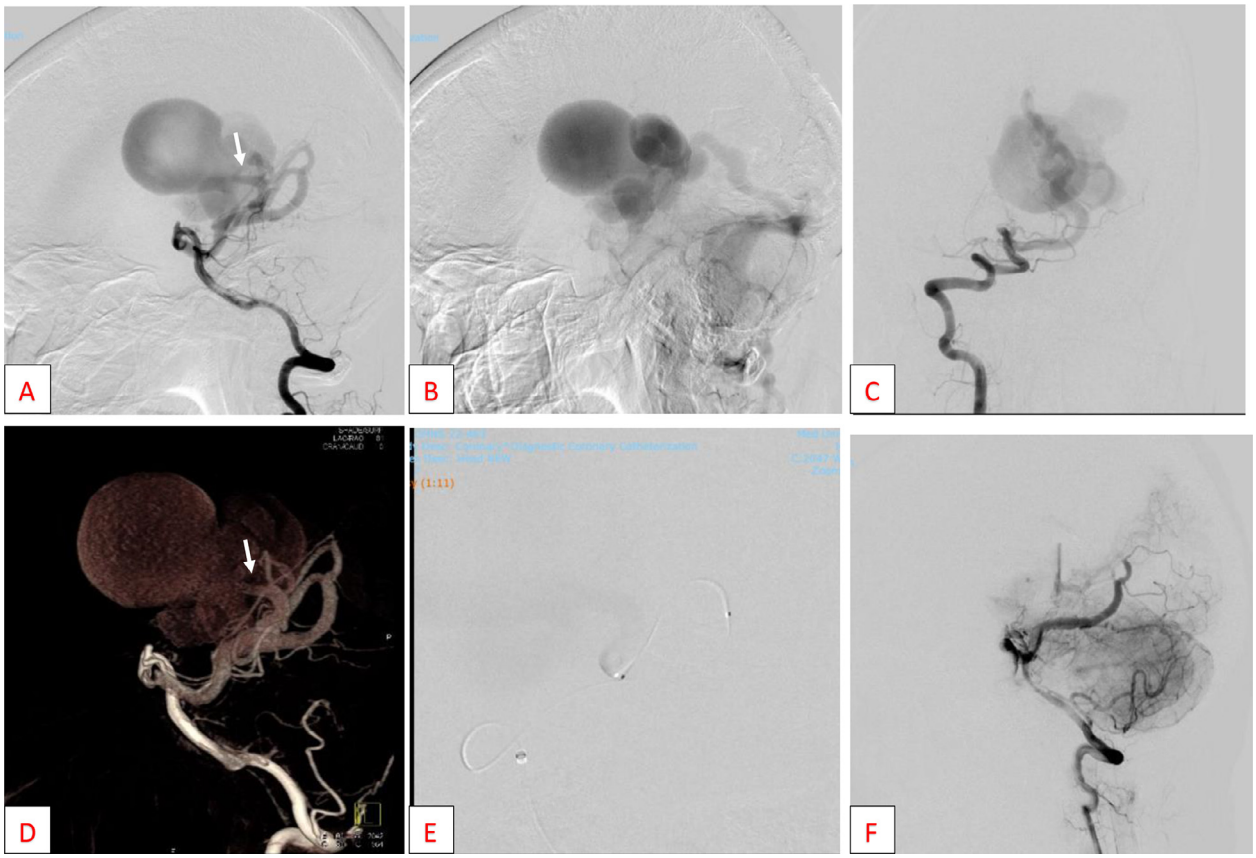

Fig. 4 – Digital subtraction angiography findings. Vertebral angiograms in lateral (A and B) and anterior-posterior (C) views confirm the diagnosis of a pial arteriovenous fistula (PAVF) associated with intraventricular giant venous varices and vein of Galen dilatation. The shunt point is demonstrated by the white arrow (A and D). During the procedure, a microcatheter was placed just proximal to the fistulous site through a feeding branch of PCA and superselective angiography was performed (E). Control angiogram after transarterial glue embolization shows near-complete occlusion of the PAVF (F).

called nidus, which is distinct from PAVF. DAVF in the Galen region is distinguished from PAVF by the location of the fistula within the dura mater and the feeding arteries from the dural rather than pial branches. Finally, true VOGM refers to an arteriovenous fistula between the embryonic choroidal arteries and the dilated embryonic precursor of the vein of Galen, the median prosencephalic vein of Markowski. This lesion develops between the 6th and 11th weeks of gestation and produces a high blood flow in the embryonic venous system, preventing the development of VOG. The dilated venous pouch of the VOGM is located midline in the Galen region, receives a bilateral and usually symmetrical arterial supply from the choroidal arteries and subependymal network originating from the posterior circle of Willis. Some anatomical variations commonly associated with VOGM include presence of occipital and marginal sinuses, absence of straight sinus with persistent falcine sinus, and persistent limbic arterial arch [5–9]. This patient in our study had an arteriovenous fistula that was fed by the pial branches from left PCA and drained into the left medial atrial vein before joining the VOG confluence and causing VOG dilatation. No nidus between the feeding arteries and draining vein, dural feeding arteries, or anatomical variations commonly seen with VOGM were found. MRI and MRA findings suggested a diagnosis of PAVF associated with intraventricular giant venous varices and VOG dilatation.

The role of MRI and MRA in diagnosis and management of intracranial high flow shunts has been shown in some reports [4,9,10]. TOF is a commonly used MRA technique which can produce high-spatial-resolution images without administration of gadolinium-based contrast agents. Owing to their high flow rates, the arteriovenous shunt as well as the feeding arteries and draining veins can be depicted as hyperintense vascular channels. CE-MRA can be performed using single phase or multiple phases techniques. Time-resolved MRA sequences have been applied to address the limitations in temporal resolution of TOF-MRA and single-phase CE-MRA techniques while maintaining spatial resolution. With this technique, imaging can be obtained at rapid time intervals, allowing distinction between the arterial and venous phases. Imaging techniques for assessing the drainage of the arteriovenous shunts as well as normal brain such as 2D-TOF-MRV, CE-MRV and SWI have been identified as useful adjunct sequences for patients with intracranial high-flow lesions. In some cases, DSA may not be able to depict normal parenchymal staining or venous drainage because of

the high shunt volume with a tendency for the contrast medium to pass directly through the lesions rather than the normal brain. MRV has been shown to better depict normal venous structures in several reports. The fistula point is identified by the abrupt change in caliber and signal intensity between the feeding arteries and the draining vein. 3D reconstructions help estimate the course of the abnormal vessels and the morphological features of the pial shunt [9,10].

In our case, we carefully analyzed the angioarchitecture of PAVF on MRI and MRA, which provided useful information not only for accurate diagnosis but also for treatment planning. Transarterial glue embolization was performed and resulted in nearly complete occlusion of the fistula and no postprocedural neurological deficit. Since intracranial PAVF often leads to high morbidity and mortality, treatment is usually attempted in most cases. The goal of treatment is to disconnect the fistula site, either by endovascularly or surgically, without the necessity of varix resection. In particular, endovascular method is increasingly considered as a safe and effective option in management of PAVF [2,8,11].

Conclusion

PAVF is a rare intracranial high-flow vascular lesion that can result in high morbidity and mortality. When the drainage of PAVF involves a dilated, but already formed VOG, it should be distinguished from other vascular lesions located in this area, including true VOGM, DAVF in Galen region, and AVM with VOG drainage and dilatation. Conventional MRI and MRA are noninvasive modalities that can provide valuable information regarding the anatomic localization of the fistula point, the feeding arteries, the venous sac, and their relationship with surrounding structures. These techniques are helpful not only for accurate diagnosis but also for treatment planning.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient

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