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SNI: Unique Case Observations

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Case Report

Atypical slow-flow paramedian AVM with venous varix

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ABSTRACT

Background: Cerebral arteriovenous malformations (CAVMs) are either clinically silent or symptomatic. The most common presentation in more than half of all CAVMs presenting patients is hemorrhage which is accompanied by long-standing neurological morbidity and mortality. This report presents a case of an atypical large, slow-flow paramedian AVM with a dilated venous varix managed with surgery. The impact of the intraoperative findings on the diagnosis and the operative technique will be discussed.

Case Description: In otherwise, healthy 26-year-old male complained of repeated episodes of generalized seizures and loss of consciousness. Brain magnetic resonance imaging (MRI) revealed a right parietal paramedian arteriovenous malformation (AVM) with signs of an old hemorrhagic cavity beneath it. Digital subtraction angiography demonstrated a slow-filling AVM with dilated venous varix drains into the superior sagittal sinus. However, the exact point of drainage cannot be appreciated. The filling of the AVM occurred precisely with the beginning of the venous phase. Intraoperatively, we noticed a whitish spherical mass, thick hemosiderin tissue, and a large cavity below the nidus; then, a complication-free complete microsurgical resection of this high-grade AVM was performed. Postoperatively, the patient suffered two attacks of seizures in the first few hours after the surgery, for which he received antiepileptics. MRI was clear during follow-up, and the patient was seizure-free and neurologically intact.

Conclusion: Parietal convexity AVMs are challenging lesions to tackle. However, the chronicity and the slow-filling of the AVM, in this case, can render the surgical pathway more direct and accessible.

Keywords: Cerebral arteriovenous malformation, Microsurgical resection, Venous varix

INTRODUCTION

Arteriovenous malformations (AVMs) of the brain are congenital lesions with <1% occurrence. AVMs are either clinically silent or appear with seizures, spontaneous intracranial hemorrhage (ICH), or headaches.[17] The diagnosis increases with the patient's age as it correlates positively with the hemorrhage incidence.[13] However, during the diagnosis, peculiar considerations should be focused on the presence of particular characteristics, such as venous varix within the lesion.

A venous varix, also known as an arterialized venous aneurysm or ectasia, is a significantly large vessel, no less than double the diameter of the draining vein.[14] In this article, we present a case of

an atypical large, slow-flow paramedian AVM with a dilated venous varix that was managed with surgery. The impact of the intraoperative findings on the diagnosis and the operative technique will be discussed, respectively.

CASE PRESENTATION

A 26-year-old male presented with repeated episodes of generalized tonic-clonic seizures with loss of consciousness for the past 2 months and was resistant to dual full-dose antiepileptic drugs. The patient had a 7-year history of severe progressive headaches with an affected mentality which resulted in school dropout. The neurological examination was intact, with no cutaneous telangiectasia or angiomas, with a Glasgow Coma Scale (GCS) of 13 (E3, V5, M5).

Brain magnetic resonance imaging (MRI) revealed a right parietal paramedian AVM, bounded posteriorly with the parieto-occipital sulcus, and accompanied by signs of an old hemorrhagic cavity beneath it which extends to the atrium of the right lateral ventricle [Figures 1 and 2]. Digital subtraction angiography revealed a slow-flow AVM with dilated venous varix emptying into the superior sagittal sinus (SSS); however, the exact drainage point cannot be appreciated. The arterial source was mainly from the middle cerebral artery's (M4) parietal cortical branches as it entered the deep part of the AVM. This AVM has a Spetzler-Martin grade of 2 (size 3.6 cm; 2 points, superficial venous drainage; 0 points, and non-eloquent area; 0 points). According to the supplementary S-M grading system, the AVM receives an additional two points (age at resection 26 years; 2 points, previous rupture; 0 points, and compact; 0 points), giving a combined grade of 4.

The filling of the AVM occurred at the end of the arterial phase, exactly with the beginning of the venous phase [Figure 3], which is not quite typical for an AVM with such a size and location and confirmed to be a slow-filling AVM.

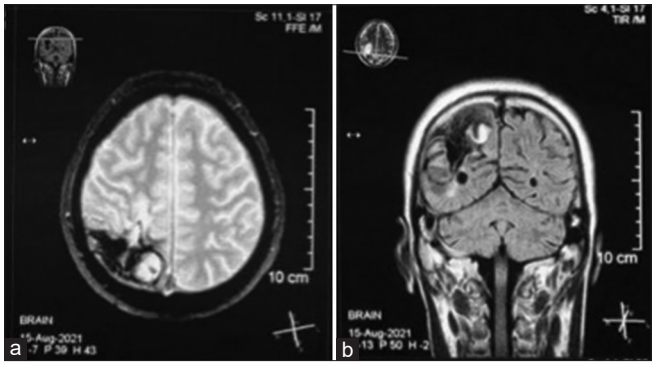

Figure 1: (a) Axial T2 magnetic resonance imaging (MRI) weighted image demonstrated a linear and round flow void features of a vascular lesion and an area of parenchymal hypointensity indicating a previous hemorrhage. (b) Sagittal view flair MRI demarcates a cavity and hemosiderin accumulation.

Microsurgical excision was performed to extirpate the AVM through a right paramedian parietal craniotomy. Intraoperative monitoring was applied in the form of somatosensory-evoked potentials, motor-evoked potential, and free-running electroencephalogram. During the initial exposure and arachnoid dissection, a whitish spherical mass (around 2 cm) was identified, dissected, and found to be attached to the top of the nidus; a large cavity was located lateral and deep to the nidus. The cavity was lined with thick white gliotic membranes and hemosiderin accumulation. After the elimination of all the feeding arteries, the nidus was dissected. The last step intraoperatively was the identification of a large draining vein that ends medially in the falx cerebri rather than drained directly to the SSS, which is then clipped and cauterized, followed by hemosiderin dissection and evacuation.

In the postoperative course, the patient had intact consciousness (GCS 15) and no weakness, with two attacks of seizures, which occurred in the first few hours after the surgery, for which he received antiepileptics. At 11 months, follow-up revealed a clear brain MRI and neurologically intact patient and seizure free as Engel scale was Class I.

DISCUSSION

The disturbed hemodynamic flow in the AVM nidus and the increased arterial pressure on the draining vein is regarded as the pathophysiological basis for the development of venous varix, which is defined as a vessel accompanying brain AVMs positioned either para- or intranidal or distant from the nidus, with diameter 2 times greater than that of the draining vein. Venous varix may be open, with an existing distal site and a proximal entrance, or "closed" with merely an entry. A venous varix is a benign, usually asymptomatic lesion;

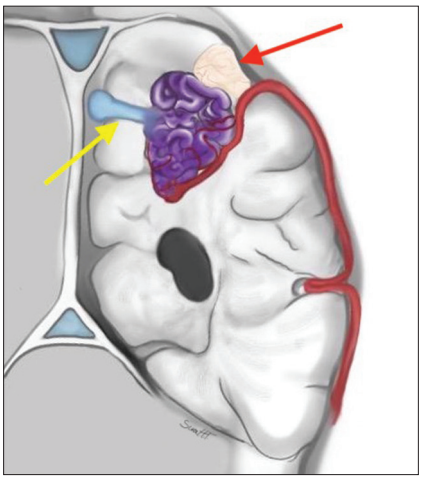

Figure 2: Artistic depiction of the paramedian arteriovenous malformation (AVM), in this case showing, arterial feeders, the AVM nidus position, venous varix (red arrow), and the draining vein (yellow arrow).

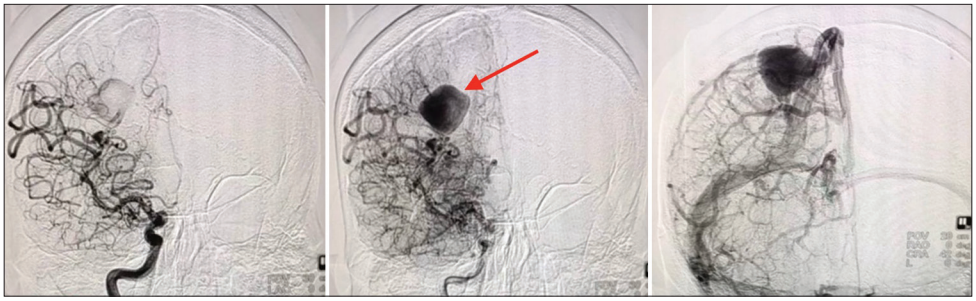

Figure 3: Serial angiography of the right carotid artery reveals a slow-filling vascular lesion in the parietal lobe. It appears to be fed by the M4 segment of the middle cerebral artery, giving branches to its base. The draining vein appears exceedingly enlarged, forms varix (red arrow), and ends in the superior sagittal sinus.

however, when it becomes symptomatic, it is suggested to carry a higher risk of AVM rupture.[3-5,7-10,12-14]

AVMs are divided into high-flow and slow-flow subtypes according to the blood flow rate. A high-flow AVM differs from the slow-flow subtype by producing structural fluctuations in the feeding and draining vessels, resulting in arterial smooth muscle hyperplasia, fibroblasts, and "fibromuscular cushions." On the other hand, slow-flow AVMs have a relatively common occurrence, and they can mimic the clinical presentations of diseases such as multiple sclerosis and brain tumors; their differentiation is crucial in determining the therapeutic implications. On imaging, slow-flow AVMs are identified by a slim rim of low to absent signal intensity encircling a central area of variable signal intensity on T1- and T2-weighted images. In some cases, the MRI only shows punctate well-defined low signal intensity or irregular and slightly decreased signal intensity in an area corresponding to calcification on a non-contrast CT scan which renders its identification difficult. Potential evidence of slow-flow AVMs includes the presence of hemorrhage or, in some cases, calcifications.[4,9] The management of high-flow AVMs includes either embolization using the Onyx alone or by surgical resection following embolization. While the treatment options for slow-flow AVMs differ, as they consist of conservative measures for asymptomatic cases, laser therapy, and sclerotherapy.[6]

Brain AVM has various clinical presentations, including seizures and ICH.[10,16] Brain AVM is considered one of the possible causes of hemorrhage in younger adults.[10] Hemorrhage accounts for part of AVM presentations and is related to increased morbidity and mortality.[5] The yearly percentage of AVM rupture is 2–4%; risk factors include AVM with deep venous drainage or deep location, infratentorial location, and presence linked with aneurysms.[7] ICH is the most frequent type of hemorrhage, which is responsible for 60% of the cases.[17] Other potential presentations of AVMs are headaches, focal neurological deficits, altered mental status, or incidental findings.[13]

Seizures constitute the most frequent presentation in cases of unruptured brain AVM, and they are the 2nd most common symptom after hemorrhage. Seizures might develop as an initial symptom or emerge during the follow-up to a hemorrhagic episode. Seizures occur in roughly 18–40% of cerebral AVMs; the outcome is favorable with antiepileptic medications. Generalized seizures are most commonly seen with AVMs (30%). On the other hand, in patients with seizures, there is a significantly low risk of enduring an AVM rupture within the follow-up period. In our case, despite the full dose of a dual regimen of antiepileptic medications, the patient experienced multiple episodes of generalized tonic-clonic seizures. Furthermore, patients that present with seizures are not more liable to suffer an AVM rupture during follow-up.[2,8]

Venous drainage plays a significant role in AVM rupture and hemorrhage. In our case, the venous drainage took an indirect pathway to drain into SSS as it was found to end in cerebral falx intraoperatively. This complexity and tortuosity of the venous drainage, in addition to the large-sized paramedian AVM, has impacted the hemorrhagic tendency of the lesion resulting in multiple previous hemorrhages, which were detected intraoperatively as gliotic mass, hemosiderin accumulation, and capsule formation based on imaging and intraoperative findings.[11] Previous unnoticed microhemorrhages, which indicate the chronicity of the case, may explain the thick gliotic capsule or membrane surrounding the paramedian AVM, which has, despite the complications of these previous silent hemorrhages and the damage to the adjacent tissue, assisted in establishing the dissection plan around the parietal convexity AVM and its isolation from the surrounding parenchyma, rendering the extirpation of the lesion more straightforwardly.[1]

There are multiple theories for forming venous varix, which is the increased arterial pressure on the draining vein and disturbed hemodynamic flow in the AVM nidus. Theoretically, varix veins are considered benign lesions, but their association with AVMs warrants careful surgical intervention due to the increased risk of rupture and uncontrolled bleeding. However, venous varix, in our case,

represents a wholly thrombosed vascular dilatation that and readily resected intraoperatively. Complete resection of the cavity membrane and the hemosiderin collections was emphasized to decrease the risk of postoperative seizures.

Angiography revealed the parietal convexity AVM with slow-filling and occult presentation, which made it first misdiagnosed as an aneurysm or cavernoma. The slow-filling is commonly seen in small-sized AVM, which counteracts our case; however, it is thought to be attributed to the compression by the pressure of hematoma and the formed occult inside the nidus.[15]

In summary, the surgery of this particular case was easy-going compared to other supratentorial AVMs in the same size and location. This mainly contributed to sufficient space provided by the previous hemorrhage and the thick membrane that prevented the adhesion of brain tissue to the AVM.

CONCLUSION

Supratentorial AVMs are difficult lesions to tackle. However, in this case, the slow-filling parietal convexity AVM and the chronicity can render the surgical pathway more accessible.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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