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Surgical Resection of a Progressive Giant Arteriovenous Malformation After 13-year Follow-Up

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Abstract: Giant arteriovenous malformation (AVM) is a complex and relatively rare congenital lesion with high morbidity and mortality. Its optimal treatment, however, remains controversial. Normal perfusion pressure breakthrough (NPPB) is a potentially devastating complication following surgical resection. Generally, strict blood pressure control is particularly recommended for preventing this phenomenon. Here we present a case of a 21-year-old patient with a progressive giant AVM who developed frequent seizures and subsequently underwent microsurgical total resection after 13-year follow-up, complicated by NPPB. Hypertensive hypervolemic treatment rather than strict blood pressure control was administered postoperatively; however thalamic infarction occurred. During the 1 year of follow-up, the patient remained seizure-free with only mild right-sided hemiparesis.

This case highlights that, in view of potential growth of the lesion, early intervention is necessary when possible. Microsurgical resection is challenging but remains to be an effective option for eliminating such giant AVM, and it is vital to keep risks associated with surgery in mind, such as NPPB. Moreover, whether blood pressure control is needed or not should be individualized.

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Abbreviations: AVM = arteriovenous malformation, ICA = internal carotid artery, MCA = middle cerebral artery, MRI = magnetic resonance imaging, NPPB = normal perfusion pressure breakthrough, PCA = posterior cerebral artery.

INTRODUCTION

Arteriovenous malformation (AVM), a lesion thought to be congenital in origin, is a major cause of hemorrhagic stroke in children with 2% to 4% annual bleeding risk and a 17% to 90% lifetime risk of rebleeding.^{1–3} The risk of disability after bleeding is 30% to 35%, whereas the risk of mortality after bleeding is 5% to 10%.¹ The most common presentation is hemorrhage, followed by seizure, headaches, and focal symptoms.⁴ The indications for surgical treatment of AVMs remain controversial. Giant AVMs (ie, those >6 cm in maximum

diameter) are a small group of AVMs but pose an especially difficult challenge, carrying a higher risk of morbidity and mortality than do smaller AVMs. Normal perfusion pressure breakthrough (NPPB), referred to the occurrence of brain edema and/or hemorrhage following removal of an AVM, is a main limiting factor for giant AVMs resection.⁵ Given this phenomenon, strict blood pressure control is particularly recommended. We herein present a case of a 21-year-old patient with a growing giant AVM who developed frequent seizures and subsequently received surgical resection complicated by NBBP and thalamic infarction after 13-year follow-up.

CONSENT

The patient signed informed consent for the publication of this case report and any accompanying images.

CASE REPORT

A 21-year-old man presented with a 2-year history of tonic-clonic seizures. At the age of 8, he was admitted with a sudden onset of headache, nausea, and vomiting. Magnetic resonance imaging (MRI) scans showed hemorrhage in the left temporal lobe associated with a Spetzler–Martin grade V AVM (Figure 1A). Conservative management was instituted, and the above symptoms gradually improved. However, he developed frequent generalized tonic-clonic seizures 2 years ago, which remained poorly controlled despite treatment with sodium valproate (500 mg three times a day). He was readmitted and neurological examination was unremarkable. Repeat MRI demonstrated a significant increase in size of the lesion (110 mm × 75 mm × 75 mm) involving the temporoccipital lobe (Figure 1B). Angiography confirmed the enlarged AVM, and it was mainly fed by branches of the left external carotid, internal carotid (ICA), middle cerebral (MCA), and posterior cerebral arteries (PCA), draining into the transverse, sigmoid and sagittal sinus (Figure 1C–E).

Pre-operative embolization failed to reduce flow to the nidus (Figure 1F). After 2 weeks, the lesion was circumferentially exposed along the feeding arteries, and thereafter was resected uneventfully. Several minutes later, however, multifocal bleeding in the surgical bed occurred, with massive parenchymal swelling. The phenomenon was considered to be the NPPB. The ICA adjacent to the origin of the posterior communicating artery was clipped proximally, and the bleeding was somewhat weakened but still uncontrollable. Ultimately, this condition was effectively held by clipping the left P1 segment. As such, collateral blood flowed to the territories of left MCA and PCA via the anterior communicating artery and left A1 segment (Figure 2A). It is of note that the A1 segment was relatively thin in diameter (Figure 2B). Instead of controlling blood pressure, hypertensive hypervolemic therapy was instituted during the initial 48 hours postoperatively in consideration of the narrow collateral vessel. Dopamine was intravenously administered to

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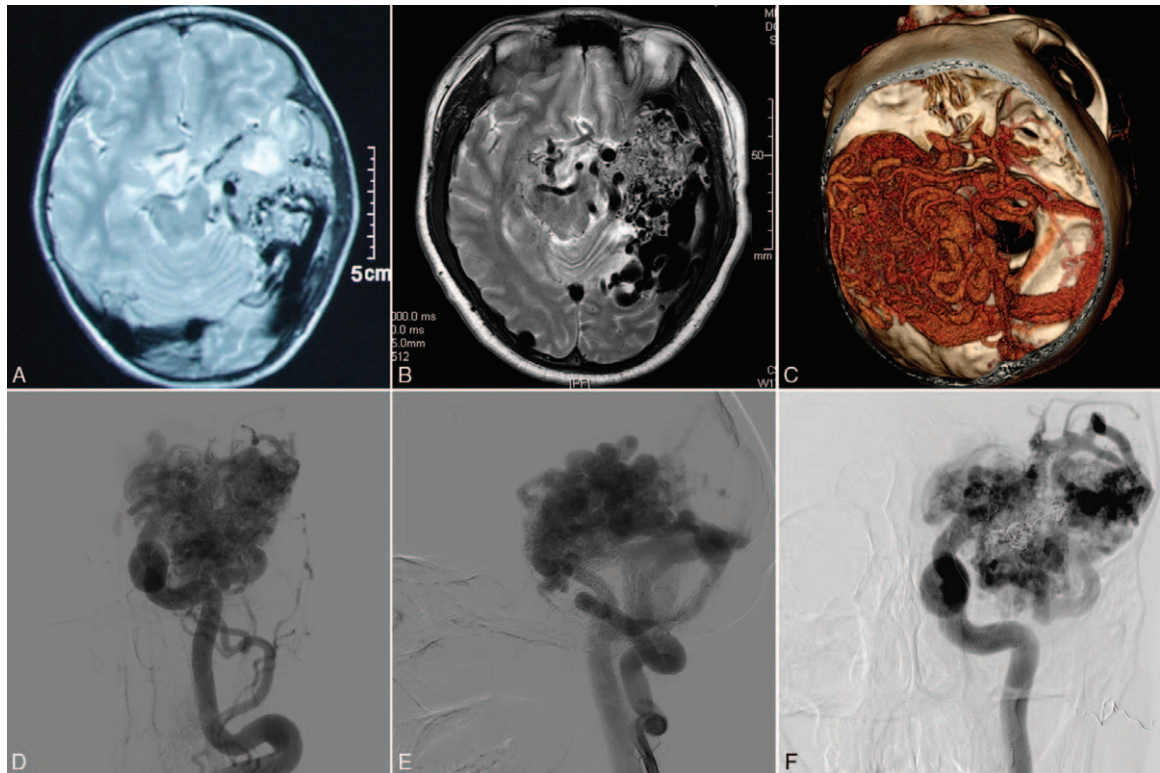


FIGURE 1. A and B, Axial magnetic resonance imaging scans show the arteriovenous malformation (AVM) increases in size during a 13-year period (2001 and 2014, respectively). C, Computed tomography angiography reconstruction reveals a giant Spetzler–Martin Grade V AVM involving the left hemisphere. D, Left common carotid angiogram shows the nidus fed by branches of the external carotid, internal carotid, and middle cerebral arteries. E, Left vertebral angiogram demonstrates the nidus also fed by branches of the dilated posterior cerebral artery. F, Left internal carotid angiogram shows that coil embolization fails to decrease the flow and the volume of the nidus.

induce a mild hypertension (systemic blood pressure, 140–150 mmHg); meanwhile, crystalloid and colloid solutions (1:3) were intravenously administered to maintain a central venous pressure of 5 to 12 cm H₂O. On postoperative day 1, however, he presented with right hemiparesis (1/5) related to a left thalamic infarct (Figure 2C). Antiplatelet therapy was instituted, and his neurologic status remained stable. He remained seizure-free with mild right-sided hemiparesis (4/5) at the 1-year follow-up, and angiography showed complete obliteration of the AVM and collateral flow to the left MCA (Figure 2D).

DISCUSSION

Giant AVM poses a significant treatment challenge, carrying a high risk of morbidity and even mortality. In terms of treatment decision making, multimodality treatments including embolization, microsurgery, and stereotactic radiosurgery were available; however, it should be individualized.⁶ In our patient, the staged embolization failed to reduce the blood flow to the AVM. Although surgical resection remains one of the most effective ways for eliminating giant AVMs, controversy exists regarding the indication.⁷ Klopfenstein and Spetzler have advanced that high-grade AVMs contributing to multiple hemorrhages or progressive symptoms should be surgically excised.⁸ Therapeutic goals include reducing the risk for bleeding, mass effect, and seizures.⁹ Our patient suffered from 2-year history of

uncontrollable frequent seizures, which subsequently halted by total resection of the lesion. Besides, in consideration of potential growth of the lesion, early intervention is essential when possible.

The NPPB theory was initially described by Spetzler et al¹⁰ in 1978 to explain the malignant edema or hemorrhage that sometimes occurred following resection of high-flow AVMs. The brain parenchyma surrounding the nidus is chronically hypoperfused; as a result, the local vessels chronically dilate and lose auto-regulatory capacity. Thus, following removal of an AVM, these chronically dilated low-resistance vessels cannot compensate for increased perfusion, and ultimately lead to hyperemia, edema, or hemorrhage.^{10,11} Currently, the occurrence of NPPB is the main limiting factor for high-flow AVMs resection.⁵ As usual, strict blood pressure control under normal values, especially systolic blood pressure (<140 mmHg), is recommended on the postoperative period to avoid NPPB occurrence.⁵ Moreover, mannitol, steroids, barbiturate coma, and hyperventilation have also been used in treatment of NPPB.¹¹

In our patient, the NPPB occurred during the surgical resection, and the proximal segments of left ICA and PCA had to be sacrificed via clipping. In that case, the blood flows into ipsilateral MCA and PCA through the narrow A1 segment. Consequently, the “2H” treatment (hypertension and hypervolemia) rather than accurate control of systemic blood pressure was administered, aiming at increasing regional cerebral blood flow, and brain tissue oxygenation. It is noteworthy that there

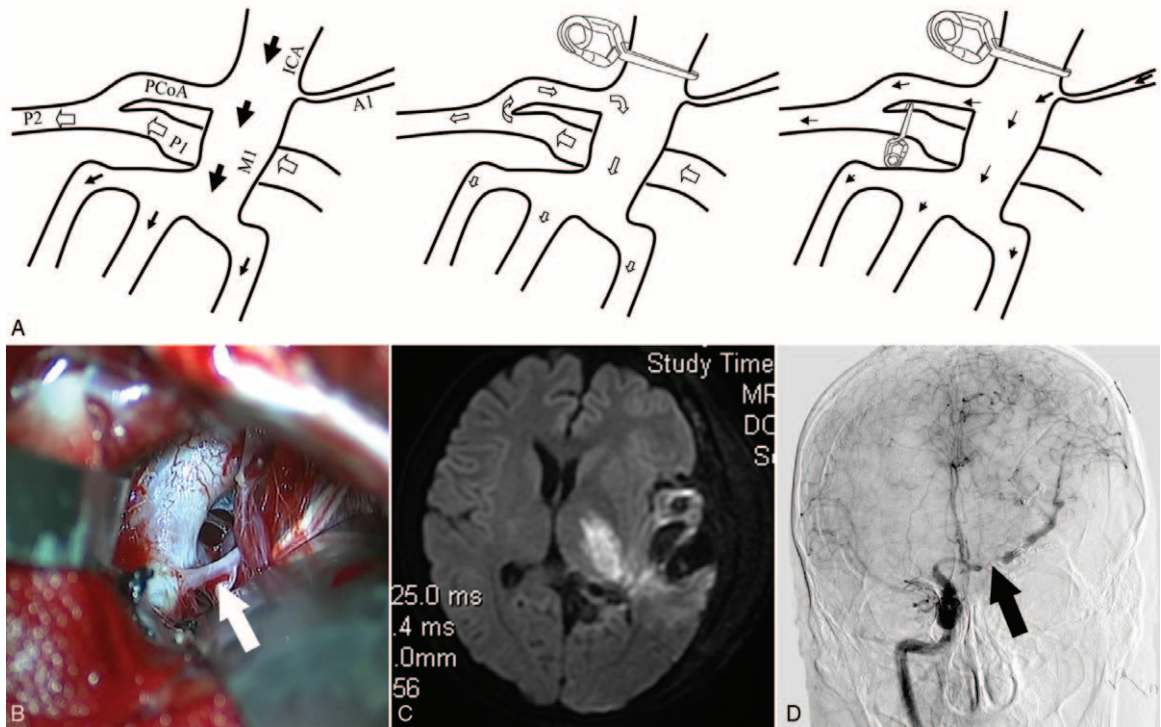


FIGURE 2. A, Schematic illustration shows cerebral blood flow changes before and after clipping the proximal internal carotid artery and P1 segment. B, Intraoperative micrograph demonstrates a left thin A1 segment (white arrow). C, Diffusion-weighted image on postoperative day 1 reveals a left thalamic infarct. D, Follow-up right internal carotid angiogram shows complete resection of the lesion, and collateral circulation filling to the left middle cerebral artery territory via the narrow A1 segment (black arrow).

are potential dangers in using this therapy such as aggravation of brain edema and increase of intracranial pressure. Unfortunately, the left thalamic infarct occurred due to the inadequate collateral circulation from the narrow A1 segment to the PCA deep branches. Such an ischemic event might be prevented if the blood flow from the left P1 segment was preserved partially.

CONCLUSION

In conclusion, early intervention of the lesion is necessary when possible in view of its potential growth. Although challenging, microsurgical treatment is an effective way to approach such giant AVMs, and the related risks, such as NPPB, should be kept in mind. Moreover, blood pressure control should be undertaken on a case-by-case basis.

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