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.

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 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 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

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 temporo-occipital 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
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. 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 loss 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 administrated, aiming at increasing regional cerebral blood flow, and brain tissue oxygenation. It is noteworthy that there...
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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