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Case Report

Spontaneous thrombosis of deep brain arteriovenous malformation in a patient with intraventricular and subarachnoid hemorrhage ☆,☆☆,★

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ABSTRACT

The most common manifestation of brain arteriovenous malformations (BAVM) is intracranial hemorrhage. The incidence of ruptured BAVM is 3.5 per 100,000 people per year. The mortality rate of ruptured BAVM within 1 month after diagnosis was 12.7%. Spontaneous thrombosis occurs in less than 1.5% of ruptured BAVMs. This phenomenon was still elusive. Up until now, the gold standard of imaging examination has been cerebral digital subtraction angiography (DSA), whose sensitivity and specificity reach 100%. We reported the spontaneous thrombosis of a ruptured deep BAVM. An 18-year-old woman presented with severe headache and vomiting. The patient also complained of seizures. There was no body weakness, skewed face, or slurred speech. Cerebral computed tomography (CT) showed extensive hemorrhage in the ventricular system and subarachnoid space. Cerebral DSA showed a left subcortical BAVM and was found to have spontaneous thrombosis 3 weeks later when the patient was about to be embolized. Spontaneous thrombosis of ruptured BAVM may occur after intracranial hemorrhage. In this patient, spontaneous thrombosis occurred within 3 weeks.

Introduction

BAVM is a lesion in the blood vessels of the brain characterized by the absence of capillary linings in an improper connection between arteries and veins [1]. This connection contains abnormal channels that tangle irregularly and widen. These irregular tangled channels are known as nidus [2]. Through the nidus, blood circulates from the arteries to the veins, causing a greater-than-normal blood flow in both the arteries and

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veins and contributing to elevated pressure on the venous side. High flow rate and shear stress, obstruction of the draining vein, arterial steal, and compartment formation are additional conditions that contribute to this complex vascular condition [3].

The total risk of bleeding in BAVM is 2%-4% per year, and the highest risk is at 5 years after the first symptoms appear. The risk of bleeding from an unruptured BAVM within 10 years is 1.3%-2.4% per year. The risk factors that need to be considered are the history of previous bleeding, deep location, infratentorial, and large size (>5 cm). If there are risk factors for previous bleeding, the risk of rebleeding is 15.4% and drops to 5.3% at 4 years and 1.7% after 5 years. The prevalence of BAVM patients with or without symptoms in the population is 10-18 per 100,000 people. A ruptured BAVM occurs in 3.5 out of every 100,000 people each year. Within 1 month of diagnosis, 12.7% of patients with ruptured BAVM passed away. Then the follow-up mortality rate in 1 year was 17.2%, and in 5 years was 22.1%. Severe disability after BAVM rupture after a 1-year follow-up was 75.3%, and after 5 years was 69.8% [2,4].

The computed tomography angiography (CTA) modality is better than magnetic resonance angiography (MRA), where the unruptured BAVM has a sensitivity of 96%. For a ruptured BAVM, the sensitivity is 87%. Meanwhile, for BAVM >3 cm, CTA has a sensitivity of 100%. Imaging examination as the gold standard so far has been cerebral DSA; its sensitivity and specificity reach 100% [2,5,6].

Spontaneous thrombosis of BAVM occurs in less than 1.5% of cases. The reason for this occurrence is not clear. It has been linked to some potential causes, including hemodynamic changes caused by hemorrhage, hypercoagulability, atherosclerosis, or thromboembolism. There have been several proposed pathomechanisms to explain spontaneous thrombosis of the main draining vein, including altered endothelium, thrombophilia/acquired coagulation problems, the mass effect associated with the nidus on the main draining vein, and venous stagnation associated with stenosis of the draining vein [7,8].

Until now, the modalities of definitive therapy for BAVM consist of 3 components: surgery, embolization, and stereotactic radiosurgery (SRS). The definitive treatment of BAVM is based on whether the BAVM is ruptured or unruptured [2,9,10].

Case reports

An 18-year-old woman was referred to the Emergency Room with complaints of severe headaches, accompanied by vomiting 2 weeks before. The headache was getting worse and worsening with activity and coughing, with a pain scale of 9-10. The patient also reported having seizures twice. The patient was unconscious during the seizure and regained consciousness after it ended. The seizure was characterized by jerking and his eyes moving upward, lasting for around 10 minutes. There was no body weakness, skewed face, or slurred speech. The history of hypertension and impaired coagulation was denied.

On physical examination, the patient exhibits normal blood pressure and is fully awake, with neurological deficits in the form of nuchal rigidity. The patient also showed normal pupillary reaction to light, round and equal, normal visual field tests, and acuity. Other cranial nerves were normal cerebellar function tests were normal.

The complete blood count laboratory examination was within normal limits. Noncontrast CT (NCCT) showed extensive hemorrhage in the ventricular system and subarachnoid space (Fig. 1). CTA of the head showed a suspected deep small AVM with a feeding artery from the left lateral lenticulostriate with a draining vein to the internal cerebral vein (Fig. 2).

The patient received conservative medications, including antivasospasm agents such as nimodipine 60 mg every 4 hours and antiseizure medication such as phenytoin 100 mg every 8 hours. No recurrent seizures occurred, and the headache improved after 3 days. The patient was planned to have cerebral DSA, with the results showing deep BAVM with a feeding artery from the left lateral lenticulostriate artery and draining vein to the internal cerebral vein and showing intranidal aneurysm (Fig. 3). The patient and family were explained regarding the findings and the embolization plan, but they decided to postpone it. The patient still received conservative medications. Three weeks later, when the patient had no symptoms and neurological deficits, she decided to do the embolization. When the patient underwent cerebral embolization, there was no deep BAVM that had previously received a feeding artery from the left lateral lenticulostriate artery and draining vein toward the internal cerebral vein, and no intranidal aneurysm was seen (Fig. 4), so the embolization

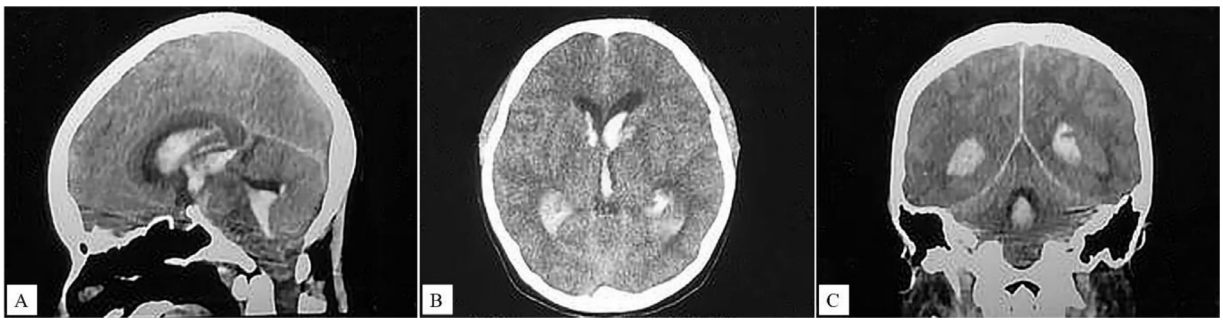

Fig. 1 – NCCT of the cerebral showed extensive hemorrhage in the ventricular system and subarachnoid space. (A) Sagittal view; (B) Axial view; (C) Coronal view.

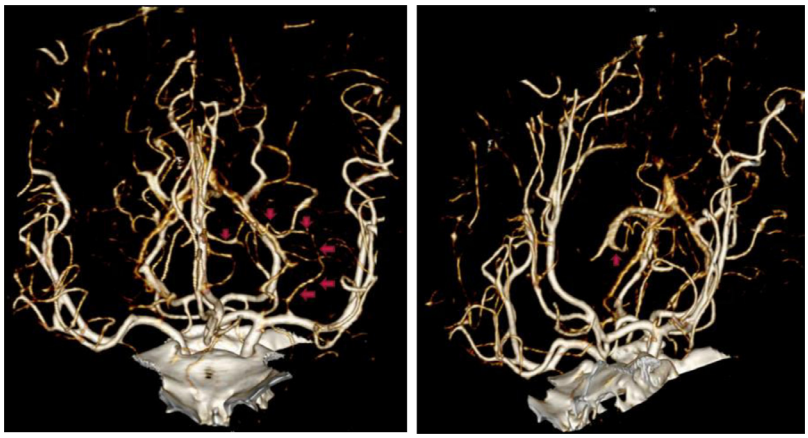

Fig. 2 – Head CTA showed suspected deep small AVM with feeding artery from the left lateral lenticulostriate with draining vein to the internal cerebral vein.

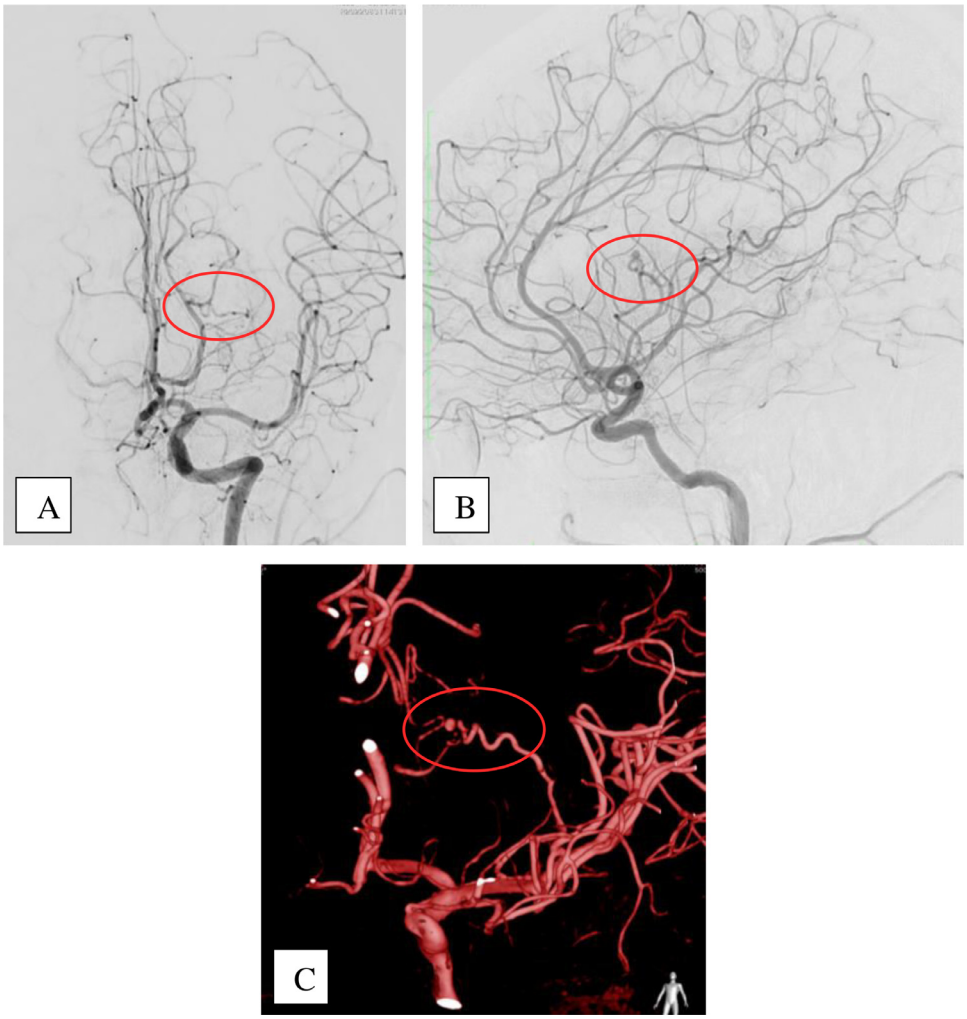

Fig. 3 – Anteroposterior and lateral view of cerebral DSA (A and B) showed deep BAVM with a feeding artery from the left lateral lenticulostriate artery and draining vein to the internal cerebral vein. The 3D reconstruction (C) showed intranidal aneurysm.

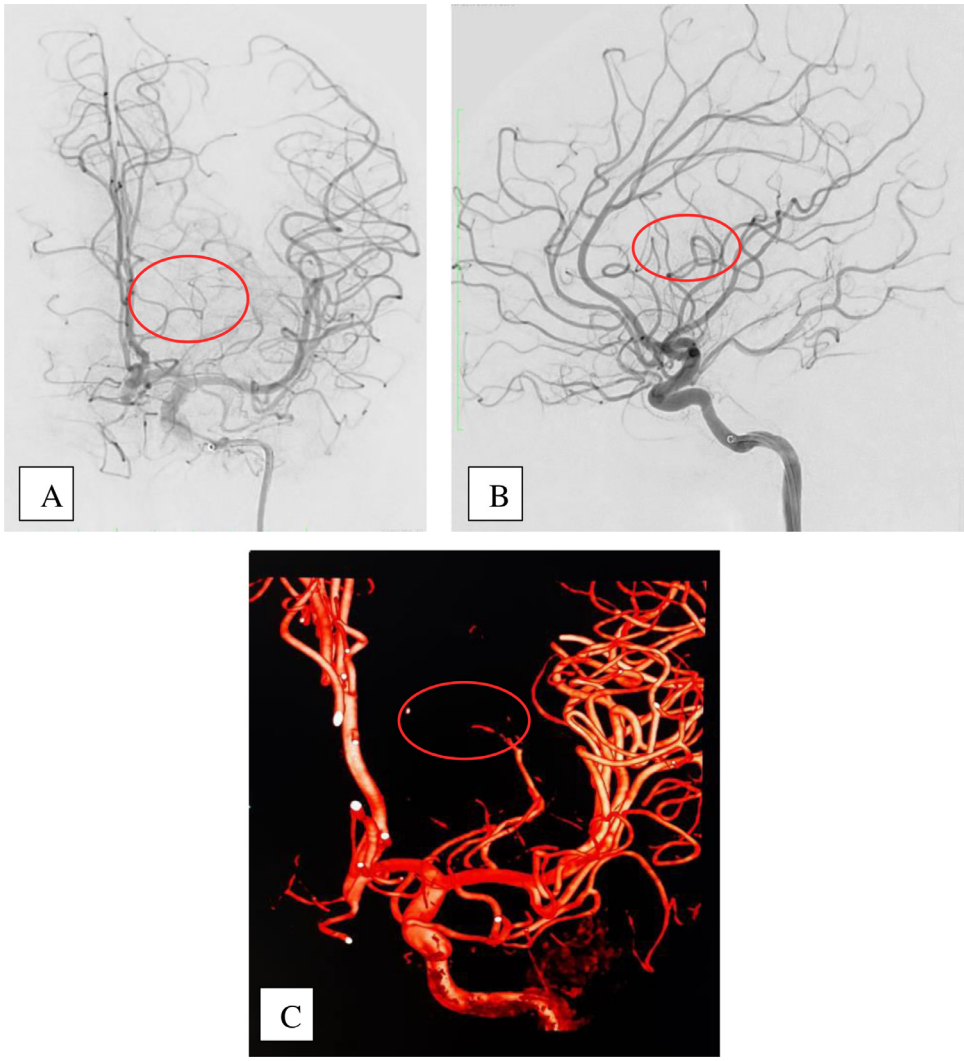

Fig. 4 – After 3 weeks, the anteroposterior and lateral view of cerebral DSA (A and B) showed there was no deep BAVM anymore, and was confirmed with the 3D reconstruction (C).

was discontinued. Three months after the last procedure, the patient was rechecked and found to be in good health with no neurological deficits or symptoms.

Discussion

The life-threatening hemorrhage, seizure, and progressive neurological deficits that indicate BAVM are widely recognized. The incidence of ruptured BAVM is 3.5 per 100,000 people per year [4,7]. Spontaneous thrombosis of BAVM is a rare but acknowledged event in the natural course of these lesions. It frequently happens unnoticed and is regarded as a fantastic result for preventing the patient from more dangerous events [11].

The age distribution at BAVM is 75% under the age of 45 years and 90% under the age of 55 years. The peak incidence is at the age of 20-30 years. BAVM usually has symptoms: 70% hemorrhage, 15%-40% seizure, 6%-14% headache, <10% neurological deficits, and 10%-15% asymptomatic (2%-4% accidental findings) [2]. In our case, the patient is an 18-year-old woman with no history of hypertension and impaired coagulation. The main complaint of our patient was headaches and seizures. This patient complained about severe headache with pain scale of 9-10; the pain had never been this severe. The patient also complained of seizures twice, but there was no history of seizures before.

For a patient with an unruptured BAVM, conservative measures are preferred. For some clinical conditions, such as headache that never improves, seizures not controlled with some antiepileptic drugs, the presence of focal neurological deficits, with angioarchitecture that indicates a high probability of rupture, definitive treatment can be considered. For patients with ruptured BAVM, definitive treatment is preferred. The modality of this therapy can be single or multimodal. The appropriate and adequate therapeutic modalities are selected based on clinical presentation, comorbidities, features of angioarchitecture, and available therapeutic modalities at the center [2,10,12].

Ruptured BAVM was the major cause of intraventricular hemorrhage and could lead to high mortality of 30%-80%, so we decided to do a further examination [13]. NCCT of our patient showed intraventricular and subarachnoid hemorrhage. The head CTA showed a suspected deep small BAVM. However, the gold standard of the imaging examination so far is cerebral DSA, its sensitivity and specificity reaches 100%. The cerebral DSA of the patient showed deep BAVM and intranidal aneurysm. The patient underwent cerebral embolization 3 weeks after, but no deep AVM and intranidal aneurysm were found, so the embolization was discontinued. Thus, we concluded that spontaneous thrombosis had occurred in this patient.

Some authors suggest that small, superficial AVMs with few arterial feeders may be more likely to experience spontaneous regression. However, the morphological and hemodynamic factors that may predispose an AVM to rupture or regression following venous thrombosis are still unclear. Another author identified 13 prior reports from the literature review and concluded that 9 patients had small lesions (77%), 3 with medium lesions, and 1 with large lesion [14]. Another literature review concluded that the majority of the reported cases had small nidus and distinctive cortical venous drainage in common [7]. Numerous angiographic series have shown that BAVM changes in size over time. The majority of BAVM grew in size, while a few of them stayed the same, and they rarely regressed [11]. However, since intracranial hemorrhage occurs in the majority of BAVM spontaneous occlusions at the time of diagnosis, we recommend aggressive treatment to prevent hemorrhage [7,15].

The occlusion of the feeding arteries or the draining veins is the possible mechanism in spontaneous thrombosis of BAVM. Some studies suggest that based on different pathological mechanisms, occlusion of feeding arteries may trigger angiogenesis that rebuilds collateral circulation, leading to BAVM recanalization, as opposed to occlusion of draining veins, which always results in permanent obliteration and is linked to a markedly elevated risk of hemorrhage. A contributing pathological aspect may be the fact that the spontaneous venous occlusion of BAVM invariably appears to occur in the presence of one or more dominant draining veins [15,16].

The majority of cases of spontaneous AVM obliteration share similar clinical symptoms with the classic subtype. A study of earlier literature revealed that intracranial hemorrhage and seizures were the most prevalent symptoms. After symptomatic intracerebral or subarachnoid hemorrhage, BAVM occludes in 75% of such conditions. The draining veins near the nidus appear to be compressed due to the increasing mass effect of the hematoma. Meanwhile, vasospasm caused by the accompanying subarachnoid hemorrhage may impair blood flow into the nidus [15,16].

The ultrasound fusion imaging (UFI) system is a new promising imaging technique that combines real-time ultrasound examinations with previously registered CT, MRI, or PET images has recently emerged in the field of neurology and has many reproducibilities. Peycheva et al. chose only healthy participants who had normal cerebral MR images to validate the diagnostic opportunities of the approach for normal anatomical structures. They integrate 3T-weighted MRI cerebral imaging with transcranial color-coded sonography in various insonation planes. They were able to discern many anatomical structures in detail, which would have been difficult to observe if merely ultrasonic transcranial color-coded sonography was used. Another advantage of the procedure is that we can repeat the examinations if we want to follow-up on our patients. During the examination, the patients also felt very comfortable. The examination also provided real-time data, such as the direction and velocity of the blood flow via the vessels, which was unable to be identified during an MR study [17].

Conclusion

Spontaneous thrombosis of ruptured BAVM is rare. It may occur after intracranial hemorrhage. In this patient, there was a small BAVM with a deep location in the left subcortical, and spontaneous thrombosis occurred within 3 weeks of being diagnosed angiographically.

Patient consent

The patient's family member gave written informed consent to the publication of this case report.

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