Treatment of a giant arteriovenous malformation associated with intracranial aneurysm rupture during pregnancy: A case report

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Received May 25, 2015; Accepted June 6, 2016

DOI: 10.3892/etm.2016.3505

Abstract. Arteriovenous malformations (AVMs) associated with aneurysm have rarely been reported in the literature. The present study reports the case of a 21-year-old pregnant female patient who presented with a subarachnoid hemorrhage and an intracranial hematoma located in the anterior end of the corpus callosum. Furthermore, an anterior cerebral aneurysm and an AVM were identified by digital subtraction angiography and magnetic resonance angiography. The aneurysm was clipped and the AVM was successfully removed by microsurgery. The diagnosis of AVM associated with an aneurysm was confirmed via intraoperative and postoperative pathological examinations. By performing a review of the current literature, issues and surgical considerations associated with AVM associated with aneurysm were analyzed.

Introduction

Arteriovenous malformations (AVMs) are uncommon vascular lesions that result from multiple abnormal connections between arteries and veins (1). The coexistence of AVMs and aneurysms has rarely been reported since its initial description in 1942 (2,3). A prospective study of 678 patients reported that the rate of AVMs associated with aneurysms was 18% (4). Stapf *et al* (5) demonstrated that 25% of 463 patients with AVMs had coexisting aneurysms. Despite the disease having severe effects on the health of sufferers, the causes and underlying mechanisms remain unknown. There exists three hypotheses that explain the pathogenesis of the disease; however, the consensus is that hemodynamic mechanisms serve an important role (6). In addition, it has been suggested that the coexistence of these two types of vascular disease in one patient may be a coincidence or the result of a congenital vascular malformation (7).

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Key words: arteriovenous malformation, aneurysm, treatment

Patients with an AVM and an aneurysm have been shown to have a greater risk of experiencing an intracerebral hemorrhage, as compared with patients with an AVM or an aneurysm alone (8,4). Thompson *et al* (6) reported that the rate of intracerebral hemorrhage in patients with AVMs and aneurysms was 27-62%. Furthermore, Brown *et al* (9) demonstrated that the incidence of intracerebral hemorrhage was 7% per year in these patients, which was 1.7% for patients with AVM alone.

This dual vascular disease presents a management challenge. The present study reports the case of a young pregnant patient with combined AVM and aneurysms who presented with a subarachnoid hemorrhage (SAH) at the 101st Hospital of Chinese People's Liberation Army (Wuxi, China). The authors of the present study experienced severe challenges when attempting to deal with the giant AVM and aneurysm in terms of the technology and management.

Case report

A 21-year-old female was admitted to Lujiang People's Hospital (Lujiang, China) on 6th June 2014 due to a short-term history of a severe headache, nausea and vomiting, in the absence of an obvious cause. The patient had no previous history of trauma. A computed tomography (CT) head scan revealed an intracranial hematoma in the right frontal lobe, and a SAH (Fig. 1). The patient was transferred to the regional center hospital where a magnetic resonance angiography (MRA) detected an AVM with a diameter of 5 cm in the right corpus callosum. The nidus was fed by the branches of the right anterior cerebral artery (ACA) and the right middle cerebral artery (MCA). In addition, there were multiple large cortical venous drains, one of which drained blood to the superior sagittal sinus, and one which drained blood to the sagittal sinus. Furthermore, a 3.0-mm enlarged blood vessel corresponding to an aneurysm was observed, although its existence could not be confirmed by an MRA (Fig. 2).

Digital subtraction angiography (DSA) confirmed the existence of a large AVM located in the right corpus callosum with multiple feeders from all branches of the ACA and MCA, and with drainage into the cavernous sinus and transverse sinus via deep temporal cortical veins. Furthermore, DSA clearly demonstrated an ACA aneurysm with a diameter of 3.0 mm adjoining the AVM (Fig. 3). Since the aneurysm was adjoining the AVM, it was difficult to evaluate the reason for the hemorrhage using CT scans. Therefore, the AVM and aneurysm

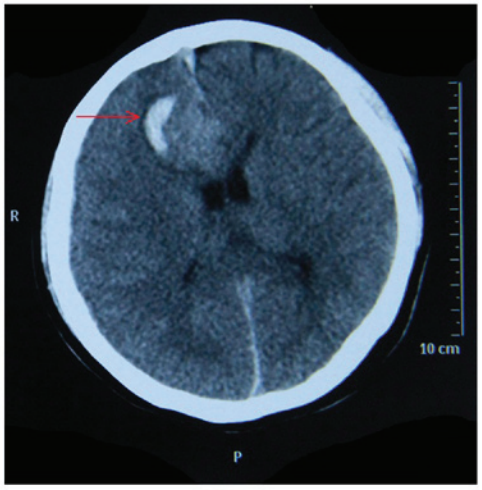

Figure 1. Preoperative computed tomography scan showed an intracranial hematoma in the right frontal lobe (red arrow) and a subarachnoid hemorrhage.

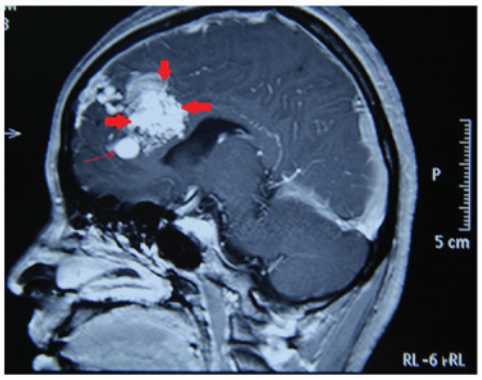

Figure 2. Preoperative magnetic resonance imaging demonstrated an arteriovenous malformation (thick red arrows) and associated aneurysm (thin red arrow).

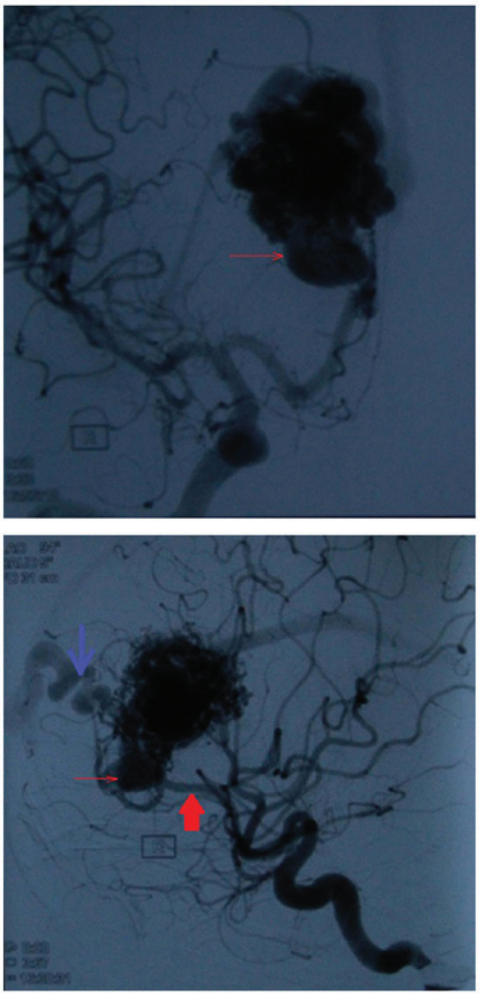

Figure 3. Preoperative digital subtraction angiography confirmed the results of the computed tomography scan and magnetic resonance imaging. (A) Venous phase, a large AVM and aneurysm are present, and the thin red arrow points to the aneurysm; (B) arterial phase, the blue arrow points to the draining vein, the thick red arrow points to the feeding artery and the thin red arrow points to the aneurysm.

were treated simultaneously to avoid re-bleeding. The hospital determined that the operation was too risky and the patient was admitted to the 101st Hospital of Chinese People's Liberation Army on day 15 following initial admission. A neurological examination involving use of the Glasgow Coma Scale (GCS) (10) detected no abnormalities (GCS score =15), and a CT reexamination showed that the SAH and intracranial hemorrhage blood had been absorbed.

A discussion involving neurology, neurosurgery and interventional radiology doctors determined that interventional vascular treatment was too risky, whereas microsurgery was considered a relatively safe method for treatment of the patient. The following day, the patient underwent a right pterional craniotomy, during which the bilateral A2 segment of the ACA was investigated using a longitudinal fissure approach to temporarily occlude the blood flow, while protecting the venous drainage system. The aneurysm was clipped completely after the parent arteries, aneurysm and main feeders of the AVM had been thoroughly explored. Subsequently, the right A3 segment was temporarily occluded (for 15 min, followed by release for 10 min) to explore the AVM, after which the part of the left A3 branches feeding the AVM were also clipped. Finally, the AVM was clipped integrally.

A pathological analysis (Fig. 4), performed as previously described (11), showed that the AVM was 5x4x4 cm (Spetzler-Martin grade 4) (12). Postoperative control of the patient's blood pressure was important and it was maintained below the normal (10-20 mmH_2g) (normal systolic pressure, 120-140 mmHg; normal diastolic pressure, 70-90 mmHg). The patient was discharged from the Neonatal Intensive Care Unit on 20th June 2014 after 3 days with a GCS score of 15/15. No nerve dysfunction was observed. A CT and CT angiography (Fig. 5) was performed and demonstrated that the surgery had been successful, that the aneurysm had been clipped completely, that the AVM had been entirely removed and that no venous drain had been injured. At the 6 month follow-up, the Glasgow Outcome Scale (13) score was 5. Informed consent was obtained from the patient.

Discussion

The coexistence of AVMs and cerebral aneurysms has rarely been reported. The annual incidence of spontaneous SAH is 10/100,000 worldwide, of which 75% are caused by intracranial

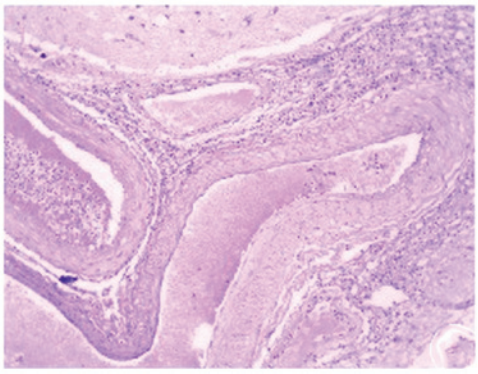

Figure 4. Postoperative histopathological examination showed abnormal, tortuously distributed vessels characteristic of an arteriovenous malformation.

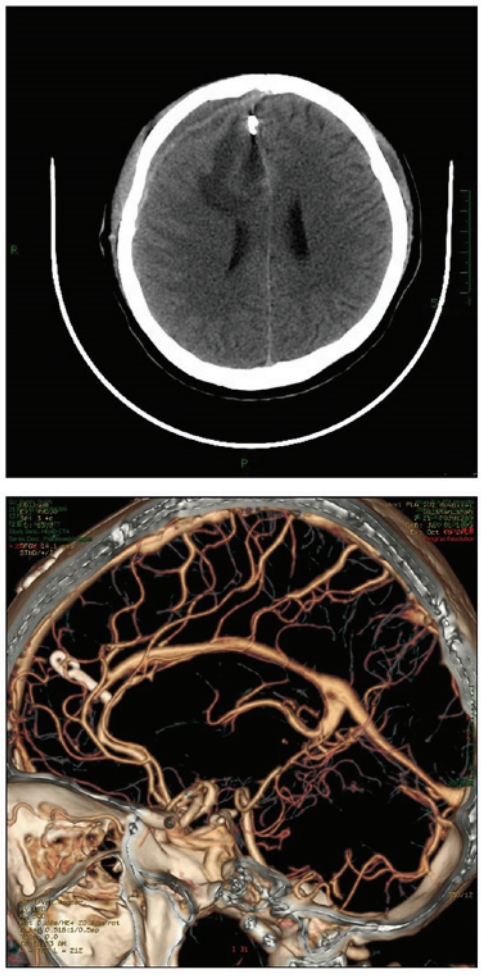

Figure 5. (A) Postoperative computed tomography (CT) scan showed that the hematoma and arteriovenous malformation had been completely removed. (B) Postoperative CT angiography demonstrated that the aneurysm had been clipped.

aneurysms (14,15). The incidence of AVM is 1.3/100,000 per year (16), and the reported incidence of AVM coexisting with intracranial aneurysm is 18% (4). Therefore, the incidence of aneurysms in patients with AVMs is higher, as compared with general patients. Furthermore, the risk of bleeding and re-bleeding are higher in patients with AVM and aneurysm, as compared with patients with either alone (8,4). In addition, in these patients, the risk of experiencing a hemorrhage increases over time (4). Marks *et al* (17) reported that aneurysms are an independent risk factor for re-bleeding. Therefore, it is important that microsurgery or endovascular treatment are performed early.

There are a number of systems used for the classification of AVMs associated with aneurysms; however, all systems are based on the location of the AVM relative to the aneurysm. The most commonly used classification system at present is that proposed by Perata *et al* (18). The patient in the present study was diagnosed with a flow-related aneurysm (type 2 classification). The position of the AVM relative to the aneurysm is important for deciding the appropriate treatment strategy and surgical approach. There has been significant debate regarding the most appropriate therapeutic strategy for AVMs associated with an aneurysm (19-23). Cunha *et al* (20) advised that priority treatment should be given to the symptomatic lesion or that both lesions should be treated simultaneously if the condition permits. Batjer *et al* (22) suggested that an intracranial aneurysm should be removed by microsurgery or endovascular embolization prior to resection of the AVM, since it may avoid rupturing the aneurysm during the surgery to remove the AVM. Conversely, Koulouris and Rizzoli (23) insisted that the AVM be removed first. Since numerous scholars consider that hemodynamic mechanisms serve an important role in the pathogenesis of the disease, the removal of AVM may lead to various hemodynamic changes causing the aneurysm to disappear spontaneously (6,7,9,14,20). In the present case, the aneurysm and AVM were adjacent and the feeding artery of the AVM was the parent artery of the aneurysm. Therefore, the AVM and aneurysm could be removed simultaneously in the same operation, according to Cunha *et al* (20). However, the aneurysm was clipped prior to removing the AVM in order to reduce the risk of the aneurysm rupturing during surgery.

At present, three treatment strategies have been described for patients with coexisting AVM and aneurysm, including microsurgery, endovascular embolization and radiotherapy. Factors, such as the size and location of the AVM, venous drainage, hospital equipment and the technique level of the surgeon, will determine the requirement for treatment and the optimum therapeutic strategy. In the present study, microsurgery was used to remove the AVM and clip the aneurysm entirely. As a result, the prognosis of the patient was good and the intracranial hematoma was cleared. However, if the lesions had been deeply seated or located within an important functional area, microsurgery treatment may not have been the ideal choice (21).

Previous studies have reported that embolization has a number of advantages, including minor damage to brain tissue, fewer symptoms, effectiveness, rapid recovery and a short hospital stay (24-26). A previous study used gamma knife radiotherapy for treatment (27); however, it has a long treatment cycle, its effectiveness has not been well documented and re-bleeding may occur (28,29). The present study describes a young pregnant woman with a large AVM, wide draining veins and a high risk of endovascular complications. Therefore, microsurgery was selected for treatment of the patient. A CTA reexamination demonstrated that the operation was

successful; the AVM had been removed completely and the aneurysm was clipped.

Previous studies have not reported particular technical difficulties associated with the removal or embolization of giant AVMs coexisting with an aneurysm. However, it is important to protect the draining veins and arteries supplying blood to the brain tissue via the AVM. Furthermore, it is important that the intraoperative and postoperative blood pressure of the patient is maintained relatively low in order to reduce the risk of re-bleeding.

In conclusion, AVMs associated with intracranial aneurysms are a complex lesion, and the treatment is dependent on the relative location of each lesion and its classification. In the present study, a patient with a type 2 AVM and associated aneurysm underwent aneurysm clipping, followed by resection of the AVM in a single operation, and showed a good clinical outcome.

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