Formation During Lifetime of Arteriovenous Shunt in Developmental Venous Anomaly That Caused Intracerebral Hemorrhage

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Key words
- Arterialized developmental venous anomaly
- Developmental venous anomaly
- DVA with arteriovenous shunting
- Vascular anomalies
- Venous angioma

Abbreviations and Acronyms
AVM: Arteriovenous malformation
AVS: Arteriovenous shunting
CT: Computed tomography
DVA: Developmental venous anomaly
MRI: Magnetic resonance imaging

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BACKGROUND: **Developmental venous anomaly (DVA) or venous angioma is a common anomaly of cerebral veins that is found incidentally in the majority of cases. There are few cases of arteriovenous shunting in DVA associated with a more malignant course of the disease. Whether these DVAs with shunts are of congenital pathology or lifetime formations is unclear.**

CASE DESCRIPTION: **We report a case of lifetime arteriovenous shunt formation in DVA that caused intracerebral hemorrhage in a child. The patient underwent 2 sequential direct surgeries: an emergency evacuation of the intracerebral hematoma and a scheduled excision of the DVA with arteriovenous shunting.**

CONCLUSIONS: **Arteriovenous shunting in DVA may develop during a lifetime and cause intracerebral hemorrhages. This case showed that localization of DVA with arteriovenous shunting in a noneloquent area enables its complete microsurgical excision with favorable functional outcomes.**

INTRODUCTION

Developmental venous anomaly (DVA) or venous angioma is a common anomaly of cerebral veins that is found incidentally in the majority of cases. In some patients, DVA may manifest with epilepsy, ischemic stroke, and intracranial hemorrhage.[1,2]

DVA evidence in cerebral angiography, computed tomography (CT) angiography, or magnetic resonance imaging (MRI) is the so-called caput medusae sign, formed via convergence of many dilated veins in an enlarged single collector vein. The differences between typical DVA and arteriovenous malformation (AVM) include the absence of arteriovenous shunt. Therefore DVA is characterized by contrast enhancement only in the venous phase during cerebral angiography.

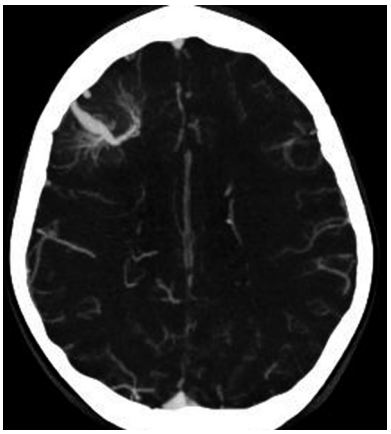

Figure 1. Computed tomography angiography: typical developmental venous anomaly in the right frontal lobe.

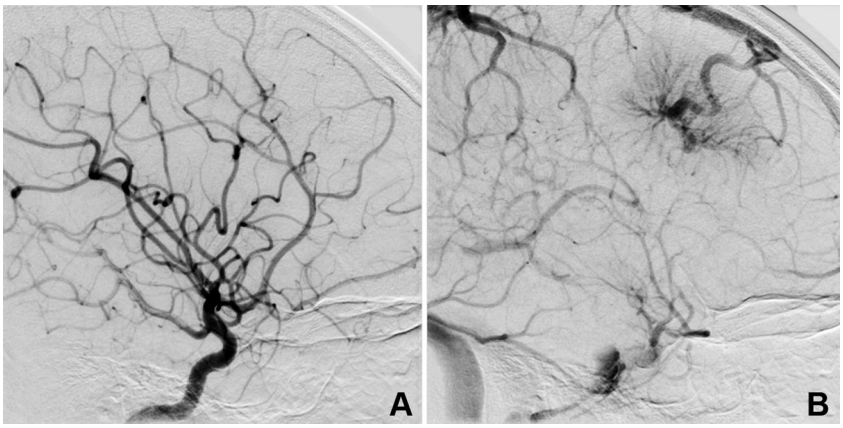

Figure 2. Right carotid angiography: no arteriovenous shunting in the malformation. (**A**) Arterial phase. (**B**) Venous phase.

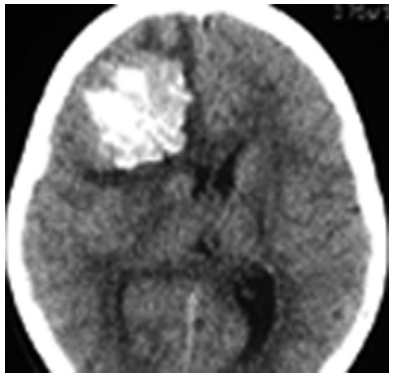

Figure 3. Brain computed tomography: intracerebral hemorrhage 40 mL in the right frontal lobe.

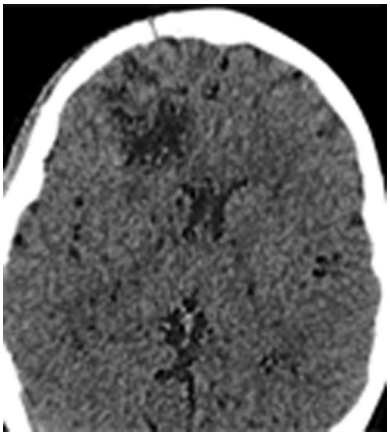

Figure 4. Brain computed tomography: posthemorrhagic cyst in the right frontal lobe.

There are few cases of arteriovenous shunting (AVS) in DVA associated with a more malignant course of the disease.[3-6] Some authors describe this condition as arterialized DVA. We believe that this term is hard to understand; therefore we use the definition "DVA with AVS." Whether these DVAs with AVS are of congenital pathology or lifetime formations is unclear.

The choice of therapeutic approach in DVA with signs of shunting is challenging. Here, we report a case of lifetime arteriovenous shunt formation in DVA that caused intracerebral hemorrhage in a child. The patient underwent 2 sequential direct surgeries: an emergency evacuation of the intracerebral hematoma and a scheduled excision of the DVA with AVS.

CASE REPORT

Patient S., female, aged 5 years, developed 2 episodes of seizures at 12-hour interval. Seizures were manifested by convulsions in the left extremities and short-term loss of consciousness. MRI and CT angiography (Figure 1) showed DVA in the right frontal lobe.

The diagnosis was confirmed via direct cerebral angiography (Figure 2). Valproic acid (Convulex) was prescribed in the age- and weight-based dosage. During 2 years of therapy, no seizures were reported. The dose was tapered to withdrawal. Subsequently, the patient felt well without any neurologic signs.

In 5 years, when the patient was 10 years old, her condition deteriorated with a sudden severe headache associated with subsequent nausea and vomiting. The patient was hospitalized in a community-based hospital, where CT revealed intracerebral hemorrhage in the right frontal lobe (Figure 3). Urgent craniotomy and evacuation of intracerebral hematoma in the right frontal lobe were performed. The postoperative period was normal. Brain CT showed no residual hematoma in 3 weeks (Figure 4). After rehabilitation the patient was admitted to the federal state autonomous institution "N. N. Burdenko National Medical and Research Center for Neurosurgery" of the Ministry of Healthcare of the Russian Federation (Moscow). No neurologic signs were evident. Cerebral angiography showed DVA in the right frontal lobe with arteriovenous shunt due to its blood supply from the branch of the middle cerebral artery (Figure 5).

Microsurgery to remove DVA with AVS was performed. During the intervention, abundant vessels with posthemorrhagic cyst were found on the cortex of the right frontal lobe after craniotomy and dura opening. A large artery from the right middle cerebral artery ran into the malformation along the cortex; it was considered a feeding vessel. At the upper pole of the malformation, a draining efferent vein to the upper sagittal sinus was identified. When the afferent vessel had been coagulated and cut, the malformation was isolated along the perilesional area. Notably, no typical node of AVM was identified, and malformation predominantly consisted of venous vessels, was easily coagulated, and slightly bleeding when cut. After complete DVA isolation, the draining vein was coagulated and cut. The malformation was removed via a single node.

During the postoperative period, the patient felt well, without neurologic deterioration. Brain CT showed no ischemic and hemorrhagic postprocedural complications (Figure 6). Postoperative cerebral angiography showed no evidence of residual DVA (Figure 7). At day 7 after surgery the patient was discharged

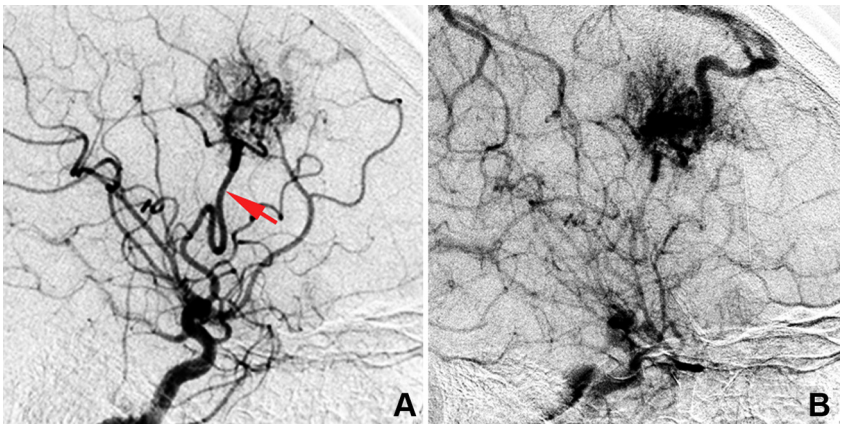

Figure 5. Right carotid angiography: arteriovenous shunting in the malformation. (**A**) Arterial phase. (**B**) Venous phase. The *arrow* indicates the main afferent vessel of malformation from the right middle cerebral artery.

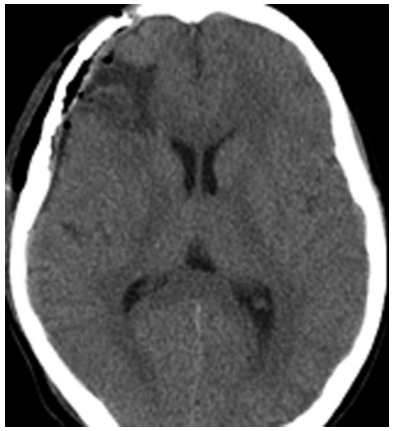

Figure 6. Postoperative brain computed tomography: no procedural complications.

home. A direct cerebral angiography in 1 year was recommended.

DISCUSSION

One hypothesis of the shunting in DVA suggests a high venous pressure leading to the capillary dilation and the formation of arteriovenous anastomosis.[1,7]

The venous hypertension and formation of an arteriovenous shunt are suggested to result from the draining vein thrombosis.[8] The increased blood flow in venous malformation due to shunting from the arteries may be a compensatory mechanism that facilitates the recanalization of clot-occluded sites. We suppose hemodynamic changes in DVA may be more severe in childhood, when human growth is accelerated. In our case it is unclear when the arteriovenous shunt has been formed: before or after a hemorrhage? Taking into account that the veins carrying the arterial blood are more prone to rupture,[1,5,7,9,10] we believe that the arteriovenous shunt has been formed before the hemorrhage.

The choice of treatment of DVA with AVS is challenging. Unlike AVM, DVA collects blood from the normal brain including eloquent areas.[4] An occlusion of the main venous DVA collector may result from the venous ischemia in this area.[3,9] This complication was defined as occlusive hyperemia.[11]

Some authors recommend the selective elimination of a DVA arteriovenous shunt with careful preservation of the draining vein patency via microsurgical excision, endovascular embolization, or radiosurgery.[1,3,5] Each of these methods is not universal and is associated with possible complications that are sometimes fatal.

In this case, intensive contrast enhancement of the abnormal malformation vessels in the early arterial phase of cerebral angiography indicates a high blood flow in the shunt. We considered this pattern DVA transformation into AVM.

The surgery was aimed to eliminate the risk of rebleeding. We decided to perform the total resection of DVA with AVS for several reasons:

1) By the time of surgery, the right frontal lobe around the DVA had been significantly damaged due to hemorrhage.
2) Because of major posthemorrhagic changes, it was unclear whether part of the vessels with arteriovenous shunting could be distinguished intraoperatively, as well as vessels carrying only venous blood. Therefore all veins around the perimeter of the main collector were cut, with the main draining vein cut last.
3) In case of partial resection, residual DVA could cause new formation of arteriovenous shunt, as it had already taken place in this patient.

This case showed that excision of DVA with AVS did not cause occlusive hyperemia and worsening of neurologic symptoms. This phenomenon has an explanation. The veins congested with arterial blood virtually do not contribute to the drainage in the normal brain; thus their excision or occlusion causes no venous hyperemia.[10] Also, it has been found that in AVM with 1 drainage—like in the presented case—the risk of occlusive hyperemia is substantially lower.[12]

Currently, we recommend repeated angio-MRI or CT angiography in all children with DVA, as well as direct cerebral angiography in cases of hemorrhage to exclude arteriovenous shunting.

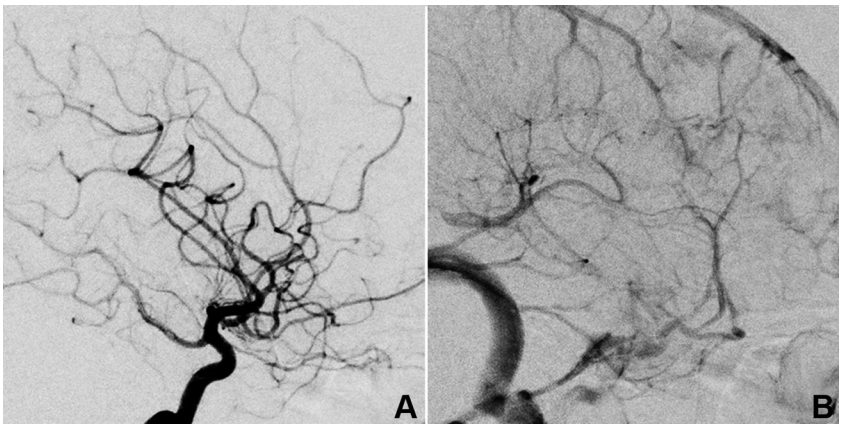

Figure 7. (**A** and **B**) Postoperative right carotid angiography: total resection of the DVA with arteriovenous shunting confirmed.

CONCLUSION

Arteriovenous shunting in DVA may develop during a lifetime and cause intracerebral hemorrhages. This case showed that localization of DVA with arteriovenous shunting in a noneloquent area enables its complete microsurgical excision with favorable functional outcomes.

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