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Angiographic Findings in Two Cases of Aneurysmal Malformation of Vein of Galen Prior to Spontaneous Thrombosis: Therapeutic Implications

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Summary: Two cases of aneurysmal malformations of the vein of Galen with later spontaneous thrombosis are reported. Angiograms before thrombosis in both cases showed mural type aneurysmal malformations of the vein of Galen with slow arteriovenous shunts and associated stagnation of contrast in the venous sac secondary to severe outflow restriction. Based on these angiographic findings, the patients were managed conservatively and the arteriovenous malformations of the vein of Galen thrombosed with good clinical outcomes. Twenty cases of spontaneous thrombosis previously reported in the literature are reviewed.

Index terms: Arteriovenous malformations; Aneurysm, arteriovenous; Interventional neuroradiology

Spontaneous thrombosis of an aneurysmal malformation of the vein of Galen (AVG) is a rare occurrence. The first reported cases were diagnosed after surgical removal (1–3). Later cases were treated by partial removal (4) or shunting (5). If a diagnostic test could predict which AVGs will thrombose and whether patients with spontaneous thrombosis do well clinically, embolization procedures or surgery could be avoided. We report two cases of mural type AVG (6, 7) where no embolization or surgical procedures were done based on the angiographic findings of slow flow and stasis of contrast material in the venous sac with severe obstruction of the venous outflow. Both AVGs thrombosed within 5 months with good clinical outcomes.

Case Reports

Case 1

A 4-month-old boy was evaluated because of increased head circumference. There was no cardiac failure. There was moderate motor developmental delay.

Computed tomography (CT) scan demonstrated ventricular enlargement with a prominent aneurysmal sac. A ventriculoperitoneal shunt was placed at 5 months of age. At age 9 months, the patient was referred for endovascular treatment. Preliminary angiography revealed a two-vessel fistula from the choroidal branch of the posterior pericallosal artery draining superiorly into the wall of a dilated median vein of the prosencephalon, the embryologic precursor of the vein of Galen (8) (Fig. 1A). The flow within the AVG was very slow, with stagnation of contrast within the venous sac. Severe outflow restriction was present in the falcine sinus. There was filling of the transverse sinus, sigmoid sinus, and jugular veins. Right and left internal carotid artery injections revealed no additional supply to the AVG.

No endovascular treatment was done because it was believed that there was a high probability of spontaneous thrombosis of the AVG. Five months later, magnetic resonance imaging (MR) revealed complete thrombosis of the AVG (Fig. 1C). At 3 years follow-up, the child is doing well, except for some mild problems with coordination. MR at this time shows almost complete resolution of any mass effect (Fig. 1D).

Case 2

A male neonate (38-week gestation) was evaluated following normal vaginal delivery because of a hypoechoic intracranial lesion seen on prenatal ultrasound 4 days prior to delivery. Head circumference was within normal limits. There was no evidence of cardiac failure at birth. The patient was neurologically intact.

MR at age 2 weeks revealed a dilated midline vein with stenosis at its junction with the falcine sinus. No evidence of cerebral atrophy or hydrocephalus was present. Since the patient was asymptomatic, no angiogram or treatment was done at this time.

Since a follow-up MR showed dilated ventricles, angiography was done at 4 months of age. The left vertebral artery injection revealed a slow flow mural fistula from the

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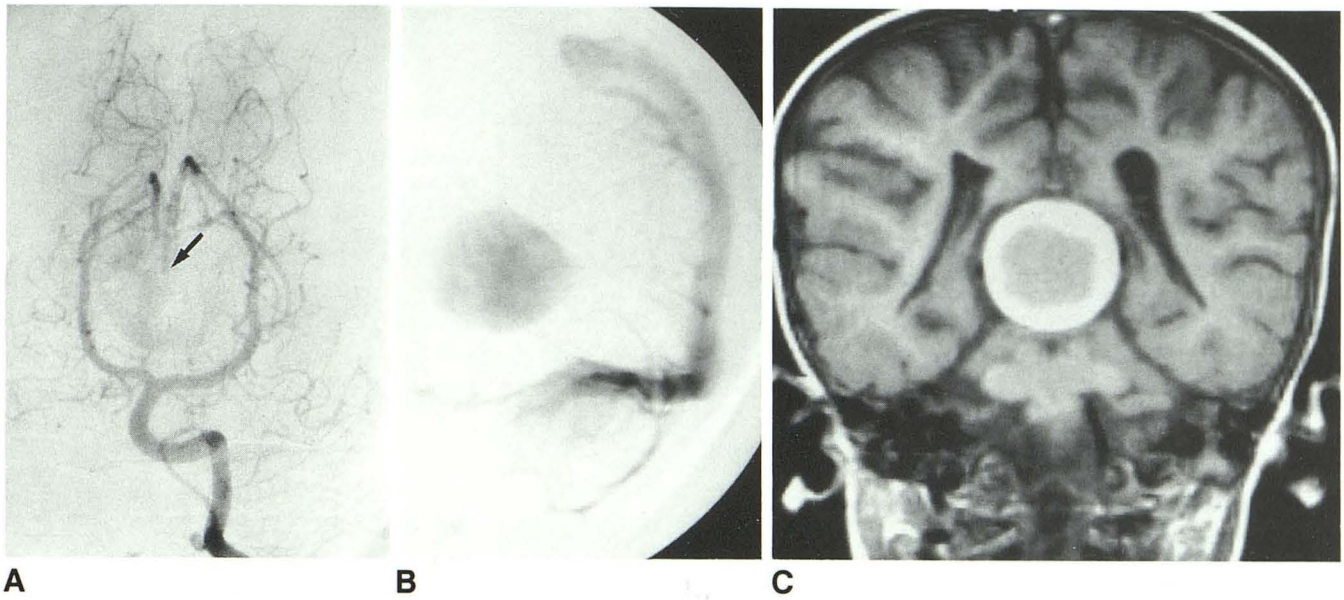

Fig. 1. Case 1 of spontaneous thrombosis of mural type AVG.

A and *B*, Left vertebral arteriograms, anteroposterior (*A*) and lateral (*B*) projections, at age 9 months. Midarterial phase (*A*) shows the fistulas from the choroidal branch of both posterior pericallosal arteries (*arrows*). Delayed view (*B*) shows stagnation of contrast in the aneurysmal sac. The falcine sinus is poorly seen.

C, Coronal MR image, 800/17/2, at age 14 months, shows spontaneously thrombosed AVG.

D, Sagittal MR image, 700/10/2, at age 5 years, shows markedly decreased size of AVG.

left medial posterior choroidal artery to the AVG (Fig. 2A). There was stagnation of contrast within the venous sac with outflow restriction (Fig. 2B). There was very poor filling of the falcine sinus, without evidence of pial venous drainage to the venous sac. This confirmed that the aneurysmal sac did not directly connect with the venous system of the brain (7). Based on these findings, no embolization or surgical procedure was done and the child was followed conservatively.

Two months later, at age 6 months, the patient was noted to have an increased head circumference. CT scan revealed hydrocephalus as well as thrombosis of the aneurysmal venous sac. Ventriculoperitoneal shunting was performed at this time. MR at age 7 months showed the thrombosed sac with compression of the aqueduct (Fig. 2C). At age 11 months, the patient was developing normally. MR at this time revealed a decrease in the size of the thrombosed venous sac so that the cerebral aqueduct could be identified (Fig. 2D).

Discussion

In addition to our two cases, 20 other cases of spontaneous thrombosis of AVGs have been reported in the literature (1–3, 9–21). The importance of the angiographic findings prior to spontaneous thrombosis has not been previously emphasized.

Nineteen patients were less than 2 years old at the time of their initial diagnosis. Three patients were over 2 years old (3.5, 4.5, and 45 years) at the time of their initial presentation.

The most common presenting symptom in the patients presenting below the age of 2 years was macrocephaly, which was present in 14/19 (78%) cases. Hydrocephalus associated with AVG can be due to obstructive mechanisms or to decreased cerebrospinal fluid absorption (7). Differentiation of mechanisms in an individual patient can be important since a shunt in the presence of distal venous obstruction may result in enlargement of the AVG. Other presenting symptoms in this age group included venous sinus thrombosis and hematoma, subarachnoid hemorrhage, sei-

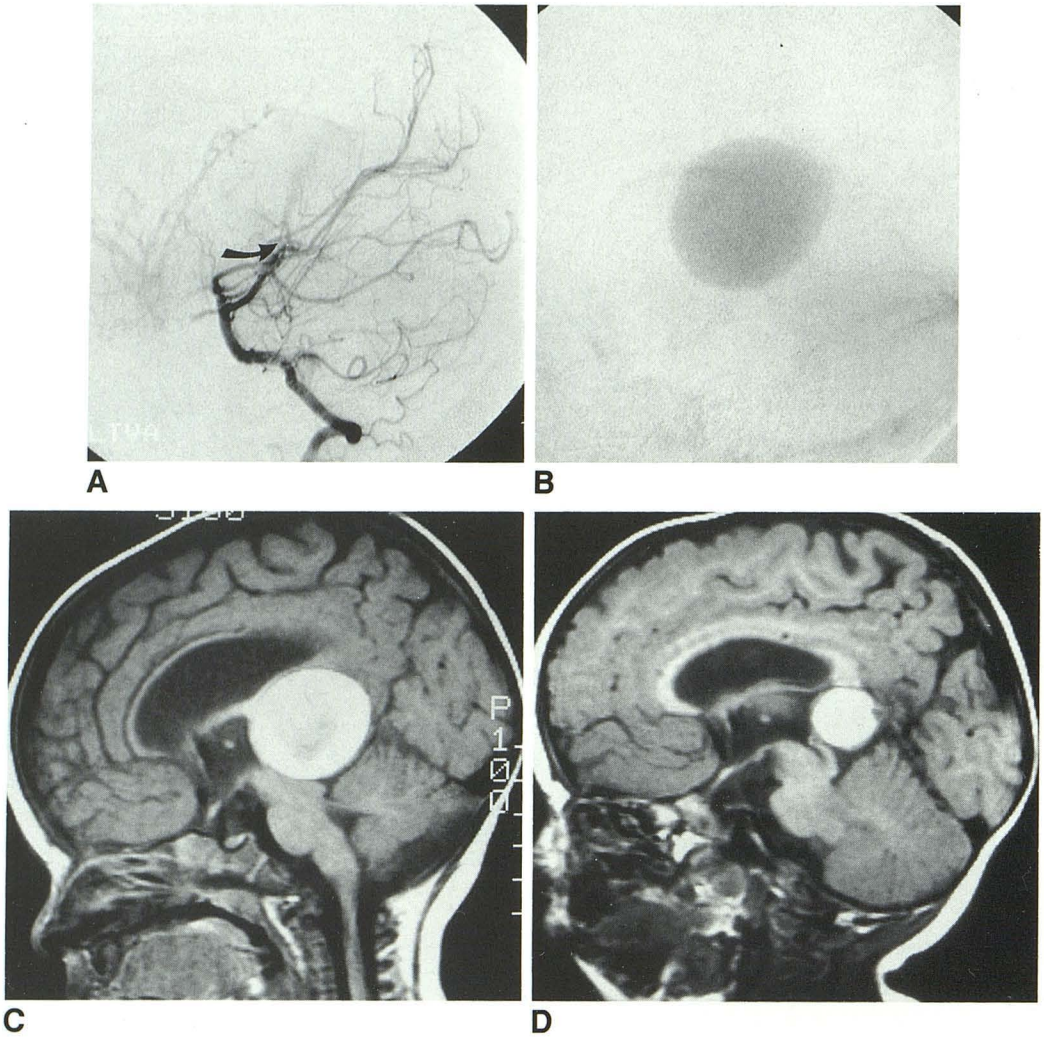

Fig. 2. Case 2 of spontaneous thrombosis of mural type AVG.

A and *B*, Left vertebral arteriogram, lateral projections, at age 4 months. Arterial phase (*A*) shows shunting from medial posterior choroidal artery (*curved arrow*). Later phase (*B*) shows stagnant flow in the aneurysmal sac.

C, Sagittal MR image, 667/20/2, at age 7 months, shows thrombosed sac and compression of the aqueduct.

D, Sagittal MR image, 667/20/2, at age 11 months, shows decreased size of the aneurysmal sac. The cerebral aqueduct can now be identified.

zures, and asymptomatic (diagnosis on prenatal ultrasound).

Patients older than 2 years of age presented with signs of increased intracranial pressure and mass effect. None of the patients reported had congestive heart failure. This would be expected, since low flow shunts in vessels that will spontaneously thrombose should not cause cardiac failure (10).

Six previously reported patients, all younger than 9 months, had undergone angiography prior to thrombosis of the AVG (5, 14, 17, 18, 21). Of these cases, only two (14, 17) give sufficient angiographic detail for comparison with the current cases. In both, stagnation of flow into the venous phase is identified with some degree of outflow restriction involving the venous sac. One was noted to have a "low flow shunt."

In addition to a case of spontaneous thrombosis of an AVG, Shirkoda et al (21) describe a 3-month-old patient who presented with microcephaly without cardiac failure. Angiography showed stasis of contrast in the sac, with obstruction of the sac and no demonstrable drainage of the sac. This was felt to represent "probable beginning thrombosis of the outflow tract and possibly of the aneurysm itself." This patient went to surgery where the feeding vessels were clipped. The aneurysm shrank and the patient did well.

In our two cases, the AVG was supplied by

direct fistulae from the choroidal vessels into the wall of the venous sac. Very slow flow into the venous sac was present with stagnation of contrast into the late venous phase. There was no normal pial venous drainage into the AVG. The venous drainage of the brain did not opacify the deep venous system and, instead, drained towards the cavernous sinus without evidence of venous congestion. In both cases, very proximal, severe outflow obstruction in an embryonic falcine sinus resulted in a large venous sac that produced obstructive hydrocephalus. In our second case, this mass effect became clinically evident only after thrombosis.

The presence of venous outflow obstruction may arise from either retention of fetal anatomical features or from acquired occlusion of dural sinuses (21). These angiographic features of AVGs have been correlated with the clinical presentation (22). Severe outflow obstruction may convert even a relatively large direct fistula into a slow flow shunt (10).

The common denominator in all four cases is the finding of stagnation of flow within the venous sac with associated outflow restriction. The absence of clinical cardiac failure reflects the low flow through the lesion.

Both of our patients had a good clinical outcome as did the previously reported patients with angiographic data.

Our second patient developed hydrocephalus when the AVG was thrombosed. As the outflow of the AVG becomes occluded, there could have been further expansion of the sac. Alternatively, the sac could have swelled after thrombosis. Although the potential for late development of hydrocephalus illustrates the need for careful follow-up of these patients, the outcomes suggest cautious optimism in the face of the described clinical and angiographic features.

The decreasing size of the sac observed on follow-up MR studies in our cases is of interest. Perhaps the need for a ventriculoperitoneal shunt could have been avoided with longer follow-up. The sac had "vanished" in one report (9) while it presented as a 6- to 7-cm calcified mass in another (3). The reason for this is unclear.

If other AVGs without the angiographic features of stagnant flow and outflow restriction could be altered by surgery or endovascular procedures to produce these effects, spontaneous thrombosis might be the expected result. This is an important consideration when performing an endovascular procedure. If obliteration of all feeding vessels is not technically feasible, thrombosis of the AVG can result if the flow in the sac is made stagnant (7).

Ultrasound and color Doppler ultrasound have been used to diagnose AVGs and monitor therapeutic procedures (23–25). If the flow in the sac is shown to be stagnant by color Doppler or MR techniques, conservative management may be indicated without the need for conventional angiography. Further correlations of angiographic studies, ultrasound, magnetic resonance angiography, and clinical outcome are needed. At this time, conventional angiography remains the best method to obtain anatomic and hemodynamic information for the proper diagnosis and management of these lesions.

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