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Case Report

Spontaneous Thrombosis of Type II Vein of Galen Aneurysmal Malformation: a Case Report

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Abstract

Vein of Galen malformations (VGAMs) are rare and complex congenital brain vascular anomalies that pose significant diagnostic and treatment challenges. The natural history of this type of vascular anomaly is very poor, with many patients succumbing to complications such as congestive heart failure, hydrocephalus, and brain parenchymal injury. Although the clinical course of most VGAMs was considered unfortunate, with meticulous imaging, a group of lesions with a more placid presentation and course can be identified.

We present a case of spontaneous thrombosis of VGAM where no embolization or surgical repair was attempted, with excellent clinical outcomes.

This case also highlights the possibility of spontaneous thrombosis in VGAM, even in the absence of clinical symptoms, and emphasizes the importance of a regular imaging follow-up in patients with known vascular malformations.

Keywords

pediatric, thrombosis, vascular, vein of Galen malformation

INTRODUCTION

Vein of Galen aneurysmal malformations (VGAMs) are a rare type of congenital vascular pathologies that represent around 30% of pediatric vascular malformations and around 1% of all cerebral vascular malformations.[1,2] The first reported pathology case is widely thought to have been published by Jaeger et al. in 1937.[3,4] Since then, many more cases have been reported, and interest in the disorder, as well as our understanding of its underlying mechanism, has expanded.[5-7] Two classifications are commonly used in VGAM evaluation: one proposed by Lasjaunias et al.[8] and the other proposed by Yasargil.[9,10] The former categorizes VGAMs into choroidal and mural types based on the origin and insertion of the feeding arteries and the place of fistula communication (direct or intramural). They observe the difference in the clinical presentation and perspective based on their proposed subtypes. The classification proposed by Yasargil makes a more precise division into four types based on the feeding arteries: type I is an arteriovenous fistula between the posterior cerebral arteries or the pericallosal arteries and the vein of Galen, type II is an arteriovenous fistula between the thalamoperforating arteries and the vein of Galen, type III is a mix between types I and II. Type IV is reserved for arteriovenous malformations that drain into the vein of Galen and directly dilate it. Although the clinical course of most VGAMs was considered unfortunate, with better imaging, a group of lesions with a more benign presentation and course has been identified. We present a case of spontaneous thrombosis of VGAM

where no embolization or surgical repair were attempted, with excellent clinical outcomes.

CASE PRESENTATION

A 1-month-old child was referred to our hospital for cranial magnetic resonance imaging (MRI) after the routine post-birth ultrasound suspected a VGAM. Brain MRI and 3D time of flight (TOF) magnetic resonance angiography confirmed the diagnosis ***(Fig. 1)****. The child underwent further neurological and cardiological examinations, which appeared normal without any deviations. The laboratory studies were also normal, and the patient's Bicêtre score was calculated to be 21* ***(Table 1)****. Following a discussion with a multidisciplinary team of neurologists, interventional radiologists, pediatricians, and neurosurgeons, a decision for observation, conservative treatment, and close follow-up was made. Parents were carefully instructed, and monthly control medical examinations were evaluated at six monthly intervals for radiology scans.*

At one year of age, the patient came to the hospital for a routine follow-up examination. The neurological examination was normal, with a slightly higher cranium diameter. A non-contrast computed tomography (CT) confirmed the macrocephaly with a more significant biparietal index than the ages. CT angiography showed an enlargement of the vein of Galen. Due to the mass effect of the malformation, we observed dilatation of the two lateral and third ventricles. There were no changes in the brain parenchyma. On the CT angiography, we could evaluate the feeders coming from the posterior and middle choroidal arteries draining into the large aneurysmal dilatated vein of Galen, dilatated torcula and significantly narrowed proximal part of the straight sinus distal to Galen. We classified the malformation as type II by Yasargil ***(Fig. 2)****.*

The patient was scheduled for digital subtraction angiography, but the parents refused to perform the test. The cardiological consultation of the child was unremarkable. After a neurological and neurosurgical examination, we assessed that the hydrocephalus was asymptomatic and placement of a ventricular-peritoneal shunt was not needed. The patient was discharged and followed closely for the next two years with periodic head circumference measurements. During this period, his neurological examination and progress were excellent.

An MRI at three years of age revealed significant but also positive changes. There was a total occlusion of the malformation due to thrombosis of the aneurysmal dilatated vein of Galen ***(Fig. 3)****. We observed an almost double shrinkage of the varix compared to the CTA made two years ago. There were no radiological signs of hydrocephalus, and the sella media index was normal. The MRI, too, showed no changes in the brain parenchyma. The patient was neurologically intact.*

Another control MRI at eight years was performed. The thrombosed aneurysmal dilatated vein of Galen continues to shrink ***(Fig. 4)****. The venous structure appeared normal, with no radiology signs of hydrocephalus. The neurological examination remained normal. The child exhibited advanced developmental milestones compared to his peers.*

DISCUSSION

The principal pathoanatomic substrate of VGAM is represented by a formed fistula between different portions of the involuting primitive median prosencephalic vein of Markowski and adjacent brain arteries of the fetus around weeks 6-11 of gestation.[10] This, in turn, halts the regression of the venous vessel due to the high blood flow that passes through it, with the malformation not rarely being associated with dural sinus stenosis due to atresia or thrombosis, further increasing the risk of complications.[8,11] An interesting observation can be made that VGAMS can be associated with a different primitive vessel noted on angiograms of patients suffering from the condition - the falcine sinus, which most likely acts as a draining shunt and is connected to the distal 1/3 of the superior sagittal sinus.[12,13] Another noteworthy point is the disruption of the function of the deep venous brain system, which drains through mesencephalic collaterals.[14]

Pathological changes associated with VGAMs can lead to several dreadful patient outcomes. The most common clinical presentation is widely reported to be congestive heart failure due to high output.[15] Another clinical manifestation is development of hydrocephalus due to either the increased intracranial blood volume or cerebral hemorrhages and direct obstruction of cerebrospinal spaces.[16] Due to the condition primarily affecting children, arrest in brain development and focal neurological symptoms can also be observed.[17]

Large strides in the treatment of VGAMs have been made. To this day, they present a clinical conundrum that requires careful consideration of the patient's characteristics and the vascular pathology. The pretreatment era of VGAMs historically cites exceptionally high mortality rates in patients. With the progression of surgical and endovascular methods, a decline in mortality can be observed[18], even with their inherent risks.

An approach for assessing newborns with VGAM involves the application of the Bicêtre score, which aids in determining potential courses of treatment.[19] This 21-point scale assigns points based on the severity of indications and symptoms related to cardiac, pulmonary, neurological, hepatic, and renal systems. The Bicêtre score is computed for neonates with VGAM by employing clinical and laboratory data. A score of less than 8 out of 21 indicates a highly critical prognosis, rendering the infant unsuitable for immediate embolization. For neonates scoring between 8 and 12, emergent embolization is likely to be beneficial. A score exceeding 12 identifies infants eligible for the administration of medical treatment to address their cardiopulmonary insufficiency. Such medical intervention is continued until the infants reach approximately 5 months of age, at which point their larger size diminishes the risks associated with

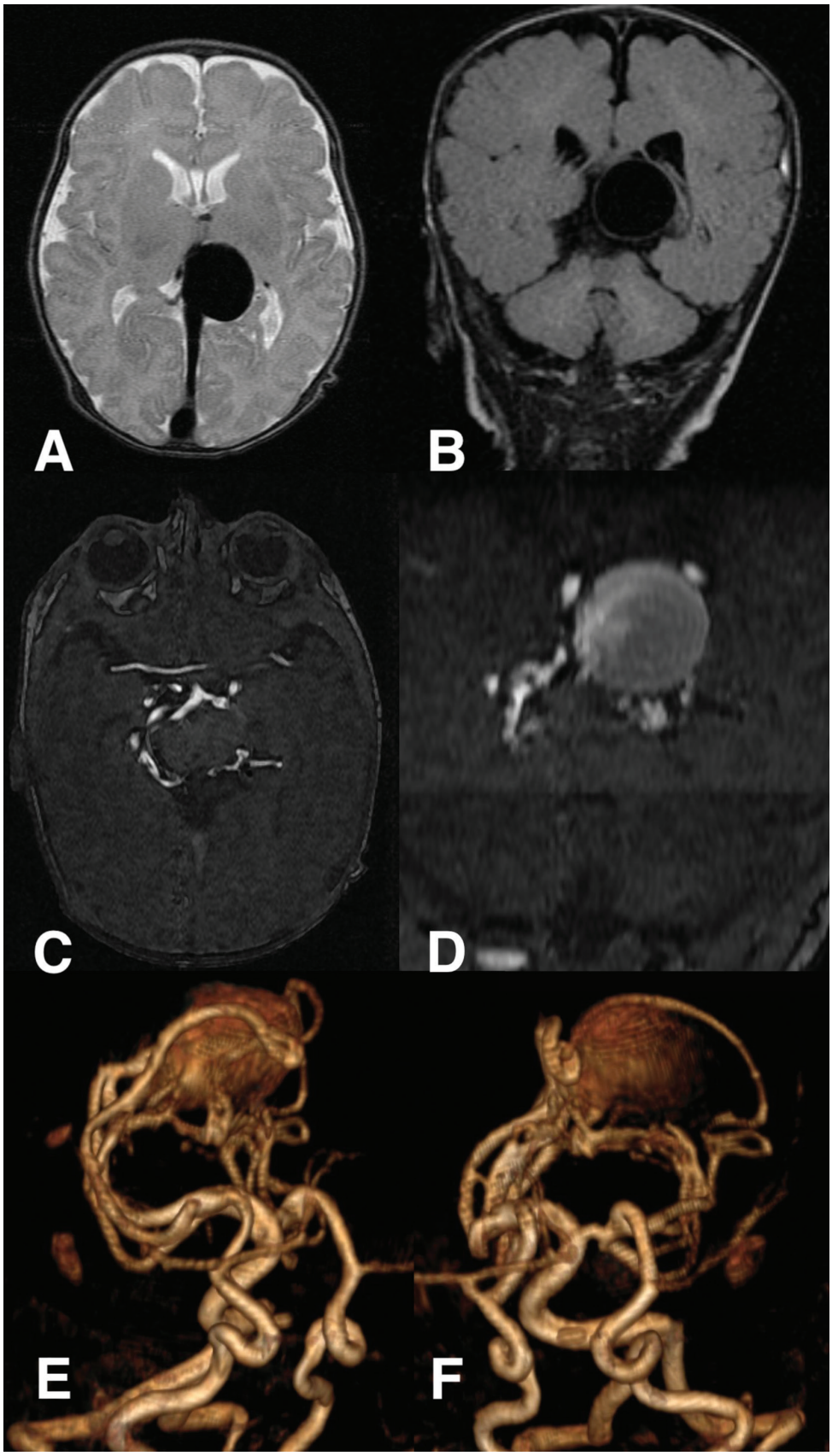

Figure 1. MRI at the age of 1 month. Axial T2 weighted and coronal FLAIR images of the brain show dilatation of the vein of the Galen up to 3 cm in diameter without any pathological findings in the brain parenchyma and ventricular system (**A, B**). 3D TOF images with VR reconstructions demonstrate the feeders coming from the posterior and middle choroidal arteries draining into the large aneurysmal dilatated vein of Galen (**C**, **D**, **E**, **F**).

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Table 1. Bicêtre score

Points	Cardiac functions	Cerebral function	Respiratory function	Hepatic function	Renal function
5	Normal	Normal	Normal	-	-
4	Overload, no medical treatment	Subclinical, isolated EEG abnormalities	Tachypnea finishes bottle	-	-
3	Failure, stable with medical treatment	Nonconvulsive intermittent neurologic signs	Tachypnea does not finish bottle	No hepatomegaly, normal hepatic function	Normal
2	Failure, not stable with medical treatment	Isolated convulsion	Assisted ventilation, normal saturation FiO_2 <25%	Hepatomegaly, normal hepatic function	Transient anuria
1	Ventilation necessary	Seizures	Assisted ventilation, normal saturation FiO_2 >25%	Moderate or transient hepatic insufficiency	Unstable diuresis with treatment
0	Resistant to medical therapy	Permanent neurological signs	Assisted ventilation, desaturation	Abnormal coagulation, elevated enzymes	Anuria

Maximal score: 5 (cardiac) + 5 (cerebral) + 5 (respiratory) + 3 (hepatic) + 3 (renal) = 21

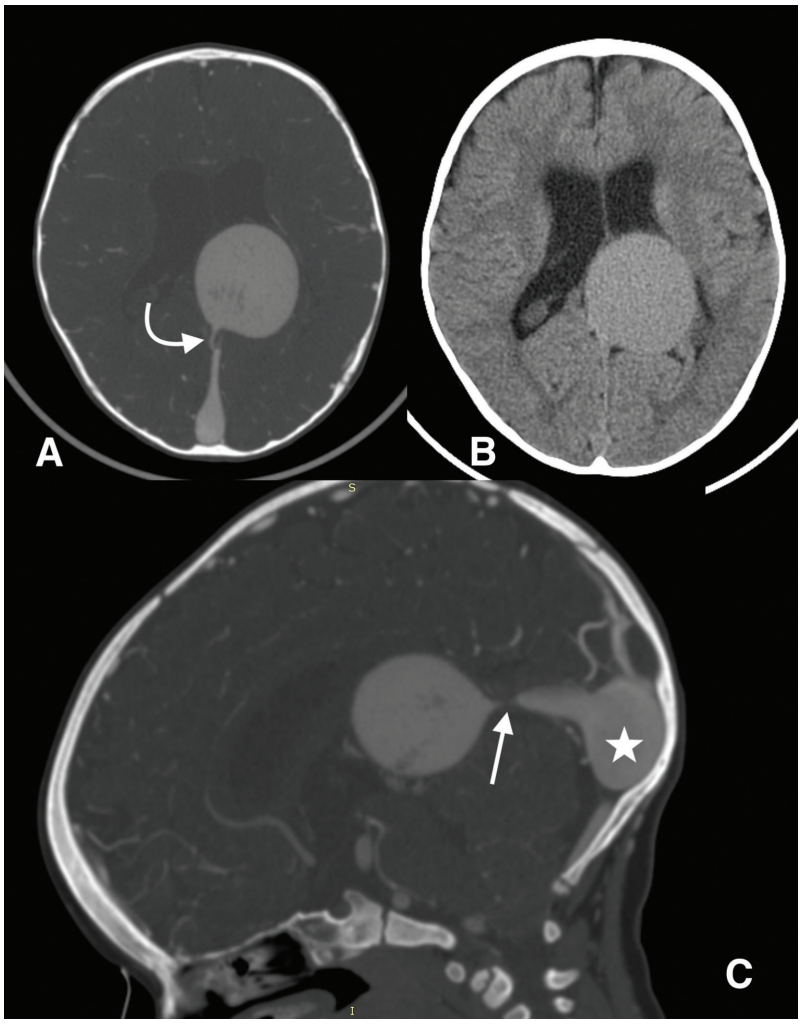

Figure 2. CT image at 1 year. Non-contrast CT demonstrated macrocephaly with elevated biparietal index and dilatated lateral ventricles (**B**). CT angiography shows significant enlargement of the aneurysmal dilatated vein of Galen compared to previous MRI, feeders coming from the posterior and middle choroidal arteries draining into it, dilatated torcula (white star) and significantly narrowed proximal part of the straight sinus (white arrows) distal to Galen (**A**, **C**).

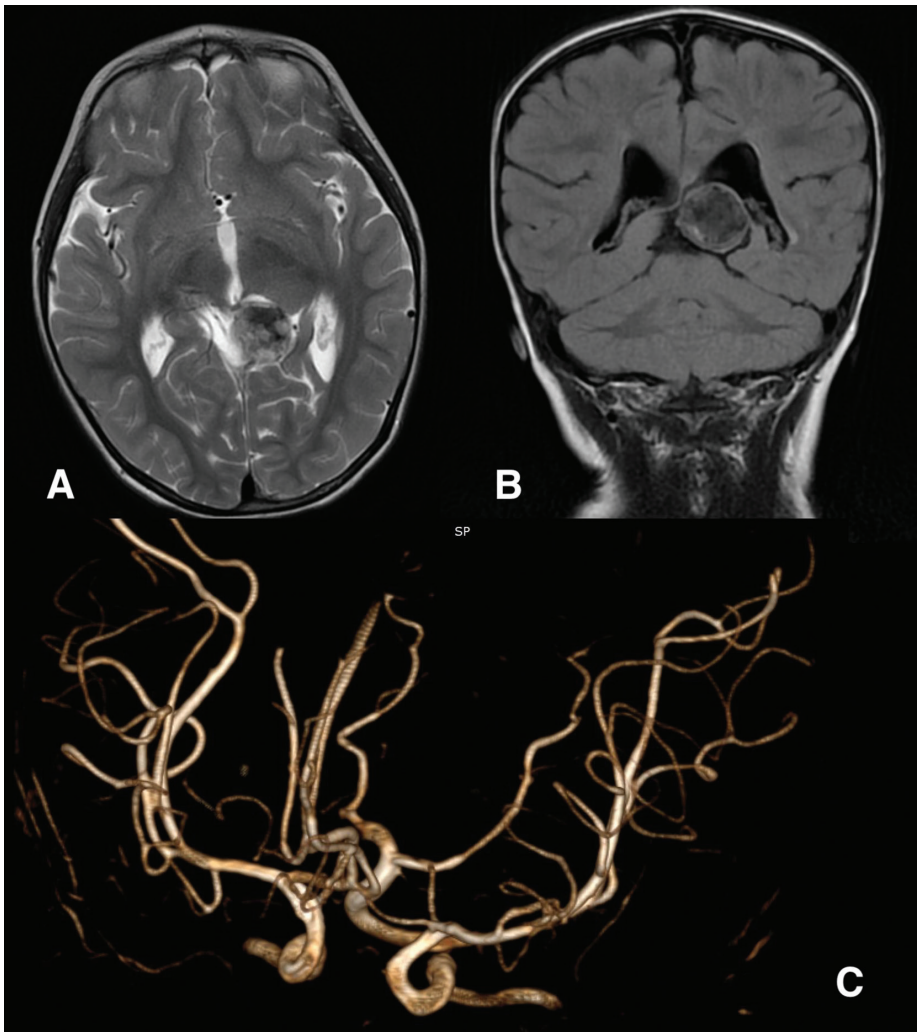

Figure 3. MRI at 3 years. Axial T2 weighted and coronal FLAIR images demonstrate total thrombosis of the malformation, double shrinkage of the varix, no signs of hydrocephalus and damage of the brain parenchyma (**A**, **B**). VR reconstruction of the 3D TOF MRI reveals a complete regression and disappearance of the pathological arterio-venous connections (**C**).

prolonged embolization. **Table 1** provides a comprehensive overview of this treatment protocol.

In one of the largest studies (317 patients) reported thus far, Lasjaunias et al. reported that approximately 19% (57 of 300; 17 lost to follow-up) were considered to be in too bad a clinical condition to be treated. Of the 216 treated patients, 73 (34%) had a bad outcome, ranging from permanent neurological symptoms and mental retardation to death. According to these numbers, the overall poor outcome is approximately 46%. Spontaneous thrombosis of the shunt occurred in only eight of 317 patients (2.5%).[8] In the study of Geibprasert et al., the poor outcome rate was 52%, and three of their reported cases (12%) spontaneously thrombosed during the follow-up period.[20]

In a recent study, Brinjikji et al. performed a meta-analysis of endovascularly treated VGAM. They reported all-cause mortality of 14% and an overall good neurological outcome rate of 62%. Their study's overall poor neurological outcome rates were 21%, in which neonates were significantly less likely to have good neurological outcomes than infants (48% versus 77%).[5]

On rare occasions, though, a spontaneous regression of the malformation can be observed. This is most likely the result of low-flow fistulas, which cause a slow filling of the dilated vascular channel and a more benign natural history, with several such cases being described in the literature.[21,22] In these cases, the commonest presenting sign was macrocephaly, but symptoms such as hemiparesis, seizures, and pyramidal and cerebellar signs were also observed. Patients presenting with spontaneously thrombosed VGAM could be classified in a discrete clinical group, as proposed by Nikas et al.[23]

Spontaneous thrombosis of intracranial vascular lesions is a well-documented phenomenon observed in conditions such as arteriovenous malformations, arteriovenous fistulas, and aneurysms. The presence of flow-related aneurysms, which can both form and regress, underscores the influence of hemodynamic alterations on thrombosis and

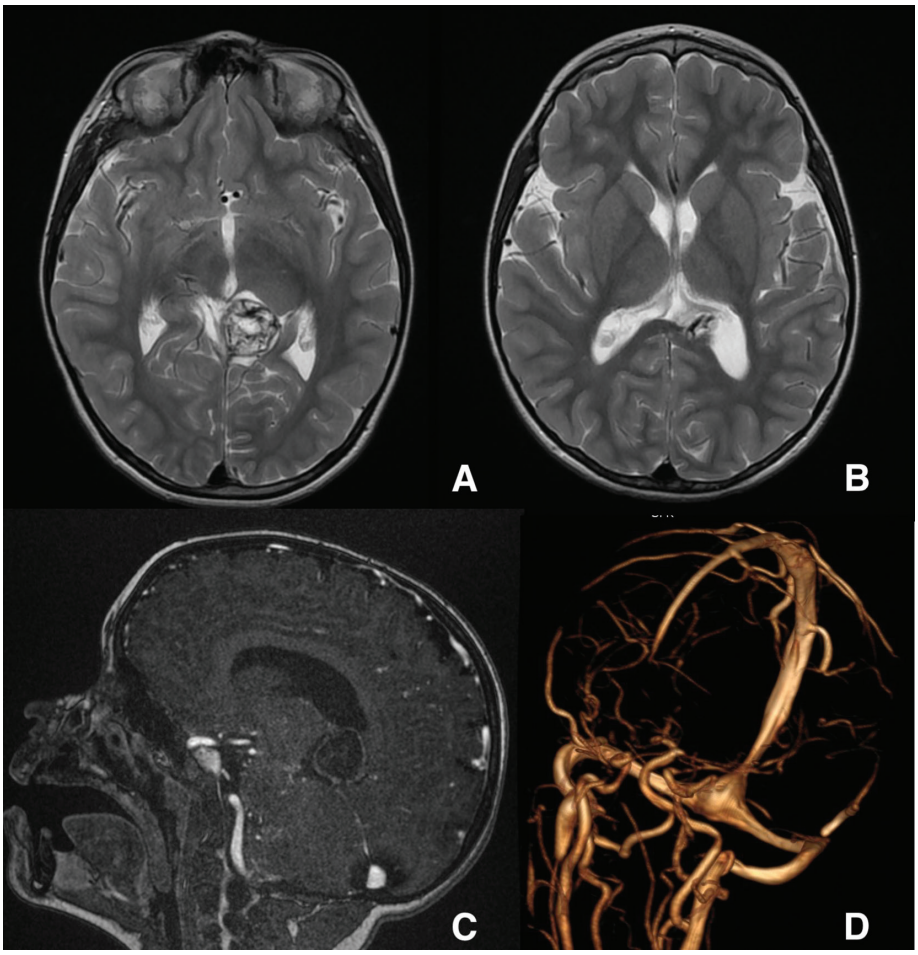

Figure 4. MRI at 8 years. Again, axial T2 and contrast MR angiography demonstrate complete occlusion of the VGAM without any abnormal finding in the brain parenchyma (**A**, **B**, **C**, **D**).

the regression of these vascular abnormalities. While the precise mechanism and clinical factors leading to thrombosis in the vein of Galen malformation are not fully understood, it can be postulated that similar to other vascular lesions, changes in hemodynamic conditions may contribute to thrombus formation. Several potential mechanisms could disrupt the hemodynamic balance in VGAM, including compression or mass effect from adjacent hematoma or intra-aneurysmal clot, posthemorrhagic edema, regressive arteriosclerosis affecting the vessel walls, vascular spasm, and gliosis resulting from fragmentary micro-bleeding.[24]

CONCLUSIONS

The case presented here highlights the possibility of spontaneous thrombosis and subsequent resolution of vein of Galen malformation in a pediatric patient. This case adds to the growing evidence suggesting that certain VGAMs may exhibit a more benign course and favorable clinical outcomes without invasive interventions such as embolization or surgical repair. The utilization of advanced imaging techniques played a crucial role in identifying this subgroup of lesions with spontaneous thrombosis. Further research and long-term follow-up studies are warranted to understand better the underlying mechanisms and factors contributing to spontaneous thrombosis in VGAMs. Nonetheless, our findings emphasize the importance of considering conservative management strategies in selected cases, which may spare patients the potential risks associated with invasive interventions.

Author contributions

S.B.: writing the manuscript; K.S.: methodology, data curation, imaging; S.S.: final and critical review

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Competing Interests

The authors have declared that no competing interests exist.

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Спонтанный тромбоз аневризматической мальформации вены Галена II типа: описание случая

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Резюме

Мальформации вены Галена (VGAMs) представляют собой редкие и сложные врожденные аномалии сосудов головного мозга, которые создают серьёзные проблемы в диагностике и лечении. Естественное течение этого типа сосудистых аномалий очень плохое, многие пациенты страдают от таких осложнений, как застойная сердечная недостаточность, гидроцефалия и повреждение паренхимы головного мозга. Хотя клиническое течение большинства VGAMs считалось неудачным, при тщательной визуализации можно идентифицировать группу поражений с более спокойной картиной и течением.

Мы представляем случай спонтанного тромбоза VGAM, при котором не было предпринято никаких попыток эмболизации или хирургического лечения, с отличными клиническими результатами.

Этот случай также подчёркивает возможность спонтанного тромбоза при VGAM, даже при отсутствии клинических симптомов, и подчёркивает важность регулярного наблюдения за пациентами с установленными сосудистыми мальформациями.

Ключевые слова

педиатрический, тромбоз, сосудистый, мальформация вены Галена