

Vein of Galen aneurysmal malformation in a neonate complicated by disseminated intravascular coagulation: A case report

Abstract

Vein of Galen Aneurysmal Malformations (VGAM) are complex arteriovenous malformations of the pediatric age group characterized by shunting of blood from the arterial to the venous system resulting in high-output cardiac failure, hydrocephalus, PHTN and irreversible neurological damage if not detected and treated promptly. Here, we report the case of a true choroidal type VGAM detected antenatally that was complicated by disseminated intravascular coagulation. We will be discussing the diagnostic approach, the in-hospital course and the treatment modalities implied to manage the associated complications along with the treatment plan tailored for our patient.

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Introduction

Vein of Galen Aneurysmal Malformations (VGAM) are rare arteriovenous malformations encountered in the pediatric population. They are characterized by shunting of an arterial flow into a dorsal dilated cerebral vein resulting in congestive heart failure in the neonatal period.¹ This median vein corresponds to a persistent embryonic channel, the median prosencephalic vein of Markowski (MProsV), normally absent at the adult stage.² VGAM occurs between the 6th and 11th week of gestation after the development of the circle of Willis.³ Here we report the case of a neonate delivered by an elective C-section who was prenatally diagnosed with VGAM along with suspected heart failure on prenatal ultrasound. This is the first case report of a VGAM complicated by DIC in Lebanon besides a case of spontaneous regression of a VGAM reported in Hotel Dieu de France university medical center in Beyrouth, Lebanon.⁴ We will be discussing the diagnostic modality used, the characteristic in-hospital course and the management plan implemented in our institution.

Case report

We report the case of a baby boy born preterm at 35 weeks and 4 days gestational age to a G3P2001 mother by c-section. Prenatally, the baby was diagnosed with vein of Galen malformation on morphological ultrasound with suspected cardiac hypertrophy. So baby was transferred to our university hospital medical center for appropriate medical intervention. Upon birth, baby had a low APGAR score of 5 then 4 at one and five minutes respectively requiring positive pressure ventilation and cardiac massage. Physical exam was unremarkable except for a head circumference of 40 cm (above the 95th percentile for his age) and a thrill heard over his anterior fontanel. Patient was stabilized, and a CXR was done at hour 1 of life that was normal with a cardiac index of 0.55. Ultrasound of the heart showed a PDA of 50%, PFO (left to right shunt, gradient 12mmHg) and a PHTN of 50 mmHg. However, at hour 30 of life,

the patient started having denaturation with respiratory distress and a decrease in his urine output so patient got incubated ventilated. Upon deterioration, CXR showed a left white lung. So infection versus cardiac decomposition was on the differential diagnosis. Laboratory workup was done showing a normal CRP and procalcitonin. He was started on meropenem and vancomycin for broad spectrum coverage. On another hand, laboratory workup showed thrombocytopenia with a platelet level of 28000, high INR of 4.6 and high PTT of 89 sec with a very low level of Fibrinogen. Patient was considered to have a disseminated intravascular coagulopathy (DIC) due to his AV malformation. He required platelet transfusion with fresh frozen plasma for 8 days. Urgent ultrasound of the heart revealed dilated cardiomyopathy, severe PHTN of more than 50 mmHg, severe tricuspid regurgitation and pulmonary regurgitation with poor right sided contractility with reversed diastolic flow of the descending aorta. So patient was started on Nitric oxide 0.2L/min and furosemide 0.5 mg q8h. Once stable, neurological workup was done. Brain MRA/MRV showed sub acute ischemic lesions in the left frontal lobe and insular and to a lesser extent, the right precentralgyrus and insula. Subependymal haemorrhage in the walls of the lateral ventricles.

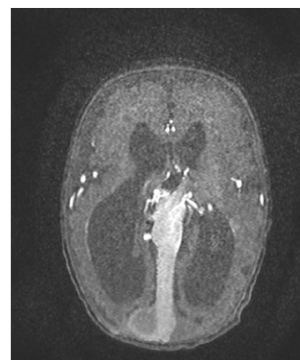


Figure 1A



Figure 1B

A Vein of Galen aneurysmal malformation was noted due to multiple arterial feeders including the thalamoperforating, choroidal and pericallosal arteries converging at its anterior aspect with subsequent obstructive hydrocephalus due to posterior compression of the third ventricle and vestibular aqueduct (Figure 1A) (Figure 1B). Head circumference was followed up on a daily basis. It wasn't increasing, on the contrary over the weeks, the head circumference was decreasing. VP shunt was not done since the hydrocephalus is stable and due to the high surgical risk. Repeated echocardiography at day 6 of life showed dilated cardiomyopathy, persistence of the poor right contractility and tricuspid regurgitation but a decrease in the pulmonary hypertension to 40 mmHg. So NO was stopped, spiro lactone was started and furosemide was tapered to 0.5mg 12h. Patient was improving; however his bilirubin level was on the rise (indirect hyperbilirubinemia). Therefore, an abdominal ultrasound with Doppler to look for other malformations was ordered. It revealed no hepatomegaly, no abdominal aortic aneurysm, normal kidneys size with no aneurysms involving the renal vessels.

Discussion

Vein of Galen Aneurysmal Malformations (VGAM) have been described using different classification systems. Lasjaunias described 2 types: The choroidal type and the mural type. Type 1 (choroidal) is the most common and most severe type which involve multiple choroidal feeding arteries the enter the prosencephalic vein of Markowski via tributary veins.⁵ This type is responsible for the high-output cardiac failure in neonates since high flow fistulas drain into the vein with low output resistance.⁶ Type 2 (mural) is associated with lower number of arterial feeders and thus smaller number of fistulas located in the infer lateral wall of the prosencephalic vein. For this reason, the dilation of the median prosencephalic vein is more pronounced but cardiac failure is less likely to develop and is generally less severe.⁵ In our case, the newborn developed signs of cardiac failure and sudden deterioration with ultrasound findings of right sided heart failure confirming the high output cardiac failure and the choroidal type classification that goes with the radiological diagnosis. Yaşargil classification divided these malformations into 4 types and was based on whether the malformation was a true AV fistula (AVF) (types I-III) or without associated AVF and the exact origin of the arterial feeders.

On the Angio MRI the newborn was classified as Yaşargil type 1 which is a pure AVF between the leptomenigeal arteries and the dilated vein of Galen.⁵ In addition to the abovementioned classifications which depict the severity and the outcome of neonates with VGAM, age has also been described as a prognostic marker with patients below 5 months of age having a poor prognosis.⁷ Our patient developed symptoms shortly after birth which adds to the burden of his disease and explains the rapid deterioration of his clinical status that led to his intubation. High-output cardiac failure is one of the main manifestations of VGAM mainly of the choroidal type and is associated with high mortality rates. It develops because of the volume overload in the right heart chambers. This volume overload along with low vascular resistance in the head leads to reduced systemic blood flow and multi organ failure with persistent PHTN of the newborn.⁷ which was the case in our patient who showed severe PHTN and reversed diastolic flow in the descending aorta which is explained by the circulatory 'steal phenomenon' to the vein of Galen. Furosemide and spironolactone were started in order to reduce the preload and nitric oxide was used in an attempt to reduce

the PHTN which improved on repeat cardiac ultrasound. Our patient presented with hydrocephalus on brain MRA which complicates most of the cases of VGAM. This is mostly due to CSF mal absorption rather than direct compressive obstructive hydrocephalus. This CSF mal absorption is thought to be the result of systemic venous hypertension and exacerbated by the presence of PHTN.⁵ During his hospitalization, our patient laboratory results showed markedly elevated PT, PTT with high D-dimer level and low fibrinogen along with anemia and thrombocytopenia suggestive of disseminated intravascular coagulation (DIC) requiring daily fresh frozen plasma (FFPs) and multiple blood and platelets transfusion until stabilization of his coagulopathy. His laboratory findings were also remarkable for indirect hyperbilirubinemia that is explained by the ongoing hemolytic process. AV malformations are known causes of DIC, however, to our knowledge; DIC was not reported previously in VGAM cases. This consumptive coagulopathy can be the result of a localized intravascular coagulopathy (LIC) due to the stagnation of blood in the AV malformation, abnormal endothelium and chronic coagulation and fibrinolysis along with consumption of other coagulation factors in the abnormal dilated vessels.⁸ Inflammatory markers were negative in our reported case, highlighting the importance of ruling out any ongoing infectious process as a differential for DIC. Pharmacologic treatment is a challenge in this case, since anticoagulation therapy poses a risk of further bleeding and anti fibrinolytic agents poses the risk of further thrombosis. Therefore, we opted for a conservative management approach with FFPs alone. Before his discharge, the patient showed normalization of his PT PTT and fibrinogen levels with a progressive decrease in total bilirubin without further need of FFPs.

Management of VGAM should involve a multidisciplinary approach and is based on whether the diagnosis was made during the neonatal period or later in infancy. Suspected cases should undergo a transfontanellar ultrasound; brain MRI, cardiac ultrasound in case of signs of cardiac decomposition and evaluation of renal and liver function. In neonates, the goal is to stabilize the patient hemodynamically and restore cardiac function in order to gain time for vascular maturation.⁹ ideally, percutaneous embolization should be delayed until 5 to 6 months of age.² However, any decomposition should prompt for early intervention. Delay in embolization may pose the risk of persistent hydrocephalus due to impaired CSF hydrodynamic.¹⁰ Management goals are directed towards preserving neurocognitive function as much as possible. Endovascular embolization is the best treatment modality. Trans arterial embolization is often favored over transvenous approach when there are small numbers of arterial feeders.¹¹ Kissing micro catheter technique is a combination approach with a various success rate. Hydrocephalus management by VP shunt or third ventriculostomy has shown to increase morbidity and mortality in VGAM population and should be only considered in case of failure of the hydrocephalus to resolve after endovascular embolization.¹² Our patient showed a good response to medical treatment by diuretics, nitric oxide and FFPs. We opted to discharge the patient once his hemodynamic status has stabilized and embolization was postponed until he is 5 months of age. Neurosurgery team decided to delay VP shunt insertion for the hydrocephalus hoping that it would resolve after endovascular embolization.

Conclusion

Vein of Galen Aneurysmal Malformations (VGAM) are rare but serious AV malformations that should prompt immediate medical intervention whenever diagnosed antenatally. A multidisciplinary

approach should be considered in order to prevent and treat serious complications, mainly high-output cardiac failure, hydrocephalus and irreversible neurological damage. Our case report highlights the importance of detecting red flags during the course of the disease namely disseminated intravascular coagulation (DIC). Workup should be directed towards ruling out infectious etiology along with other causes of this consumptive coagulopathy in order to adequately manage this serious condition. Management plan of VGAM should be individually tailored according to a case-by-case fashion since endovascular embolization largely depends on the anatomical malformation and the course of the disease.

Acknowledgments

None.

Conflict of interest

The authors declared there is no conflict of interest.

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