

Vein of Galen Aneurysmal Malformation revealed by a severe heart failure and pulmonary hypertension in an infant

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Abstract

Neonatal and pediatric heart failure is commonly caused by congenital heart diseases, especially by large left-to-right shunting. It is rarely related to a Vein of Galen Aneurysmal Malformation (VGAM). Diagnosis and management of these cerebrovascular malformations in infancy are still difficult and remain a medical challenge.

We report the case of a 2-month-old infant hospitalized for heart failure. In addition to symptoms relating to heart failure, the child presented in an agitated state with opisthotonic posture. Echocardiography showed dilated right heart cavities with pulmonary hypertension and no heart defect. Brain magnetic resonance imaging scanning confirmed the diagnosis of VGAM, showing multiple cerebral parenchyma damages. The baby was treated with diuretics and an angiotensin-converting-enzyme inhibitor leading to stabilization of her heart failure. Given the gravity of the neurological sequelae, no embolization procedure was planned for this child. She died 5 years later of acute heart failure.

Keywords

Vein of Galen aneurysm, heart failure, pulmonary hypertension, infant.

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Introduction

Vein of Galen Aneurysmal Malformation (VGAM) is a rare congenital vascular malformation. It represents less than 1% of all cerebrovascular malformations [1]. It is specified as a midline intracranial vascular fistula with aneurysmal dilation of the vein of Galen [2]. In infancy, the clinical presentation features include signs related to volume overload, congestive death.

We report a child with a VGAM revealed by severe heart failure and Pulmonary Arterial Hypertension (PAH).

Case report

A 2-month-old female baby was admitted to our department for dyspnea and cyanosis. She was a full-term baby born by normal delivery. Birth and postnatal histories were uneventful. Upon admission, a physical examination showed hypotrophy with a bodyweight of 3,600 g (-2.4 standard deviations [SD]), a length of 55 cm (-0.5 SD) and a head circumference of 38 cm (0 SD). The body temperature was 38.9°C. The child had tachypnea at 55 breaths/min with oxygen saturation in the air at 92%, her heart rate was at 120/min, her blood pressure was at 90/45 mmHg, and she had a mild systolic murmur. The physical examination also revealed hepatomegaly and hepatic jugular reflux. The neurological examination showed an agitated baby with a paroxysmal opisthotonic posture and no bulging anterior fontanel.

Laboratory investigations done to investigate the child's fever had shown no evidence of bacterial infection and the fever was discovered to be related to a viral infection. However, a chest X-ray detected cardiomegaly (cardio-thoracic ratio = 68%). Congenital heart disease was initially suspected and echocardiography was intended. Thereafter cardiac echography revealed enlarged right cardiac cavities with tricuspid regurgitation, normal left cavities with a normal systolic function of the left ventricle. There was evidence of PAH with a high systolic pulmonary arterial pressure of 110 mmHg. No heart defects were detected. An extra-cardiac shunting especially vascular cerebral fistula was strongly suspected since the neurological examination was abnormal. This diagnosis was thereafter confirmed by radiological imaging.

Non-enhanced brain computed tomography showed a midline cystic mass posterior to the third ventricle with supra-tentorial obstructive

hydrocephalus. Urgent brain MRI with contrast administration was performed. It confirmed the suspected diagnosis showing a large vein of Galen aneurysm (**Fig. 1** and **Fig. 2**) with compression of the Sylvius aqueduct and third ventricle anteriorly causing obstructive hydrocephalus. The inferior sagittal sinus was draining into the aneurysm directly

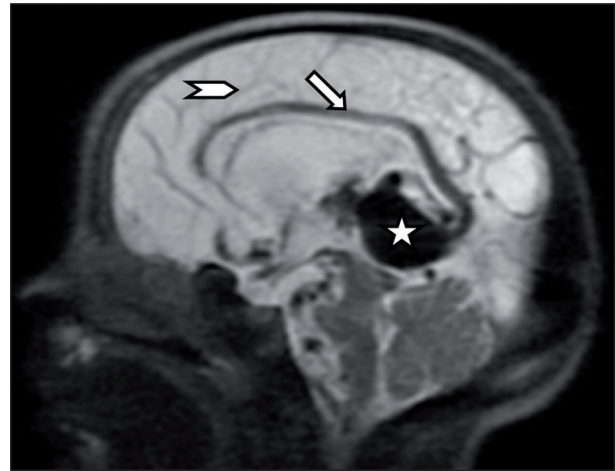


Figure 1. Sagittal T2 weighted image showing a large vein of Galen aneurysm (**star**). The inferior sagittal sinus (**arrow**) is draining directly into the aneurysm. Encephalomalacia (**arrow ribbon**) is also seen appearing as diffuse areas of cystic parenchyma.

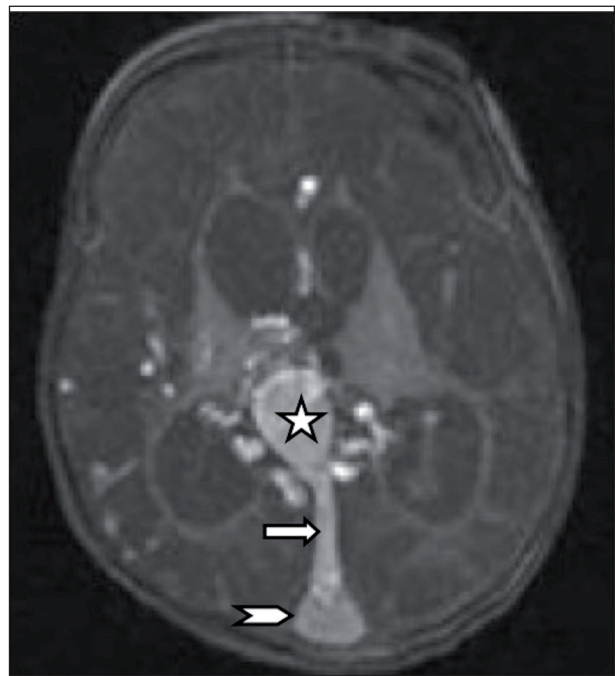


Figure 2. Axial 3D T1 gradient echo post contrast weighted image showing a large vein of Galen aneurysm (**star**) causing compression of the third ventricle with obstructive hydrocephalus. Straight sinus (**arrow**) is draining out the aneurysm associated to dilation of the torcular (**arrow ribbon**). Encephalomalacia is also seen appearing as diffuse areas of cystic parenchyma.

and straight sinus draining out was associated with dilation of the torcular and the superior sagittal sinus. Diffuse supra-tentorial encephalomalacia was also noticed. An electroencephalogram showed a depressed right activity. The baby was treated with diuretics and angiotensin-converting-enzyme inhibitor leading to stabilization of her heart failure associated with valproate. Given the gravity of the neurological sequelae, no embolization procedure was planned for this child and 5 years later she died of a refractory acute heart failure.

Discussion

The VGAM involved an arterio-venous malformation of the diencephalon draining into a dilated vein of Galen [2]. Two major types must be differentiated: true VGAM and Arterio-Venous Malformations (AVM) of the brain with dilatation of the vein of Galen [3]. In the first type, one or more congenital vascular fistulas are connected directly to the wall of the vein of Galen or in the choroid fissure [3]. They are all extracerebral [3]. In the second type (brain AVMs with dilatation of the vein of Galen), there is an AVM that drains into a tributary of the vein of Galen. The dilation of Galen's vein expresses the overload and a secondary or acquired occlusion of the venous outlet, resulting in its overload and dilatation. In this malformation, the AVM is choroidal or parenchyma (supra- or infra-tentorial) in position [3]. It is usually diagnosed at a later age with neurological symptoms such as hydrocephalus, neuro-psychomotor decline, seizures and hemorrhaging [3]. True VGAM manifests early in life (in infancy and the neonatal period) with systemic manifestations and congestive heart failure [2]. The impact of such malformations mainly affects two organs: the brain and the heart. In our case heart failure and neurological disorders were the major evocative signs of VGAM. At the cerebral level, the arterio-venous shunt carries a vascular steal phenomenon hampering the normal vascularisation of some brain areas and leading to cerebral venous hypertension [4]. Hydrocephalus can occur either by decreasing the absorption of the cerebral spinal fluid, a consequence of this cerebral venous hypertension, or by direct mechanical compression of the aneurysm on the cerebral aqueduct [5]. In infants, a macro cranium by hydrocephalus is usually associated. However, hydrocephalus may be, in some cases, associated with microcephaly. In fact, the decrease of the cerebral vascularization induced

by the massive blood shunt can compromise the cerebral growth and result in brain atrophy and microcephaly. In our case, although the head circumference was normal, the patient presented an obvious hydrocephaly associated with a diffuse supra-tentorial encephalomalacia.

At the heart level, the massive left-to-right shunt resulted in an important increase in the cardiac venous return. This hemodynamic load increased the pulmonary vascular resistance and resulted in a PAH [1]. A high-flow vascular shunt was also responsible for heart failure [6]. The severity of heart failure is correlated with the importance of the shunt.

Antenatal consequences of this shunt are mitigated as low placental resistance protect the fetus by a phenomenon of competition with low fistula resistances [7]. These hemodynamic protection mechanisms can sometimes be exceeded and heart failure can be installed even antenatally. To resolve this critical situation, the only solution is to occlude the fistula. The endovascular embolization is often performed in several stages for overload shunting. Different prognostic criteria have been provided to select patients eligible for an embolization procedure. Lasjaunias et al. [8] proposed a neonatal prognostic score assessed on the 8th day of life based on the systemic functions of these children. A score of less than 8 out of 21 leads to the decision not to treat; a score of between 8 and 12 out of 21 entails emergency endovascular treatment; a score of more than 12 out of 21 results in a decision for medical management until the child is at the age of 5 months [9]. At such time, a decision is made to proceed with endovascular treatment no matter what the symptoms are. Embolization at the age of 5 months is associated with the maximum efficacy and the minimum risk of cerebral maturation delay. Lasjaunias et al. report that the presence of severe congestive heart failure with multi-organ failure, seen as a neonatal score of < 8, is a criterion indicating withholding treatment in a patient [8]. This condition relates to our patient. In fact with congestive heart failure, PAH and severe neurological sequelae, the prognosis of the child was estimated to be poor; therefore no therapy had been considered for her.

Conclusion

VGAM is a rare vascular malformation. Its management remains difficult. Embolization is the most appropriate therapeutic procedure for this disease. Heart failure and PAH associated with this

malformation are correlated with a poor prognosis. Therapeutic options are very limited in these situations and associated with a high mortality rate.

Declaration of interest

No Author of this paper has a conflict of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included in this manuscript. Source of funding: unfunded.

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