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Case Report

Vein of Galen aneurysmal malformation presenting as severe heart failure in a neonate [☆]

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ABSTRACT

The vein of Galen aneurysmal malformation (VGAM) is a rare cerebral arteriovenous malformation that can be life-threatening if not diagnosed and treated early. VGAM usually presents in the neonatal period with high-output cardiac failure. We report the case of a full-term male neonate who presented with respiratory distress, and a fontanel bruit soon after birth. A chest radiograph revealed marked cardiomegaly. Transthoracic echocardiography showed dilatation of all four cardiac chambers and a patent ductus arteriosus. Transfontanellar doppler ultrasound and brain computed tomography confirmed the diagnosis of a VGAM. Clinical worsening took place despite aggressive hemodynamic and ventilatory support. The patient's Bicêtre Neonatal Evaluation Score for embolization was 2. Endovascular treatment could not be performed. The patient regrettably passed away. VGAM should be considered in the differential diagnosis of neonatal congestive heart failure with a structurally normal heart. Early diagnosis and treatment improve prognosis considerably.

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Introduction

VGAM is a direct arteriovenous fistula between choroidal arteries and the embryonic median prosencephalic vein (precursor to the vein of Galen) [1]. It accounts for less than 1 % of the abnormalities of the fetus cerebral arterio-venous system [2]. Prenatal ultrasound or MRI diagnosis is possible usually during the third trimester [3]. Neonates (< 1 month) present

with congestive heart failure, infants (1 month- 2 years) and young children (> 2 years) with macrocrania, seizures, hydrocephalus and developmental delay, older children with headaches and subarachnoid hemorrhage [4]. 2 VGAM types are described: mural and choroidal [5]. Cerebral angiography is the gold standard for diagnosis [6]. Embolization is the treatment of choice and improves outcome [6].

We report the case of a full-term male neonate who presented with respiratory distress and a fontanel bruit soon af-

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Fig. 1 – Frontal chest x-ray demonstrating cardiomegaly

ter birth. Transfontanellar Doppler ultrasound and brain computed tomography confirmed the diagnosis of a VGAM. Clinical worsening took place despite aggressive hemodynamic and ventilatory support. Endovascular treatment could not be performed. The patient regrettably passed away.

Case presentation

A male neonate was born to a non-consanguineous couple, at full term, via spontaneous vaginal delivery, weighing 3140 g. The Apgar scores were 4, and 5 at 1, and 5 minutes, respectively. Physical examination noted cyanosis, a hyperdynamic precordium and a fontanel bruit. A chest radiograph revealed marked cardiomegaly (Fig. 1). Transthoracic echocardiography showed dilatation of all four chambers and a patent ductus arteriosus.

Transfontanellar doppler ultrasound demonstrated a sonolucent mass posterior to the third ventricle, with turbulent pulsatile flow within it (Fig. 2).

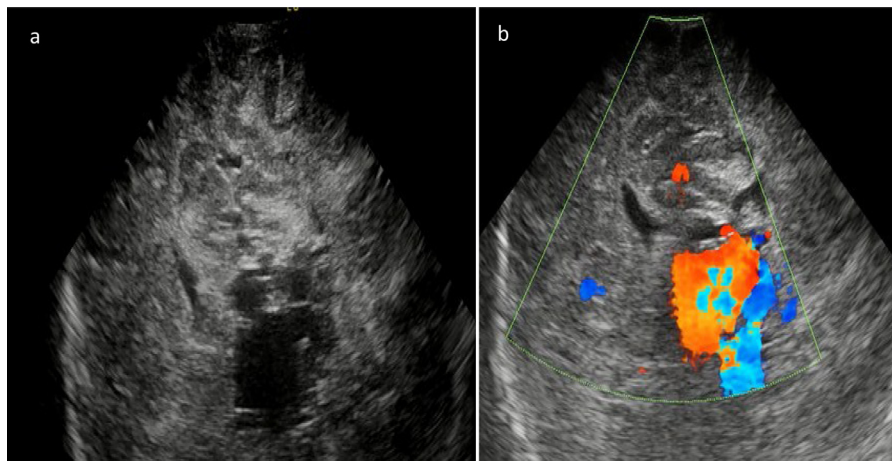


Fig. 2 – Transfontanellar greyscale ultrasound image (A) showing a prominent anechoic, midline structure. The same level doppler examination image (B) on the left demonstrates prominent, turbulent flow within it

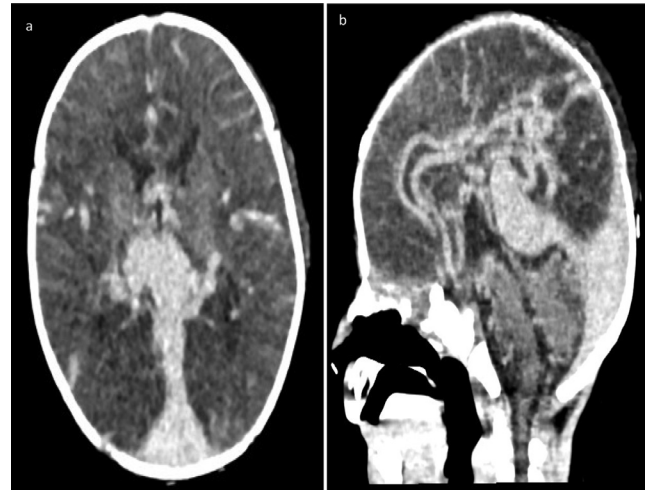


Fig. 3 – Axial (A) and sagittal (B) sections of a brain CT demonstrating marked dilatation of a vessel in the region of the vein of Galen. Drainage is via the straight sinus. The posterior part of the superior sagittal sinus, torcula herophili, the transverse sinuses and sigmoid sinuses are enlarged. Bilateral dilated tortuous tuft of vessels are noted in the quadrigeminal cistern. Diffusely bilateral edematous hemispheres are noted. -> Vein of Galen aneurysmal malformation (VGAM)

Brain computed tomography (Fig. 3) displayed marked dilatation of a vessel in the region of the vein of Galen drained via the straight sinus, enlarged posterior part of the superior sagittal sinus, torcula herophili alongside with the transverse and sigmoid sinuses. Bilateral dilated tortuous cisternal tuft of vessels were present in the quadrigeminal cistern, and diffusely bilateral edematous hemispheres were noted. A vein of Galen aneurysmal malformation (VGAM) was the retained diagnosis.

Laboratory data showed: Hemoglobin 112 g/L, White cell count 8.980×10^9 /L and platelet count 92×10^9 /L. Biochemi-

cal studies showed impaired hepatic and renal functions: (ALT 267 U/L, AST 800 U/L, and serum creatinine 20.1 mg/L), and coagulopathy: prothrombin time 17 seconds.

Despite aggressive hemodynamic support (inotropics-diuretics) and mechanical ventilation, clinical worsening took place. The patient's Bicêtre Neonatal Evaluation Score for embolization was 2. Endovascular treatment could not be performed. The patient regretfully passed away.

Discussion

Vein of Galen aneurysmal malformation (VGAM) is an unusual vascular abnormality, accounting for less than 1% of all the intracranial vascular malformations [7].

It originates from persistent arteriovenous shunting between primitive choroidal arteries and the median prosencephalic vein of Markowski, the embryonic precursor of the vein of Galen, which fails to regress between 6 and 11 gestation weeks [8]. The arteriovenous fistula (AVF) increases the venous system pressure, thus impairing the cerebrospinal fluid (CSF) drainage. CSF accumulates in the ventricles leading to hydrocephalus and in the white matter where congestion causes subependymal atrophy and ventriculomegaly [1]. Mechanical compression of the mesencephalic aqueduct is not the primary cause of hydrocephalus, the aqueduct remaining patent in nearly all patients [9]. Macrocrania results provided that the skull sutures are patent [1].

VGAM has a prevalence of less than 1/25.000 deliveries [8], and a male predominance [2].

Two types of VGAMs are described: The mural type consisting of a direct arteriovenous fistula, with a hole in the persistent median vein receiving arterial inflow, and the choroidal-type VGAM where arteriovenous connection is complex, and small niduses transport arterial flow into the anterior facet of the median vein [1]. Mural VGAM induces heart failure less commonly than choroidal VGAM [1].

Symptoms vary depending on age and VGAM anatomy. In neonates, the choroidal type dominates [2]. Cranial bruit can be auscultated. High output cardiac failure leading to multi-organ dysfunction predominates. In infants, the mural type prevails [2]. Manifestations comprise hydrocephalus, macrocrania, and developmental retardation. Seizures are rare [2]. Subarachnoid and intracerebral hemorrhages can occur due to blood redistribution to pial veins [10].

Prenatal diagnosis of VGAM is feasible by ultrasound or MRI generally throughout the third trimester of pregnancy [3]. Post-natal confirmation relies on transfontanellar ultrasound, computed tomography, MRI, and angiography [11]. Cranial ultrasound is the first imaging modality to consider. It identifies the venous sac as a sonolucent midline mass posterior to the third ventricle. Color and/or pulsed Doppler demonstrate a turbulent pulsatile flow within it, and allow distinction between a VGAM and other midline brain cystic lesions (arachnoid cyst, Dandy Walker malformation...). Hydrocephalus can be observed [12]. Cranial CT with and without contrast, and magnetic resonance imaging confirm the diagnosis and study the adjacent brain parenchyma [13]. Cerebral angiography is the gold standard for the diagnosis [10]. Other imaging tools

to consider are: Chest radiography that may reveal cardiac failure signs, followed by echocardiography that suggests an extra-cardiac shunt in the setting of significant right-sided volume overload, and retrograde diastolic 'aortic runoff' flow, with no intra-cardiac structural lesions [13].

VGAM presents high mortality rates and significant morbidity in survivors [14]. Its negative prognostic factors are: the choroidal type, larger shunts, the presence of cardiac failure in-utero, marked cardiomegaly, tricuspid regurgitation, right atrial enlargement and superior vena cava dilatation, brain injury, and cerebral tissue calcifications [2,12,13].

Endovascular embolization to occlude the shunt is the treatment of choice and is associated with a better outcome [6]. Multiple embolization stages are usually necessary [5]. Endovascular techniques development, patients selection and treatment timing, have considerably bettered patients' prognosis [15]. The primary treatment goal should be amelioration of the patient's physiologic and neurologic status. Neonates' treatment presents a high risk of technical complications and poor neurologic outcomes. When the neonate can be stabilized, postponing the treatment for a few months is beneficial. To select patients for treatment, The Bicêtre neonatal evaluation score (BNES) -a 21-point score that evaluates cardiac, neurologic, respiratory, hepatic, and renal functions- is followed. Endovascular treatment is withheld when there is evidence of brain damage or severe multi-organ failure (BNES<8) due to unavoidable poor outcome. Aggressive medical therapy is given to manage neonates with congestive heart failure. When congestive heart failure's medical management fails, and no severe cerebral parenchymal damage is evident on MRI (BNES 8-12), urgent endovascular treatment is feasible [5]. Once the medical condition is stabilized (BNES 13-21), regular developmental and MRI evaluations (every 3 months for the first year) are advisable. A gradually increasing head circumference suggesting hydrocephalus or early signs of developmental delay indicate endovascular treatment at this point.

Conclusion

Although of rare occurrence, VGAM diagnosis should be considered in neonates with high output cardiac failure and a structurally normal heart. Transfontanellar Doppler ultrasound is suggestive. Cerebral angiography is the gold standard for the diagnosis. Early diagnosis and treatment are crucial as delays result in poor outcome. Patients' selection and treatment timing rely on prognostic factors. When feasible, endovascular embolization is the treatment of choice and is associated with better outcome.

Patient consent

Informed consent was obtained for publication.

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