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Case report

Vein of Galen malformation: prenatal diagnosis, postnatal monitoring and treatment

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Abstract

Vein of Galen aneurysmal malformation (VGAM) is a very rare congenital vascular malformation. Also known as the "median prosencephalic arteriovenous fistula", VGAM is a subtype of dural arteriovenous fistula. It is believed that the development of VGAM occurs between weeks 6 and 11 of embryo development. Prenatal diagnosis is based on fetal ultrasound, between the second and third trimesters. Despite prenatal diagnosis, this congenital malformation is associated with high morbidity and mortality. We report the case of a 38-year-old primigravida referred at 32 weeks of gestational age to our Prenatal Diagnosis Unit for a suspected VGAM. The remaining fetal assessment was normal. At 38 weeks, a male newborn weighing 3,825 g was born. The postnatal evaluation confirmed VGAM. Endovascular treatment was performed at 4 and 5 months, without complications. The patient is currently 9 months old and has a normal neurodevelopment.

Keywords

Vein of Galen aneurysmal malformation, arteriovenous malformation, congestive heart failure, hydrocephalus, prenatal diagnosis, endovascular embolization.

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Introduction

Vein of Galen aneurysmal malformation (VGAM), also named "median prosencephalic arteriovenous fistula", is a rare congenital malformation. It is a subtype of dural arteriovenous fistula, with shunting of arterial blood to an enlarged dilated vein [1, 2].

Prenatal diagnosis is set between the second and third trimesters and is based on fetal ultrasonography.

Fetal magnetic resonance imaging (MRI) plays an important role in diagnosis confirmation, malformation characterization and assessing possible complications [3].

In the presence of VGAM, high-output congestive heart failure is a possible fatal outcome. Therefore, fetal heart assessment is compulsory [4].

Treatment with endovascular embolization is the preferred option, associated with better prognosis [2-5].

We present a case describing the prenatal diagnosis, postnatal monitoring and treatment of a VGAM.

Case description

A 38-year-old primigravida was referred at 32 weeks of gestation to our Prenatal Diagnosis Unit for a suspected VGAM on fetal ultrasound (**Fig. 1**). No other fetal abnormalities were seen. The fetal echocardiogram was normal. Fetal MRI at 33 weeks of gestation confirmed the diagnosis of VGAM. The remaining pregnancy was uneventful. At 38 weeks, a cesarean section was performed electively in a Neonatal Cardiac reference Centre. A male newborn weighing 3,825 g (92nd percentile) was born with an Apgar score of 8 and 9 at 1 and 5 minutes, respectively. At birth, auscultation of the head revealed a cranial bruit. The remaining neonatal evaluation was unremarkable, with head circumference on the 85th percentile. Echocardiogram showed a normal cardiac function. Cranial ultrasound confirmed the prenatal diagnosis of VGAM. Postnatal magnetic resonance angiography (MRA) at 1 month of age showed VGAM and the course of its feeders and drainage (**Fig. 2**). Serial clinical surveillance with particular attention to head circumference and cardiovascular evaluation was performed, and at the age of 1 month his head circumference was above the 97th percentile. MRA was repeated and showed ventricular obstruction with hydrocephalus. Endovascular embolization was performed twice, at 4 and 5 months of age, without complications.

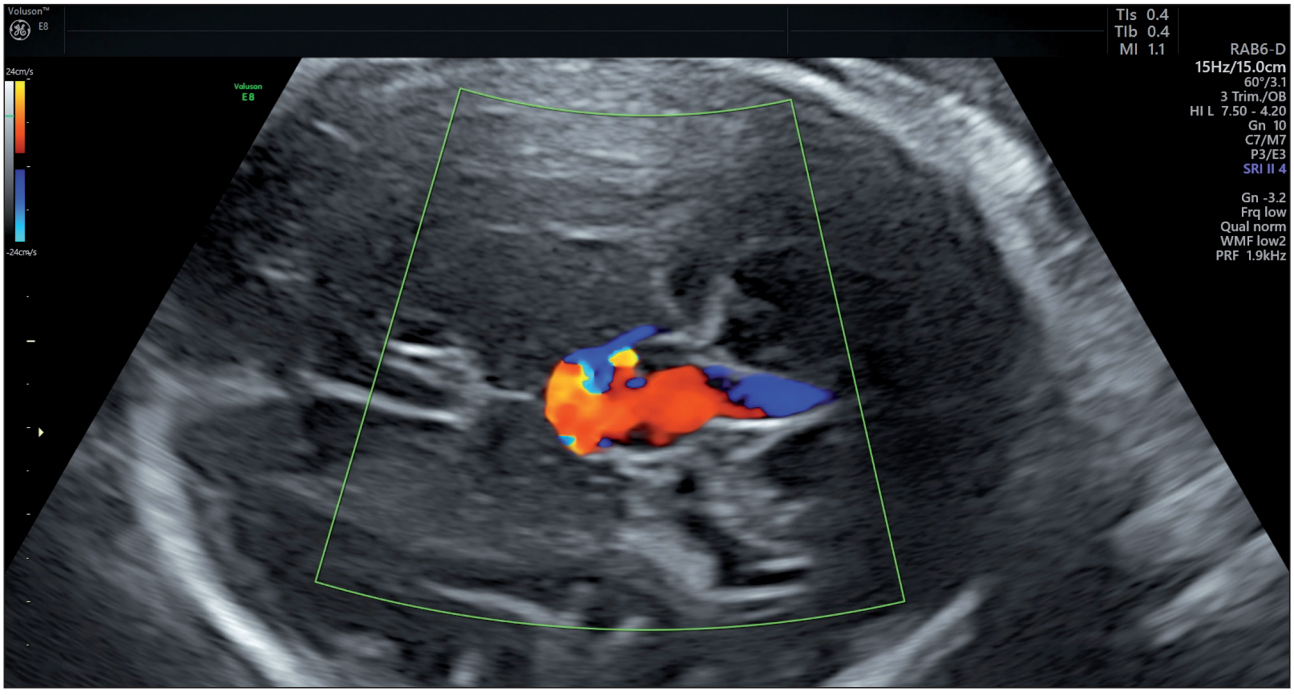

Figure 1. Prenatal ultrasonography at 32 weeks. Axial view of the Vein of Galen aneurysmal malformation (VGAM) with colour Doppler.

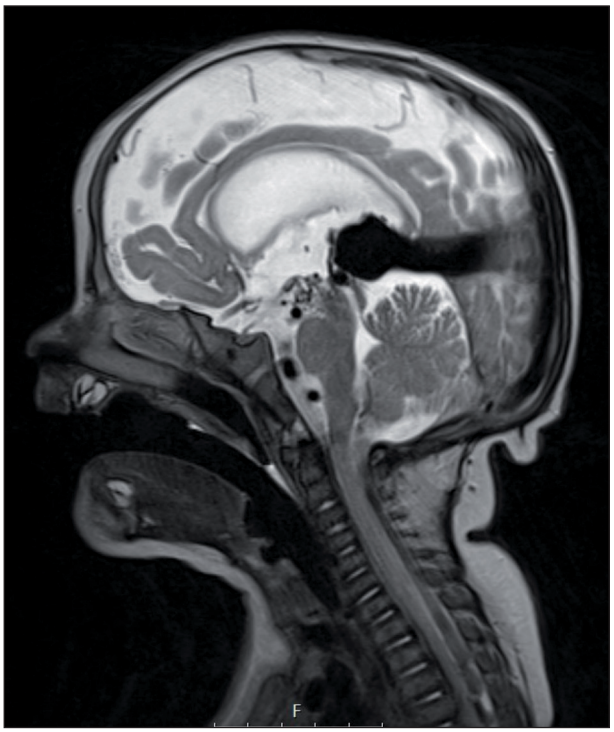

Figure 2. Postnatal magnetic resonance angiography (MRA) at 1 month of age showed Vein of Galen aneurysmal malformation (VGAM).

The child is currently 9 months old and has a normal neurodevelopment. Head circumference is still above the 97th percentile, but stable.

Discussion

VGAM, or median prosencephalic arteriovenous fistula, is a rare congenital vascular malformation, representing 1% of the intracranial malformations [1, 2]. In the first trimester, between weeks 6 and 11, abnormal persistence of the prosencephalic vein of Markowski with aberrant arteriovenous shunts leads to aneurysmal formation of the vein of Galen [4-6]. The majority of VGAMs are diagnosed after birth; however, prenatal diagnosis of VGAM is set between the second and third trimesters and is based on fetal 2-dimensional ultrasonography with colour Doppler. A cystic midline lesion with turbulent flow is identified behind the third ventricle [3]. In the absence of colour Doppler, aneurysmal malformation can be misdiagnosed as a cystic lesion, such as arachnoid cyst, cavum vergae, hydrocephaly and porencephalic cyst [6, 7]. Fetal ultrasound also allows identification of other neurological associated anomalies and assessment of fetal cardiac function (fetal ascites, cardiomegaly, tricuspid insufficiency and jugular vein distention are signs of cardiac failure). Prenatal MRI is helpful for prenatal diagnosis confirmation and characterization of the malformation, identifying the course of its feeding arteries and draining veins. Fetal MRI plays an important role in assessing cardiac and vascular complications. Postnatal confirmation is possible with transfontanellar ultrasound, MRI or angiography [3, 4]. Major complications of VGAN are high-output cardiac failure (as a consequence of increased venous return, contractility, cardiac output and, later, higher oxygen need) and hydrocephalus (due to ventricular obstruction or venous hypertension). Long-term complications include development delay and focal neurological deficits. Endovascular treatment, between 4 and 6 months of age, is the preferred option, associated with better outcomes [2-5].

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Declaration of interest

The Authors declare that there were no conflicts of interest in conducting this work. There were no external funding sources for the realization of this paper.

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