

Case Report

Vein of Galen aneurysmal malformation: Prenatal diagnosis and early endovascular management

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Abstract

A prenatally diagnosed patient was referred to our hospital at 35 weeks of gestation. Antenatal ultrasonography demonstrated cardiomegaly and aneurysm of the vein of Galen. A 3,290 g male baby was delivered by elective cesarean section at 37 weeks of gestation. Physical examination was remarkable for tachypnea, hyperdynamic precordium with a continuous murmur, cranial bruit and bounding carotid pulses. Magnetic resonance angiography confirmed vein of Galen aneurysmal malformation and demonstrated the vessels feeding the aneurysm. Postnatal management included aggressive medical treatment of cardiac failure and transarterial embolization of the vessels feeding the aneurysm at 3 days of age. Posthemorrhagic hydrocephalus developed after embolization, and a ventriculo-peritoneal shunt was placed at 29 days of age. Presently, the patient is 4 years of age and has no neurological abnormality at follow-up. Vein of Galen aneurysmal malformations can cause severe morbidity and mortality in neonates. However, careful obstetric follow-up and early postnatal endovascular treatment of these malformations may lead to a favorable outcome.

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1. Introduction

Vein of Galen aneurysmal malformations (VGAMs) are rare intracranial arteriovenous malformations, which can cause severe morbidity and mortality in neonates. Prenatal diagnosis may be achieved by real-time and Doppler sonography with color flow imaging or magnetic resonance imaging (MRI). Antenatal MRI can show the malformation in three dimensions and depict the exact anatomy of the dilated channels and thrombosis if any.^{1–3}

Patients with VGAM have varying age-related clinical presentations. Fetal manifestations have included nonimmune hydrops, hydrocephalus and intracranial hemorrhage.⁴

The clinical manifestation consists predominantly of high-output cardiac failure in the neonatal period. An elevated cardiac output may be correlated with the magnitude of the cerebral arteriovenous shunt.^{2,5} The low systemic resistance of the fetus *in utero* can decrease the flow through the malformation and minimize cardiac decompensation, but the sudden increase in systemic vascular resistance encountered at the time of delivery will result in a much greater diversion of flow through the malformation.⁶ Evidence of progressive cardiac dysfunction *in utero* is a grave sign, indicating a high-flow lesion that may not respond to therapy.⁷

Management is a major therapeutic problem. Neuroradiological intervention has been shown to control cardiac failure, but there is a perception that neurological outcome in survivors is poor.⁸ Here, we present a male neonate with VGAM diagnosed prenatally and treated with early endovascular

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embolization. At 4 years of age, he is growing normally without neurological deficits or developmental retardation.

2. Case report

A prenatally diagnosed patient with VGAM was referred to our hospital at 35 weeks of gestation. Both pulsed-wave Doppler and color–velocity imaging studies confirmed aneurysm of the vein of Galen and cardiomegaly. Prenatal ultra-fast MRI was used to view the malformation in three dimensions.

A 3,290 g male baby was born at 37 weeks' gestation by elective cesarean section to a 31-year-old woman. Apgar scores were 9/9 at 1 and 5 minutes, respectively. Physical examination was remarkable for tachypnea, hyperdynamic precordium, a grade 2/6 continuous murmur and bounding carotid pulses. A loud continuous bruit was audible all over the cranium. The baby's head circumference was 36.5 cm (>90 p), and cranial sutures were separated.

Chest radiography immediately after birth showed cardiomegaly. An echocardiogram performed at 1 hour of life revealed pulmonary hypertension, tricuspid regurgitation and patent foramen ovale. Head ultrasound scan demonstrated a large, midline venous structure. MRI and magnetic resonance (MR) angiography showed VGAM and the vessels feeding the aneurysm. The rest of the brain parenchyma and ventricles were normal (Fig. 1). Since the baby had signs of cardiac decompensation, in addition to medical treatment of cardiac failure, transarterial embolization was performed at 3 days of age (Fig. 2). Microcoils were placed into several feeding arteries by an arterial approach, resulting in a marked decrease of flow through the malformation. However, the patient had a left focal seizure, signs of severe cardiac failure and hypertension after the embolization. He required



Fig. 2. This picture shows coils that have been placed during transarterial embolization of vein of Galen malformation.

ventilatory support. Cardiac failure was treated with digoxin, dobutamine, and furosemide. Captopril was used to treat hypertension.

Repeat head ultrasound demonstrated right intraventricular hemorrhage at 4 days of age, and posthemorrhagic triventricular hydrocephalus developed consequently. A ventriculo-peritoneal shunt was placed at 29 days of age. After shunting, requirement for medical treatment decreased. The patient was on room air, feeding well, and gaining weight. At 44 days of age, MRI and MR angiography showed that the aneurysmal dilatation of the vein of Galen had markedly thrombosed and the degree of hydrocephalus had decreased (Fig. 1). He was discharged at 65 days of age. At 4 years of age, there is no evidence of cardiac failure, he does not require any cardiac medications, and he has normal head circumference and neurologic development.

3. Discussion

VGAM is known to be associated with various structural cardiac anomalies, of which sinus venosus atrial septal defect and coarction of aorta seem to be the most common.⁹ McElhinney et al.¹⁰ reported partial anomalous pulmonary venous connection, ventricular septal defect and atrioventricular canal defects to be other frequent associations. Our case had pulmonary hypertension, tricuspid regurgitation and patent foramen ovale which were not structural cardiac lesions.

Management is a major therapeutic problem, in spite of the recent advances in radiologic diagnosis, neonatal intensive care, and neurosurgery. The principal approaches to treatment in the newborn involve attempts to eliminate the high flow through the vascular lesions, either by arterial embolization, usually with a liquid adhesive agent or microcoils, or by venous embolization, with placement of metal coils.¹¹

Aggressive medical treatment of cardiac failure and early neurointervention combined with modern neuroanesthetic and neurointensive care results in good survival rates with low morbidity, even in cases of high-risk VGAM presenting in the immediate perinatal period with cardiac failure.¹² If medical management of cardiac failure fails, and there is no evidence of gross cerebral parenchymal damage on imaging, urgent endovascular treatment is feasible and can reduce the almost-100% mortality otherwise expected, without invariably severe morbidity.⁵

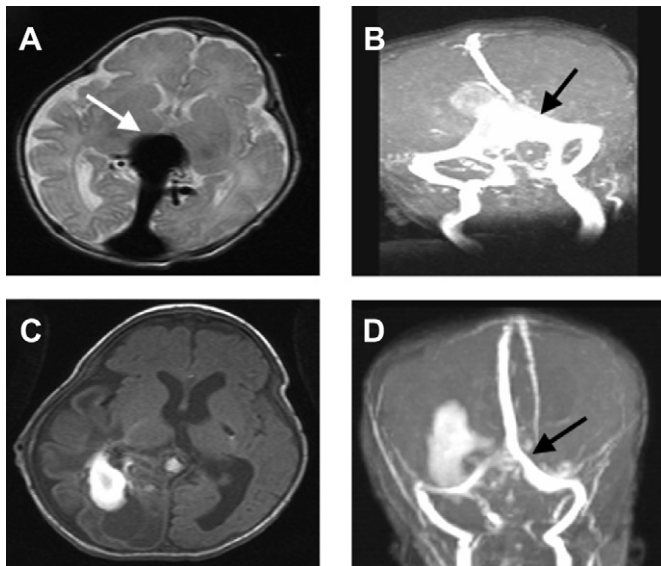


Fig. 1. (A) Magnetic resonance imaging and (B) magnetic resonance angiography show vein of Galen aneurysmal malformation and the vessels of the feeding. (C) Magnetic resonance imaging and (D) magnetic resonance angiography show vein of Galen aneurysmal malformation has decreased in size after embolization.

Table 1
Clinical presentation and outcomes of 15 neonates with vein of Galen aneurysmal malformations

Authors	Time of diagnosis	Symptoms	Management	Outcome
Has et al. ³	Antenatal (34 wk)	CCF	Aggressive treatment of CCF	Death (3 d)
Sasidharan et al. ⁴	Antenatal (33 wk)	CCF	Aggressive treatment of CCF	Death (6 d)
	Postnatal (30 hr)	CCF, HS	Aggressive treatment of CCF	Death (2 mo)
Mitchell et al. ⁵	Postnatal (8 hr)	CCF	Multiple embolization	Normal
	Antenatal (31 wk)	Mild CCF	Multiple embolization	Normal
	Postnatal (2 hr)	CCF	Embolization	Death (2 d)
	Postnatal (36 hr)	CCF	Multiple embolization	Moderate developmental delay
Frawley et al. ¹²	Postnatal (38 hr)	CCF	Multiple embolization	Normal
	Postnatal (38 hr)	CCF	Embolization	Normal
	Antenatal (36 wk)	CCF	Multiple embolization	Death (24 d)
	Antenatal (38 wk)	CCF	Multiple embolization	Normal
	Antenatal (32 wk)	CCF	Multiple embolization	Death (29 d)
Maheshwari et al. ¹⁴	Antenatal (28 wk)	CCF	Embolization	Death (29 hr)
Jones et al. ¹⁵	Antenatal (36 wk)	CCF	Embolization	Death (24 hr)
	Postnatal	Mild CCF	Multiple embolization	Normal

CCF = congestive cardiac failure; HS = hydrocephalus.

Lasjaunias and colleagues¹³ reported that 216 patients were treated with endovascular embolization. Of 216 patients, 23 died despite or because of the embolization (10.6%). Twenty out of the 193 (10.4%) surviving patients were severely retarded, 30 (15.6%) were moderately retarded, and 143 (74%) were neurologically normal on follow-up. Their data demonstrated that most treated children survive and undergo normal neurological development. Our case has no neurological abnormality at 4 years of age.

Our patient underwent MRI with MR angiography as part of his diagnostic evaluation. MRI is mandatory before endovascular treatment, to assess the brain parenchyma. MRI has a prognostic value, allowing the decision for therapeutic approach. MR angiography is mandatory at the time of endovascular treatment. It not only confirms the aneurysm, but also provides details of vascular anatomy and helps to plan the treatment.¹⁴

We observed intraventricular hemorrhage after embolization in our case. In neonates, the bleeding is presumed to arise from immature vessels in the choroid plexus or germinal matrix that cannot tolerate the significant increase in pressure and volume associated with occlusion.¹⁵

Table 1 summarizes the clinical details and outcomes of 15 neonate cases published in the past 10 years. Without interventional neuroradiology and embolization, the prognosis is poor for neonates whose primary presentation is severe cardiac failure secondary to VGAM. When cardiac failure cannot be controlled with medical therapy, early endovascular embolization may allow successful treatment of these neonates.

Prenatal diagnosis and early intervention by transarterial embolization produced a good outcome in our case. He recovered very well without any sequelae. Prenatal diagnosis provides the opportunity to plan the delivery of the baby at a center where immediate and definitive care can be provided. Patients diagnosed with VGAM should be transferred to a tertiary neonatal intensive care unit where a pediatric neurologist, a cardiologist, an interventional neuroradiologist

and a neurosurgeon for management of the infant are available. Timely diagnosis and appropriate interventions of this condition may lead to a favorable outcome.

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