

Vein of Galen Malformation: Prenatal diagnosis to postnatal outcome

Brinda Sabu, Dhanya R Shenoy, Manish Kumar Yadav, KIMSHEALTH Trivandrum, Kerala, India

INTRODUCTION

- Vein of Galen malformation (VOGM) is a rare congenital malformation of the cerebral blood vessels.
- Though they account for less than 1-2% of all intracranial vascular malformations, they constitute up to a third of intracranial vascular malformations among the pediatric age group.
- There are two types of VOGM are described in the literature - Mural and Choroidal type.
- Most babies are symptomatic in the neonatal period
- If left untreated have an almost 100% morbidity and mortality.
- Prenatal diagnosis allows for the delivery of the baby in a tertiary care centre and early intervention which would significantly improve the neurodevelopmental outcome of the baby.
- The introduction of endovascular embolization techniques has paved the way for successful treatment of VOGMs

CASE REPORT

We report a case of 32 yr old second gravida, who was referred to us at 38 weeks for II opinion VOGM. A detailed prenatal ultrasound at our centre revealed a supratentorial midline cystic lesion measuring 2.1 x 2.1 cm superior to the thalamus. Colour flow mapping revealed turbulent flow and spectral Doppler showed high velocity flow- suggestive of Aneurysmal Malformation of Vein of Galen. Sulcal pattern appeared normal for the period of gestation. There was associated bilateral ventriculomegaly, prominent cisterna magna, and dysmorphic Cavum Septum Pellucidum, but no features of hemorrhage, necrosis, porencephaly, schizencephaly or cortical malformations. A fetal echocardiogram was done which revealed cardiomegaly with Cardiothoracic ratio 0.52 and persistent left SVC draining into the dilated coronary sinus. No other fetal anomalies were noted. The growth velocity of Biparietal diameter and head circumference were more than the 99th centile, but rest of the fetal parameters showed normal growth. There was polyhydramnios with an AFI of 25 and Dopplers were normal. The limitations of genetic screening at this gestational age was explained to her and the need for delivery in a tertiary care center with detailed postnatal evaluation and close postnatal followup was advised.

She went into spontaneous onset of labour and delivered a term, male baby of 3.82 kg by vaginal route.

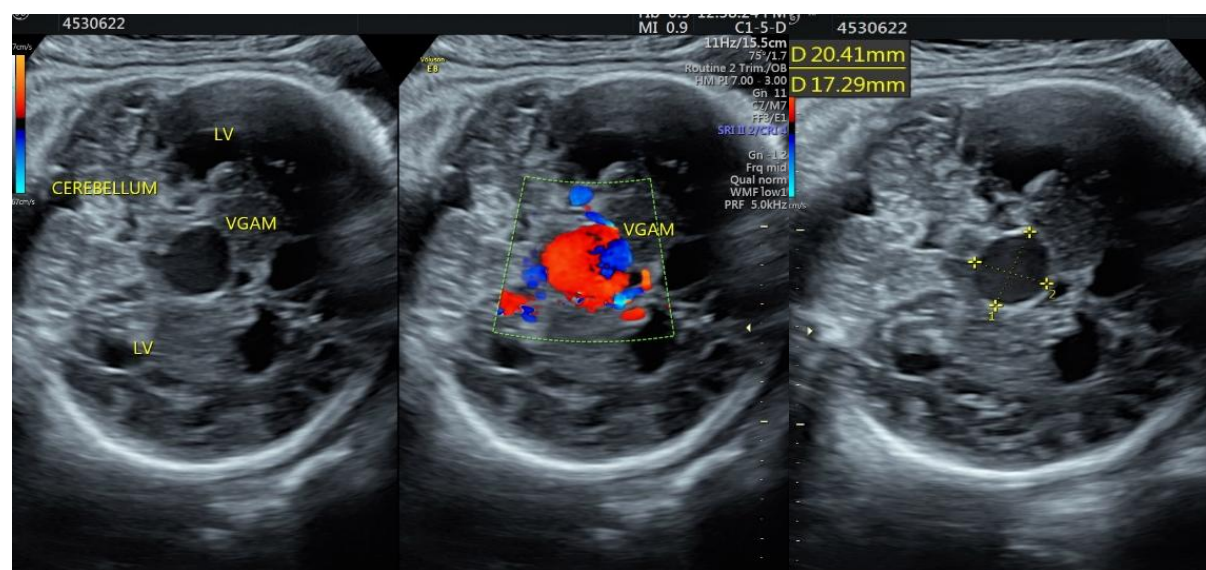
Postnatal cranial ultrasound confirmed the antenatal findings. The baby was also started on acetazolamide from postnatal day 2 to decrease the cerebrospinal fluid formation.

Postnatal MR Angiogram at 1 month showed a mural-type VOGM with stenosis at the junction of the sac and falcine sinus with obstructive moderate hydrocephalus. Serial clinical surveillance and close follow-up

of the baby was done. At 3 months of life, the baby presented with increasing head circumference. MR Angiogram was repeated and showed ventricular obstruction with increasing hydrocephalus. Endovascular embolization was performed with N-butyl 2 cyanoacrylate gel (80 % NBCA) by an Interventional Radiologist. As the postprocedure MRI head showed a significant increase in hydrocephalus, a Ventriculoperitoneal shunt was placed following which the child gradually improved. The child is currently 9 months old and is under close neurological follow-up.

IMAGING

Prenatal Ultrasound



VGAM in Grey Scale (Figure 1) and Color (Figure 2)
Figure 3 shows the size of VGAM



Figure 4 depicts the dilated lateral ventricles; Figure 5 shows the feeder vessel and draining sinus of VGAM; Figure 6 showing blood flow across the VGAM

Postnatal MR angiogram



Axial images of the brain confirming VOGM with moderate hydrocephalus



Sagittal section of VOGM with its feeder vessel and draining sinuses

Postembolisation MRI



Increased hydrocephalus post-embolization.

Hence ventriculoperitoneal shunt was performed on postembolisation day 4

DISCUSSION

- VOGM, also called as Vein of Galen aneurysmal malformation (VGAM), is a type of dural arteriovenous fistula, with shunting of arterial blood to an enlarged dilated vein.
- The vein of Galen which is located under the cerebral hemispheres drains the anterior and central regions of the brain into the posterior cerebral fossa sinuses. At 11 weeks of gestation, the median prosencephalic vein of Markowski usually regresses and its posterior part joins the internal cerebral veins and basal veins to form the vein of Galen. VOGM occurs when the prosencephalic vein does not regress but persists beyond 12 weeks. An arteriovenous shunt between this vein and the arteries supplying the brain induces hemodynamic changes and causes the steal phenomenon which leads to high output cardiac failure.

Prenatal diagnosis

- Though majority of VOGMs are diagnosed after birth, prenatal diagnosis is possible. Though the defect develops in the early 1st trimester, the aneurysm becomes sonographically apparent in the 3rd trimester only.
- In a prenatally diagnosed VOGM, a detailed Neurosonography should be done to rule out other brain abnormalities. A detailed Fetal Echocardiography should also be done to look for cardiomegaly, evidence of cardiac overload, or high output cardiac failure.
- VOGMs are not usually associated with chromosomal abnormalities or genetic syndromes.

Differential diagnosis of cystic midline lesions with turbulent flow identified behind the third ventricle include arachnoid cyst, cavum vergae, hydrocephaly and porencephalic cyst; however, they don't take up Color Doppler unlike VOGMs.

Prenatal Followup & delivery

- Weekly follow-up scans should be done mainly to determine the time of delivery depending on the risk of developing heart failure. Delivery should be in a tertiary care centre. Mode of delivery is suggested to be Caesarean Section and it is usually timed at around 38 weeks.

Postnatal management-

- Most of the neonates with VOGMs become symptomatic within 1 year of life and if left untreated have an almost 100% morbidity and mortality.
- In the immediate postnatal period, babies can show features of respiratory distress, cardiac failure and may require prolonged oxygen support. Major complications of VOGM include high-output cardiac failure (VOGM acting as a steal and causing increased venous return, increased contractility, increased cardiac output and, thus higher oxygen need) and hydrocephalus (due to ventricular obstruction of cerebrospinal fluid flow or venous hypertension).
- Long-term complications include neurodevelopment delay and focal neurological deficits
- Endovascular embolizations have shown great success in treating VOGMs. Endovascular treatment, between 4 to 6 months of age, is the preferred option today. However, congestive cardiac failure in a neonate which is refractory to medical treatment is an indication for an emergency embolization.

CONCLUSION

- Though a rare entity, VOGMs carry very high morbidity and mortality.
- Prenatal diagnosis provides an opportunity to plan the delivery of the fetus at a tertiary care center and thus provide the best of care to the baby through a multidisciplinary team approach.