Case Report: Vein of Galen Aneurysmal Malformation in the Fetus and Neonatal Outcome

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Vein of Galen aneurysmal malformation [VGMA] is a rare and abnormal vascular malformation of the fetal vascular cerebral system. We present a case at 37 weeks of gestation which has been referred to our institution with suspicion of VGAM associated with early fetal cardiac congestive failure. After confirmation of the diagnosis the parents were informed about the diagnosis by multidisciplinary team consisting of a feto-maternal specialists, Neurosurgeon and Neonatologist. Patient had lower segment cesarean section at 38 weeks of gestation. The newborn was admitted to Neonatal Intensive care unit and due to sustained cardiac failure neonatal death occur after 48 hours.

INTRODUCTION

The merging of the two internal cerebral veins with the basal veins of Rosenthal forms a single vein termed the 'Vein of Galen' which is located midline and posterior to the splenium of the corpus callosum. In normal cases the vein passes posteriorly to empty into the straight sinus. During development, abnormal connections / fistulas arise between cerebral arteries and the Vein of Galen leading to the formation of Vein of Galen Aneurysm (Padget, 1956). The pressure gradient between the abnormal connections act as a supporting factor for its persistence and enlargement.

Aneurysm Vein of Galen also called Vein of Galen aneurysmal malformation (VGAM) is a rare developmental abnormality of the fetal cerebral circulation, representing around 1% of the fetal cerebral arteriovenous system abnormalities and approximately 30% of pediatric vascular malformations[1,2].

This malformation is usually isolated, but there are some reported cases where other anomalies are also present. The VGAM arises around the 6th and the 11th week of pregnancy[3]. In his study of Anatomic analysis of 23 cases of Vein of Galen aneurysm, Raybaud found that the venous aneurysm most probably represents persistence of the embryonic median porencephalic vein of Markowski rather than the Vein of Galen (Raybaud, et.al 1989).

The size of the aneurysm of the vein of Galen will determine its clinical presentation. If the aneurysm is large, the arteriovenous connection will shunt 50% to 60% of the cardiac output (Cumming, 1980). This shunt leads to high-output congestive heart failure, hydrops in utero, or with cardiac failure in early neonatal life. Other cases of vein of Galen aneurysm are not associated with cardiac failure and may not present until the first year of life.

CASE REPORT

A pregnant woman aged 30 years old, gravida 3 para 2 at 37 weeks of gestation referred to our institution [Salmaniya Medical Center] from a private clinic with suspicion of VGMA associated with dilatation of the superior vena cava and dilated right atrium of the hearts.
A consent from the patient to present the ultrasound images and the case was obtained.

The 2D real time ultrasound scan was repeated, and midline tubular cystic structure was identified, color doppler showed high vascularity consistent with dilated Vein of Galen measuring [1.25 x 4.2 cm]. There were no evidence of other structural abnormalities in the brain. [Figure 1]

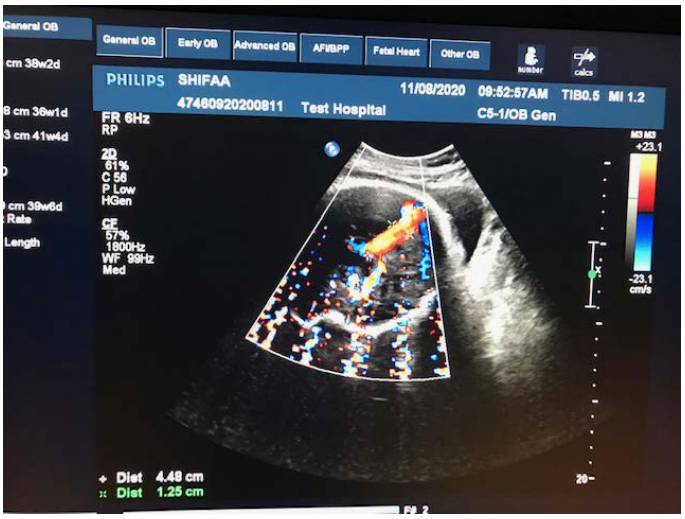

Figure 1: Color Doppler of VGAM

A fetal echo was performed by pediatric cardiologist which showed dilated superior vena cava, right atrium, right ventricle and pulmonary artery. Volume overload on the right side with preserved systolic function and pulmonary hypertension was noted. These changes are secondary to systemic atriovenous malformation of Vein of Galen.

The heart at the 4-chamber view was dilated and occupying two third of the chest diameter indicating volume overload. No polyhydramnios or any other fetal abnormalities were noticed during scanning.

The case was discussed with neonatal intensive care unit (NICU) and neurosurgeon regarding the management of this case. Decision was made to admit this patient to Salmaniya Medical Center and proceed with delivering the baby by elective cesarean section. The aim is to stabilize the baby in terms of treating the heart failure in order to

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proceed with radiological intervention by transvenous and transarterial embolization technique if possible.

The baby was delivered by cesarean section on 13th of August 2020, intubated and shifted to (NICU). immediate postnatal portable chest x ray showed cardiomegaly.

Fetal brain MRI was arranged. MRI confirms the antenatal diagnosis of Vein of Galen malformation. There is a large aneurysmal median prosencephalic vein (Vein of Galen Malformation) (2x2.4 x4.5cm) (Trans x CC x AP) at the pineal region of the brain draining into the dilated straight sinus and the sinus confluence at the Torcular Herophili with multiple collateral vessels around the brain stem and posterior fossa.

There are bilateral symmetrical multiple tortuous arteries at the brainstem more marked at the brain along the ambient and interpeduncular cistern. Dilated and tortuous vessels are seen at the circle of Willis and also prominence of the vessels along the lateral wall of the ventricles. No evidence of hydrocephalus is seen. Mass effect is seen at the supra vermian cistern.

The frontal horn of the lateral ventricles measures 0.3cm. The third ventricle is not dilated. The fourth ventricle is normal. No evidence of intracerebral hemorrhage is seen Normal myelination is seen at the posterior limb of the internal capsule, the dorsal aspect of the brainstem.

The brain is otherwise structurally normal however the corpus callosum appears to be thinned out due to pressure effects from the dilated vessels. Signal intensity is seen at the pituitary gland. The posterior fossa structures are otherwise normal.

The visualized part of the orbits and cavernous sinus are normal. The extra-axial CSF spaces.
MRA and MRV shows the Vein of Galen aneurysmal malformation with dilated collateral arteries along the circle of Willis.

CT Brain angiography with 3D reformatted images was performed to delineate the connections of the (aneurysmal median prosencephalic vein) Vein of Galen Malformation and the large draining vessels, this explicitly displayed the multiple dilated collateral vessels with choroidal arteries. [Figures2,3 4,5]. Baby expired on 17/08/2020 due to congestive heart failure.

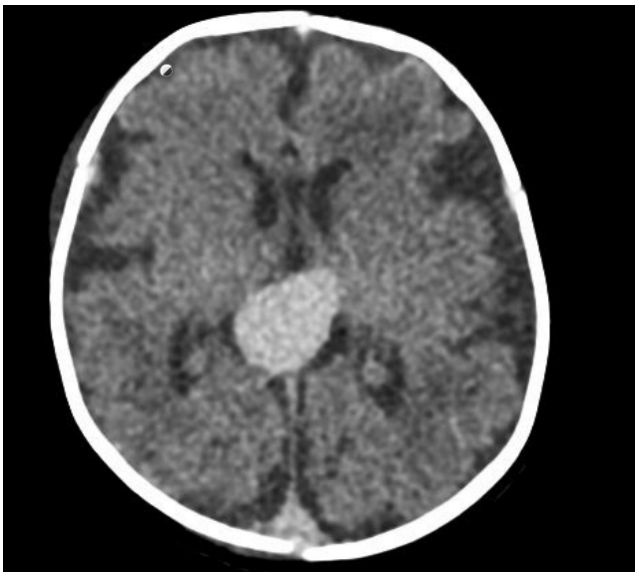

Figure 2: VGAM in MRI Brain

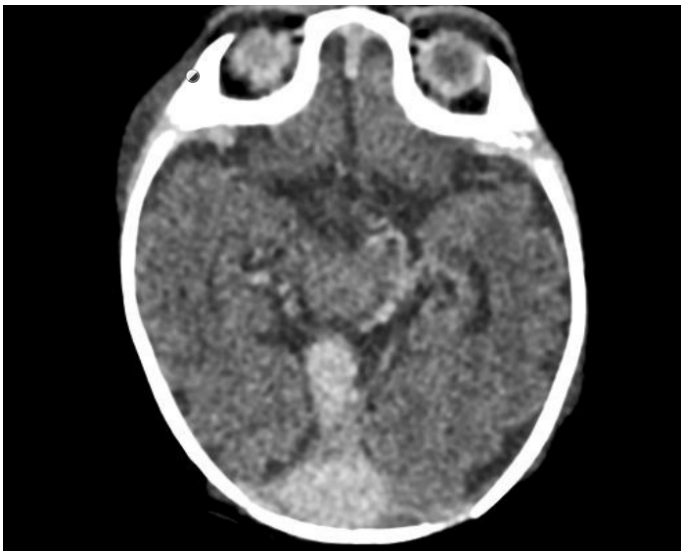

Figure 3: VGAM in MRI Brain

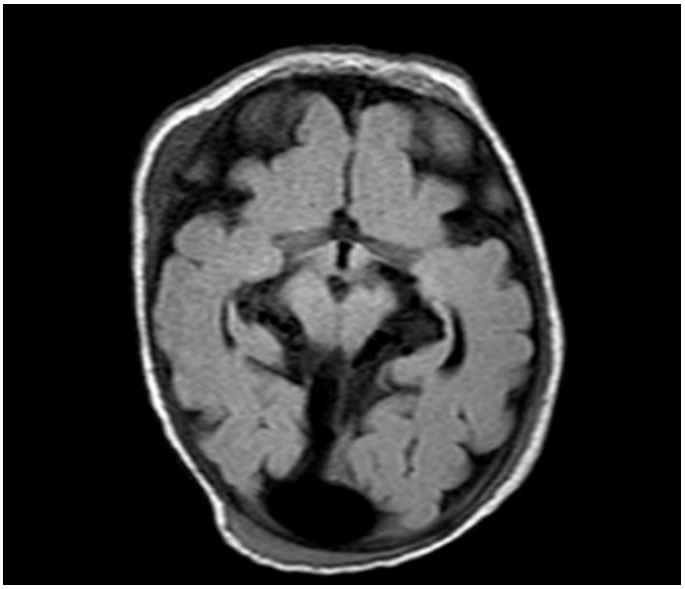

Figure 4: VGAM inb rain MRI Brain

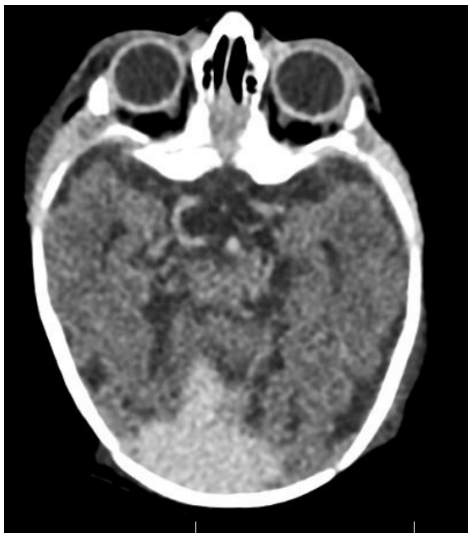

Figure 5: CT angio Brain

DISCUSSION

Vein of Galen malformations are rare anomalies of intracranial circulation that constitute 1% of all intracranial vascular malformations. In pediatric age group they represent 30% of vascular malformations[1,3].

Although any vessel can be affected the vein of Galen is most frequently affected. Congenital malformation develops during the weeks 6-11 of fetal development as persistence embryonic pros encephalic vein of Markowski. Pros encephalic vein drain to vein of Galen. These lesions are characterized by the presence of anurosmally dilated midline deep venous structure fed by abnormal arteriovenous communications[4,5].

There have been several classifications of VGAM, based on, type of supplying arteries, complexity and the location of fistula[6].
One of the favored classifications is by lasjaunias who divides VGAM into two types: choroidal or mural. The choroidal type is characterized by multiple feeders from the choroidal arteries and other deep midbrain arteries that form a fistula and causes fetal congestive heart failure. The other type is mural lesions, characterized by a fistula. They typically have less degree of heart failure because of fewer feeding arteries[7].

VGMA are occasionally diagnosed antenataly by ultrasound at 25 weeks of gestation. It appears like a cystic structure in the midbrain, and with aid of Color Doppler and MRI the diagnosis and the extension of brain tissue damage will be confirmed. In our case the diagnosis was confirmed at the late third trimester with development of cardiac failure.

A team approach is very important for successful management. Ideally the management is conservative. Embolization of the newborn is a high-risk procedure. It is advisable to treat the neonate medically for cardiac failure until the age of six months then embolization can be done for the arteriovenous malformation. Surgery has small role in the management of VGMA as it has severe morbidity and high mortality rate[2,8].

CONCLUSION

We presented a case of VGAM with cardiac failure antenataly which was diagnosed in the third trimester. The 2D real time ultrasound scan with Color and pulsed Doppler accurately identified the lesion. When there is VGMA with preexisting cardiac failure a poor outcome is inevitable.

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