Case Report

Vein of Galen Aneurysm: Antenatal Diagnosis: a case report

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Abstract

Aneurysm of vein of Galen is a rare congenital malformation with reported incidence of less than 1% of cerebral vascular malformations. Most cases present in neonatal life with congestive cardiac failure. We report a case of aneurysm of vein of Galen diagnosed in intrauterine life at 36 weeks of gestation. The importance of third trimester Doppler ultrasound is highlighted.

Introduction

Vein of Galen aneurysm is a rare congenital brain malformation[1], and a rare cause of congestive cardiac failure. In a patient presenting with congestive cardiac failure, vein of Galen aneurysm should be considered a diagnostic possibility when cardiac causes have been excluded. Although the abnormality develops between the 6th and 11th weeks of gestation[2], prenatal diagnosis is usually possible in the third trimester of pregnancy.[3] Most cases are diagnosed postnatally.[4]

Case report

A 33 year old woman reported for a routine ultrasound evaluation at 36 weeks of pregnancy. Since she had 2 previous caesarean sections, she was referred for localization of placenta and possible placenta accreta. Her 2 previous ultrasound examinations done at 16 and 26 weeks of gestation were normal. Her present ultrasound examination

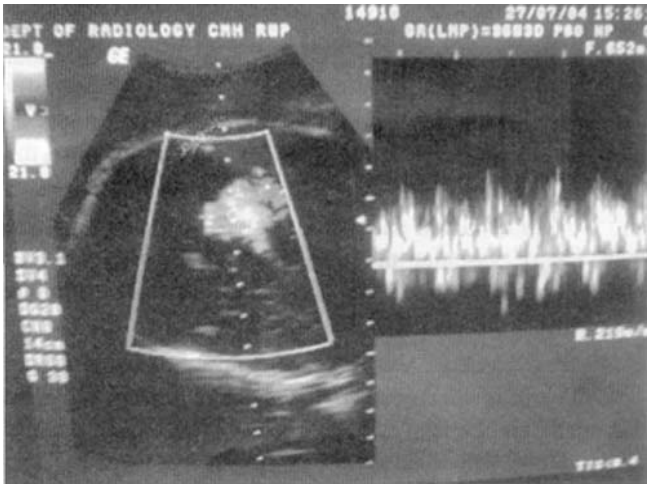

Figure 1. Doppler ultrasound of brain showing a cystic area in right thalamus with turbulent flow in it.

revealed a large cystic area in region of right thalamus extending posteriorly below the splenium (Figure 1). Colour Doppler sonography showed turbulent flow in the cyst. The venous sinuses were enlarged. The large vascular channel drained into an enlarged and prominent Internal Jugular Vein and Superior Vena Cava (SVC) was dilated.

Examination of the foetal heart showed an enlarged, hypertrophied right ventricle and enlarged pulmonary artery. E/A ratio was reversed across the Mitral valve and equalized across the Tricuspid valve (E/A ratio is the ratio of passive

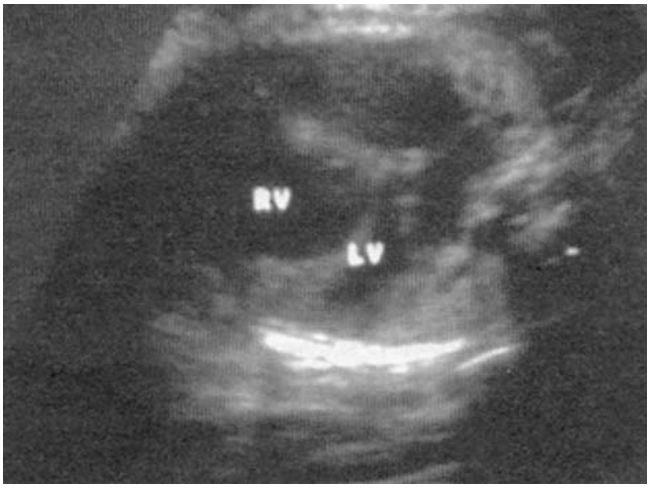

Figure 2. Fetal echocardiogram showing an enlarged right ventricle with interventricular septum deviated to left side.

ventricular filling to the filling due to atrial contraction. Normally in foetal life, ventricular filling due to atrial contraction is more than the passive filling of the ventricles). On the basis of ultrasound findings a diagnosis of vein of Galen aneurysm was made and parents were counseled for the outcome. A baby boy was delivered by Caesarean section, who was cyanosed at birth and died after four hours of birth.

Discussion

Vein of Galen is a short midline venous structure formed by the confluence of the two internal cerebral veins and the basal veins of Rosenthal. It represents the caudal remnant of the median prosencephalic vein, a centrally located vessel that drains the choroid plexus. Malformations of Galenic vein are believed to result from an insult to the cerebral vasculature between 6 and 11 weeks of gestation.[2]

Vein of Galen aneurysm (VOGA) accounts for less than 1% of vascular malformations of the brain but in paediatric population, 30% of vascular malformations of brain are due to vein of Galen aneurysm.[1]

Few cases of VOGA are diagnosed in utero. About 40% of cases are diagnosed in the neonatal period[4] while the rest are diagnosed later in life. Antenatal diagnosis is usually made after the 30th week of gestation because the malformation grows as pregnancy advances. Therefore, the importance of third trimester ultrasound is emphasized.[3] In our patient, 2 previous ultrasound examinations were normal and this abnormality was detected in 36th week of gestation, when the patient was referred for an unrelated problem.

The differential diagnosis includes conditions such as cavum vergae, arachnoid cyst, and porencephalic cyst. All these entities do not show flow on colour Doppler imaging.[5]

VOGA may be associated with congenital cardiac defects such as sinus venosus atrial septal defect, coarctation of aorta, partial anomalous pulmonary venous return, patent foramen ovale, ventricular septal defects and atrioventricular canal defects.[6,7] In our patient, we identified right ventricular hypertrophy and dilatation of Pulmonary artery secondary to high venous return from the brain. No other structural cardiac defect was identified.

This malformation is an entity which is known to regress spontaneously at times. It has been documented that Galenic aneurysm with low flow states thrombose and regress with time. Beltramello et al have reported thrombosis of Galenic aneurysm in 11 out of 21 patients, as demonstrated on follow up angiography.[8]

Various treatment options are available for patients which include embolisation and surgical correction. The prognosis also depends on the stage at which the patient presents. The earlier the patient presents with symptoms, the worse is the prognosis. Children usually present with high output cardiac failure. The child in our case report was cyanosed at birth and had severe respiratory distress. Echocardiography in these children fails to reveal structural cardiac defects. In such children, besides other causes of respiratory distress, VOGA should be considered a diagnostic possibility and appropriate modalities should be employed for diagnosis, depending on the stage at which the patient presents (Doppler ultrasonography of brain in a neonate, Magnetic Resonance Imaging (MRI) in older children). Treatment outcome depends upon the expertise of the center where the treatment is carried out.

Vein of Galen aneurysmal malformations are rare congenital anomalies. Prenatal diagnosis is usually possible only in the third trimester, hence the importance of a third trimester ultrasound is emphasized. A cystic brain lesion always requires Doppler evaluation to differentiate from other brain cysts. Children with these malformations usually present with high output cardiac failure. In the absence of a structural cardiac defect to account for the patient's symptoms, VOGA should also be considered as a diagnostic possibility.

References

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