Annals of Cardiology and Cardiovascular Medicine

Case Report
Published: 15 May, 2024

Fetal Cardiac Failure due to Vein of Galen Malformation

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Abstract

Vein of Galen malformation is a rare cerebrovascular anomaly and is diagnosed in a fetus on routine antenatal ultrasound or anomaly scan. Fetal cardiac failure occurs due to volume overload of the right ventricle resulting from increased cerebral venous return. Fetal echocardiography can help in the diagnosis of cardiac complications. Cerebral complications can also occur and can be diagnosed on fetal neuroimaging. We present the case of a 33 weeks fetus with vein of Galen malformation and evidence of cardiac failure which is a poor prognostic sign.

Keywords: Vein of Galen malformation; Fetal cardiac failure; Hydrops fetalis; Fetal imaging

Introduction

Vein of Galen Malformation (VGM) is a rare and complex cerebrovascular anomaly representing 1% of the abnormalities of the fetal cerebral vascular tree [1]. This is an arterio-venous malformation that results in shunting of arterial blood into veins at the cerebral cortex and vein of Galen [1]. As a consequence, increase venous return comes from cerebral veins to the superior vena cava, right atrium and right ventricle. The ensuing increase in preload is poorly tolerated by the fetal myocardium which is inherently restrictive in filling. This results in tricuspid valve regurgitation and right-sided chamber dilatation, thereby causing in cardiomegaly and right ventricular dysfunction, and cardiac failure [2]. We describe a 33-week-old fetus with vein of Galen malformation diagnosed on screening ultrasound and detailed on fetal echocardiography.

Case Presentation

A 29-year-old lady with 33 weeks pregnancy was referred for fetal cardiac evaluation due to the routine antenatal ultrasound diagnosing a vein of Galen malformation. A fetal echocardiogram was done with the echocardiographic machine GE Vivid E95 Ultrasound System, (GE Healthcare Life Sciences, Pittsburg, USA) using the curvilinear 4C probe and standard fetal software for analysis. 2D, M-mode, color flow mapping, and pulsed Doppler were used to analyze the fetal cardiac structure and function according to the criteria mentioned elsewhere [3,4]. There was cardiomegaly (CT ratio measured as area was 0.54, Figure 1), superior vena cava was dilated with normal sized inferior vena cava (Figure 2) with a hugely dilated right atrium (right atrium to left atrium diameter ratio 2.4, normal being <1.2) [4]. There was a moderate enlargement of both ventricles (right more than left) with mild pericardial effusion, depicting a failing heart (Figure 3). There was an associated mid-muscular ventricular septal defect, 2.3 mm in size, with right to left shunt (Figure 4). Moderate tricuspid regurgitation was present with a velocity of 2.9 m/s (Figure 5) and mild coarctation of aorta was seen with a reversal of flow in the arch and isthmus (Figure 6).

Ultrasound examination of the head showed an arterio-venous malformation seen as an aneurysm and a channel arising from it (Figure 7).

Cardiac output of the left and right ventricles was calculated using aortic and pulmonary valve diameters and velocity time integral across them, fetal heart rate, and applying the formula CO = HR × VTI of semilunar valve x {2π D2/4} where, D is the diameter of the valve. The cardiac index was calculated by dividing it by the estimated fetal weight which was 1.6 kg. Cardiac indices were 600 ml/Kg/min for the left ventricle and 660 ml/Kg/min for the right ventricle. The combined cardiac index for both ventricles was 630 ml/Kg/min (normal being 425 ml/kg/ min) [2]. The Tei index for the left ventricle was 0.44 and for the right ventricle was 0.48 (both normal).

Parents were counseled for guarded prognosis in a combined meeting with a multidisciplinary team comprising of an Obstetrician, Neonatologist, Pediatric Cardiologist, Pediatric Neurologist and Interventional Radiologist. Concerns highlighted in that meeting were the presence of cardiac failure and hydrops fetalis, and fetal growth restriction, estimated fetal weight being 1.6 Kg. The

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Received Date: *05 Apr 2024*

Accepted Date: *07 May 2024*

Published Date: *15 May 2024*

Citation:

Atiq M, Korar AA, Waqar F, Ahmed F. Fetal Cardiac Failure due to Vein of Galen Malformation. Ann Cardiol Cardiovasc Med. 2024; 7(1): 1055.

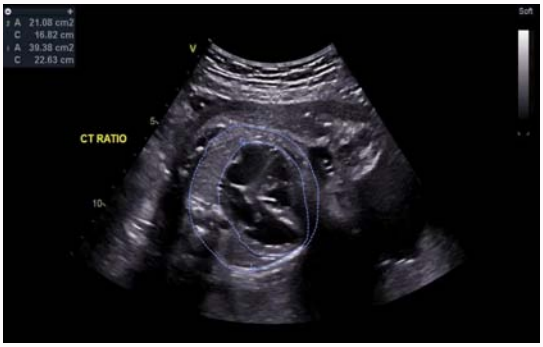

Figure 1: Cardiothoracic (CT) ratio of 0.54.

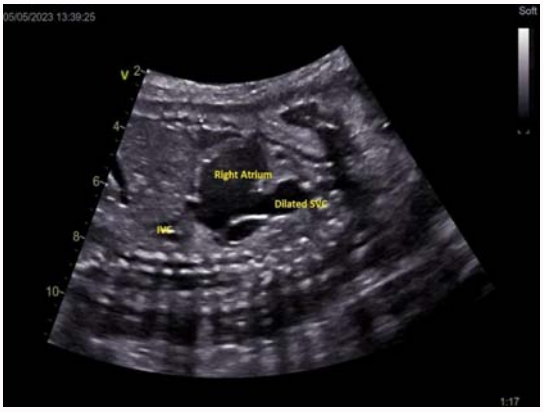

Figure 2: Enlarged Superior Vena Cava (SVC), normal Inferior Vena Cava (IVC) and enlarged right atrium.

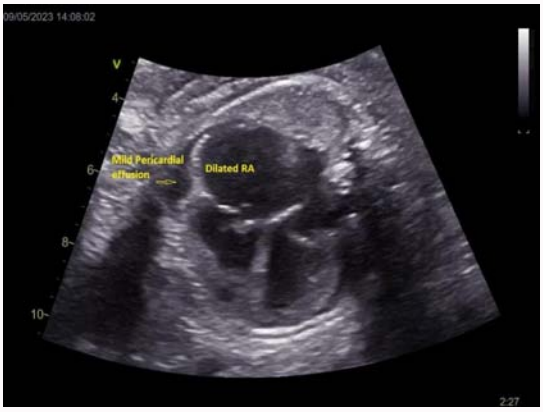

Figure 3: Dilated right atrium and mild pericardial effusion (arrow).

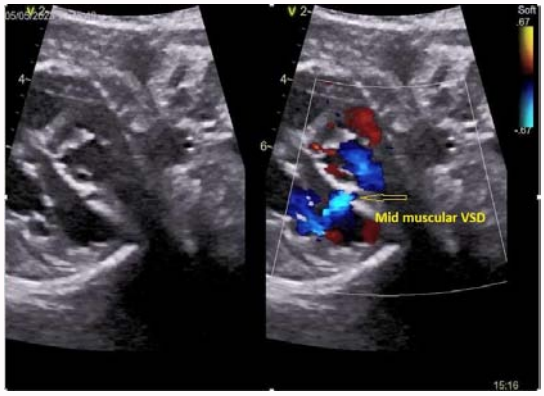

Figure 4: Small mid muscular ventricular septal defect (VSD, arrow).

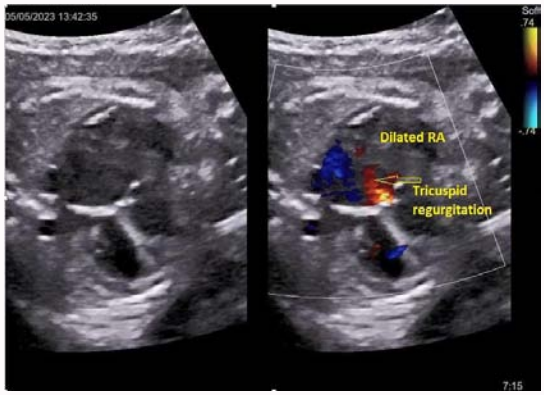

Figure 5: Tricuspid valve regurgitation (arrow at the red jet).

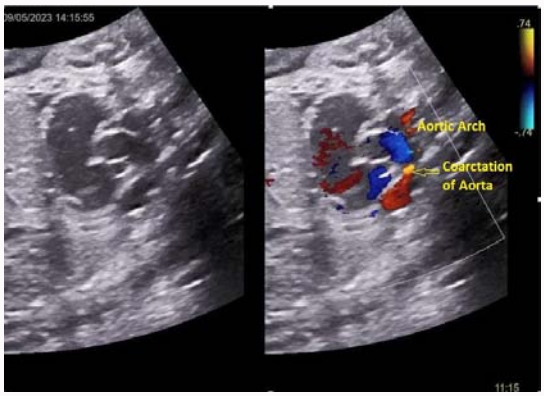

Figure 6: Aortic arch with coarctation of aorta (arrow) with flow reversal (red color).

low fetal weight precluded immediate postnatal cerebrovascular intervention for the vascular malformation. However, it was agreed to provide full medical support at and after birth.

Discussion

Malformation of the vein of Galen is an uncommon cerebrovascular anomaly comprising 1% of cerebral vascular defects [1] with a prevalence of 1 in 25,000 live births [5]. The male-to-female ratio is 3:1 [6]. In this anomaly, one or more arterio-venous fistulae develop between the 6th to 11th weeks of gestation and connect with a dilated median prosencephalic vein of Markowski, which is the embryological precursor of the vein of Galen [5].

It is usually diagnosed in the third trimester of pregnancy [6]. It can occur as an isolated abnormality but one-third may have associated congenital heart defects. Sinus venosus atrial septal defects, ventricular septal defects, and coarctation of aorta [1] have been described. Our patient had a small muscular ventricular septal defect as an associated congenital heart defect with mild coarctation of aorta.

The arterio-venous fistula creates a low resistance and high flow pathophysiology. The ensuing high cardiac output state causes an increase in venous return through the superior vena cava thereby producing a dilatation of that vein as well as right-sided cardiac

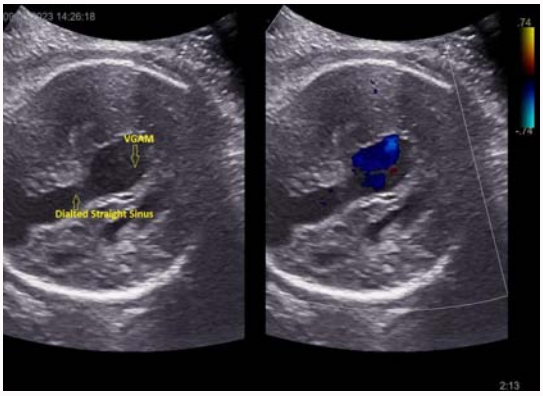

Figure 7: Cerebral vessels showing dilated straight vein (upward pointing arrow), vein of Galen malformation (downward pointing arrow).

chambers and pulmonary artery. Fetal echocardiography is done to assess cardiac failure [7].

Due to the restrictive nature of fetal myocardium, tricuspid regurgitation ensues. Cardiac failure is diagnosed when the cardiothoracic ratio is >0.5 [8], tricuspid regurgitation velocity is >2 m/s, right ventricular dysfunction is present, and hydrops fetalis is seen as pericardial effusion and ascites [9]. There is a reversal of blood flow at the aortic isthmus and is due to a steal phenomenon in the cerebral circulation caused by excessive cerebral blood flow [6]. In the report by Paladini et al. [8], the detection of aortic isthmus flow reversal on fetal echocardiography was present in 14 of the 29 fetuses and it co-related with poor neurological prognosis. Our patient had a high combined cardiac output with all echocardiographic features of cardiac failure and reversal of flow in the aortic isthmus.

Neurological complications are due to increased pressure in the venous sinus, cortical and medullary veins. This results in excessive fluid in the brain parenchyma leading to impaired oxygenation and sub-ependymal brain atrophy which may progress to the so-called melting brain syndrome [6]. Hence it is recommended that these fetuses should not only have detailed echocardiography but also neuro-sonography and brain MRI.

Correlates of poor outcome in the fetus were a blood volume of VGM of ≥ 20,000 mm^3, major brain abnormalities/damage on brain MRI, or tricuspid regurgitation. Tricuspid regurgitation is also associated with an increased risk of associated major brain lesions [8]. Other studies found an increased combined cardiac output and heart failure as a risk factor for poor outcome [2,8]. We did not have the facility to perform a fetal brain MRI at our institution.

Conclusion

The vein of Galen malformation is a rare anomaly of cerebral vasculature with variable outcomes. A detailed evaluation by fetal echocardiography and fetal neuroimaging is important to prognosticate this complex anomaly. Poor outcomes are expected if there is cardiac failure, tricuspid regurgitation, and high volume of blood in the vascular malformation or associated brain abnormalities.

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