Vein of Galen aneurysmal malformation: Prenatal ultrasound diagnosis

A 23-year-old primigravida from a non-consanguineous marriage was referred by her primary obstetrician for a routine third trimester fetal ultrasound at 33 weeks of gestation. The patient had no previous surgical history or significant medical history. The previous fetal ultrasound images were normal.

Imaging Findings:

Our patient’s fetal ultrasound examination revealed dilated cardiac cavities, mainly in the right heart, along with a cardio-thoracic (C/T) area ratio of 0.5 (>1/3), consistent with cardiomegaly (Fig. 1). Dilatation of the superior vena cava (Fig. 2) and the left brachiocephalic vein (Fig. 3) was also noted. No other cardiac abnormalities were present. In addition, an axial B mode ultrasound image through the fetal brain (Fig. 4) showed a cerebral midline anechoic tubular structure above the midbrain with a positive blood flow on Doppler examination (Fig. 5) draining into a dilated sagittal sinus.

These ultrasound findings were consistent with Vein of Galen aneurysmal malformation together with its subsequent high output cardiac failure. Neonatal brain MR images (Fig. 6) confirmed VGAM diagnosis with hypoplasia of the straight and superior sagittal sinuses and the presence of a fistula between the deep choroidal arteries and the median prosencephalic vein. No significant parenchymal abnormality was found.

Discussion:

Vein of Galen aneurysmal malformation (VGAM) remains a rare intracranial cerebral vascular abnormality, although being the most common symptomatic cerebrovascular malformation presenting in neonates and infants [1]. The malformation is due to an arteriovenous fistula between the primitive choroidal vessels and the median prosencephalic vein (MPV) that develops during the 6th to 11th weeks of gestation [2]. Subsequently, the MPV fails to regress and becomes aneurysmal. The shunted blood flow will be drained through the persistent MPV into the sagittal sinus via a persistent falcine sinus or less commonly via the straight sinus (often absent). This will lead to a delay in the MPV involution, preventing proper development of the vein of Galen [3]. Endothelium proliferation is induced by the rapid and turbulent blood flow as well as anomalous dural sinuses.
resulting in secondary sinus stenoses, commonly associated with VGAM [4]. The high output left-to-right shunt will generally result in cardiac failure leading to hydrops fetalis in severe cases [5]. Despite their different aetiology, the true VGAM and the aneurysmal dilatation of the vein of Galen are sometimes considered to be in the same category. Several systems are used to classify the VGAM; these are either based on the clinical presentation, the malformation complexity (Yasargil classification) or the number and origin of feeding arteries (Lasjaunias classification) [3].

With the advent of antenatal ultrasound, the diagnosis of VGAM is generally made in utero [6]. It is mostly reported in the 3rd trimester since it usually appears late in pregnancy.

Detection of a midline supra-thalamic cerebral anechoic tubular structure showing a prominent flow on Doppler examination and contiguous with a dilated sagittal sinus is generally sufficient for the diagnosis [6]. Associated ultrasound features include cardiomegaly, enlarged neck vessels and ventriculomegaly [6]. Complementary intrauterine MR imaging provides additional information, allowing better evaluation of the malformation and the associated cerebral parenchymal damage [7]. Neonatal MR imaging and MR angiography are helpful for planning of endovascular procedures and for follow up [8].

The main presenting postnatal feature is cardiac failure [5]. Several perinatal prognostic predictors have been suggested, nevertheless, a full clinical evaluation is mandatory to determine the intervention timing [8].

VGAM is fatal in 100% of untreated cases [9]. Before the era of endovascular interventions, surgical attempts failed to save 90% of VGAM patients [10]. Nowadays, in the absence of gross cerebral parenchymal damage, endovascular treatment techniques using multiple embolization strategies are feasible, showing promising results, thereby increasing the chances of surviving this once deadly malformation as well as reducing morbidity [8, 11].

**Differential Diagnosis List:** Vein of Galen aneurysmal malformation, Fetal interhemispheric cyst, Cerebral cystic teratoma, Thrombosed subdural vascular malformation, Total anomalous pulmonary venous return (TAPVR)

**Final Diagnosis:** Vein of Galen aneurysmal malformation.

**References:**

Figure 1

Description: Four chamber cardiac view - showing cardiomegaly (Cardio-thoracic area ratio $a/b = 0.5$) predominantly in the right cavities. Origin: Abou Jaoude Hospital, Department of Radiology, Jal El Dib, Lebanon.
Description: Three-vessel view - showing a dilated superior vena cava. ao: Aorta, pa: Pulmonary artery, svc: Superior vena cava. Origin: Abou Jaoude Hospital, Department of Radiology, Jal El Dib, Lebanon.
Description: Axial view slightly above the three-vessel view – demonstrating a dilated left brachiocephalic vein (arrow) draining in the dilated superior vena cava. svc: Superior vena cava. Origin: Abou Jaoude Hospital, Department of Radiology, Jal El Dib, Lebanon.
Description: B mode image just above the midbrain level demonstrating a midline anechoic tubular structure (arrow). Origin: Abou Jaoude Hospital, Department of Radiology, Jal El Dib, Lebanon.
Description: Color Doppler image just above the midbrain level showing a positive turbulent blood flow in the midline anechoic tubular structure. Origin: Abou Jaoude Hospital, Department of Radiology, Jal El Dib, Lebanon.
Figure 6

Description: Brain MRI (T2). Axial slice across the lateral ventricular antra (a) and mid-sagittal slice (b) showing median interventricular large flow void compatible with the dilated prosencephalic vein. Sagittal image showing straight sinus hypoplasia. Origin: Hôtel-Dieu de France - Radiology department, Achrafie, Beirut, Lebanon.