Case Report

A Severe Case of Aneurysmal Malformation of the Vein of Galen, Cardiac Failure and Intrauterine Fetal Death

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ABSTRACT: Aneurism of the vein of Galen is a rare congenital anomaly, where complex arteriovenous malformation are identified between multiple choroidal arteries and the median prosencephalic vein of Markowski, which is a precursor of the vein of Galen, with subsequent aneurysmal enlargement of the respective arteriovenous system. The congenital malformation develops during week 6 to 11 of fetal development. Infants often die from high-output congestive heart failure. We present a case of a 40 years old patient, presented the first time at hospital at 28 weeks of gestation for lower perception of fetal movements. The patient was referred to our Prenatal Diagnosis Unit for a suspected cardiac malformation. We diagnosed cardiomegaly along vein of Galen aneurysmal malformation (VGAM) with severe cardiac failure, ascites, and critical fetal distress. Fetal demise was noted 24 hours later, during the corticosteroid procedures for fetal maturation. A stillborn weighting 2000g with a severe hydropic aspect was born after labor induction. Autopsy was performed following injection of colored gelatin in carotid vessels. The aneurysmal defect was evident in the vein of Galen and straight sinus. We also found the transverse, sagittal and the occipital sinus seriously dilated. We could not highlight the communication with the middle cerebral artery, because of the degradation of the cerebral tissue, despite an optimal preparation.

KEYWORDS: *vein of Galen, fetal aneurysm, cardiomegaly, cardiac failure, intrauterine death*

Introduction

VGAM is a rare fetal anomaly, of which the neurological outcome may be favorable in certain cases and appropriate perinatal management.

The background of this defect that develops during 6-11 embryonic weeks, involves presence of one or more arteriovenous fistulas directing blood flow toward a persistent, dilated, proximal part of median prosencephalic vein of Markowski, a precursor of the vein of Galen, with subsequent aneurysmal enlargement of the arterial and venous system [1].

Although the fetuses/infants often die from high-output congestive heart failure [2], since endovascular treatment became a therapeutic option the survival rates have significantly increased [3].

Regarding the prognostic value of some prenatal features, the group of Palladini found that major brain lesions, tricuspid regurgitation and, to a lesser extent, VGAM volume ≥20 000mm3 are associated with poor outcome in fetal VGAM [4].

VGAM may be missed or mistaken for a cystic lesion of the brain if color Doppler is not used to differentiate cystic lesions from vascular aneurysms. It can be misdiagnosed as arachnoid cyst, cavum vergae, porencephalic cyst, hydrocephaly or intracranial hemorrhage [5].

The heart abnormalities may mimic cardiovascular malformations such as coarctation of aorta [6] and other findings as isolated relapsing bilateral hydrothorax were also described [4].

Thus, a carefully complete assessment of the malformation site and the secondary modifications should be done.

The 3D ultrasound can be used as advent image technique in prenatal diagnosis of VGAM, but the benefit was not proven. 3D power Doppler has the potential to map the aneurysm and its vascular connections, and this technique can also assess the relationships between vascular and brain structures [7,8].

Case report

We present a case of a 40 years old patient, with 4 previous uneventful births and a 28 weeks pregnancy on date of admission with no prior antenatal care, when she described lower perception of fetal movements.

10.12865/CHSJ.44.01.13

Ultrasound examination showed a living severely hydropic fetus, with cardiomegaly and hepatomegaly.

Further assessment of the heart revealed normal structural relations, persistent left superior vena cava, and cardiac failure with markedly decreased ventricular contractility, absent/reversed "a"-wave of ductus venosus flow and massive tricuspid and mitral regurgitation (Fig.1).

The fetal brain evaluation complemented with color Doppler found an aneurism of the vein of Galen, with a shunt from the middle cerebral artery (Fig.2).

Therefore, the heart failure was considered due to the Galen shunt, leading to increased inflow to the right atrium.

The severe fetal distress was shown by umbilical artery reversed end-diastolic flow and a poor Manning score, with absent active and respiratory movements.

According to our institution customs, immediate extensive exploration and fetal maturation corticosteroid therapy was decided. Standard blood samples, TORCH analysis and glucose tolerance test were normal.

The following day after admission fetal demise occurred. After labor induction was completed, an overweighed for gestational age stillborn of 2000g was delivered, with a severe hydropic aspect (Fig.3A).

Fetal genetic assessment was declined. The autopsy report confirmed the Galen vein aneurism. We used a special tracing technique [9] by injecting colored gelatin in the venous and arterial system of the head to demonstrate the aneurism of the vein of Galen with conventional autopsy (Fig.3B,C).

After catheterization of the right and persistent left superior vena cava and aorta, we injected blue colored gelatin in the venous system and green colored gelatin in aorta.

We remarked a very good passage of colored gelatin substances in the venous system and high resistance pressure in the arterial system due to massive thrombosis.

Dissection was made after 24 hours. The aneurysmal defect was shown in the vein of Galen and the straight sinus, while transverse, sagittal and occipital sinuses were also seriously dilated (Fig.3D,E).

The degradation of the cerebral tissue impaired the proper evaluation of the middle cerebral artery communications.

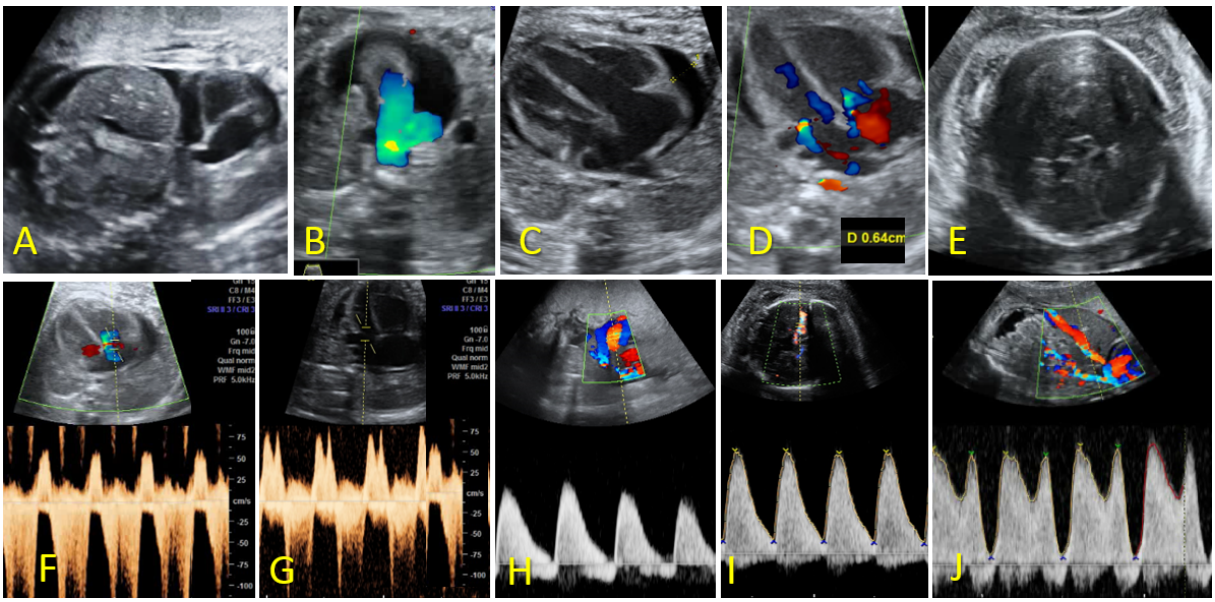

Fig.1. Cardiac failure highlighted by ultrasound evaluation. A: hydropic fetus, with ascites, hepatomegaly and pericarditis; B,C: pericarditis, evident in four-chamber view and outflow tracts view; D: cardiomegaly, tricuspid and mitral regurgitation; E: fetal scalp edema; F,G: tricuspid and mitral regurgitation; H: reversed end-diastolic flow in umbilical artery; I: abnormal cerebroplacental ratio; J: reversed "a"-wave/increased ductus venosus pulsatility index

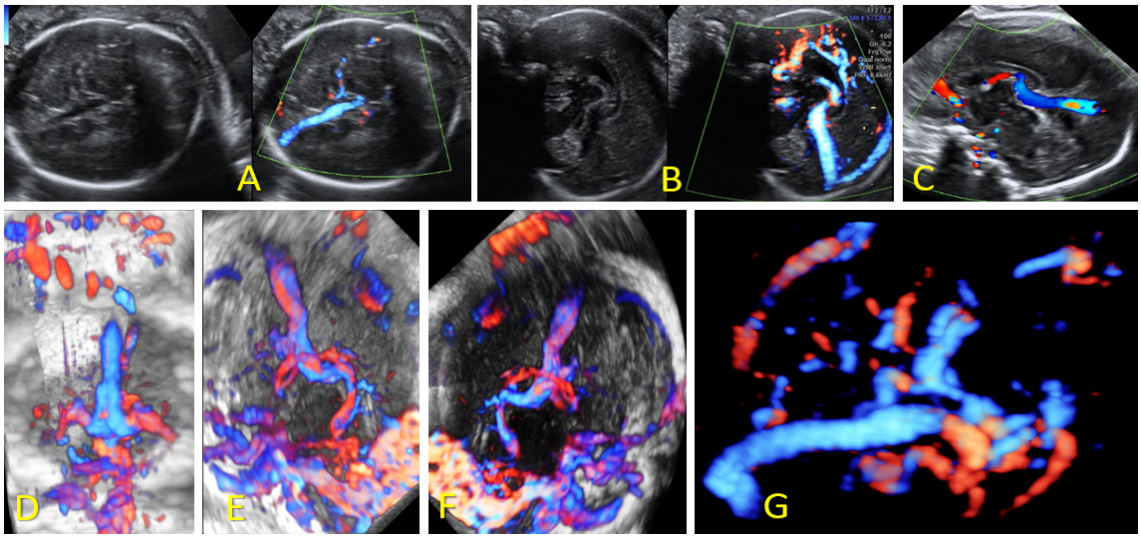

Fig.2. 2D axial (A), and sagittal (B,C) views of the vein of Galen aneurysm. 3D vascular map show the aneurysm in the vein of Galen and the straight sinus, while transverse, sagittal and occipital sinuses appears also seriously dilated in axial (D,E,F), and sagittal (G) planes

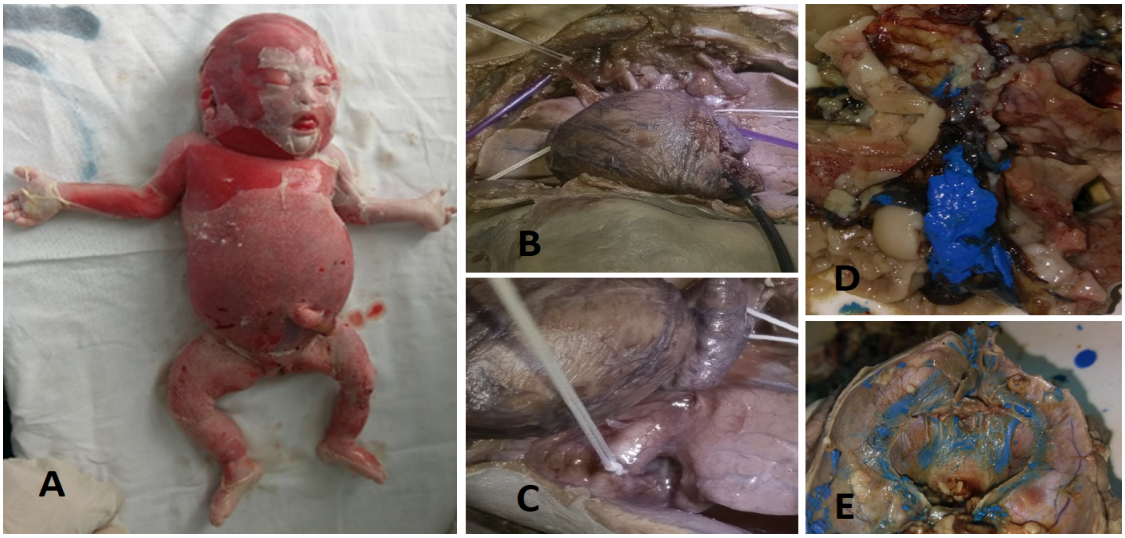

Fig.3. A: hydropic stillborn. B: catheterization of the right and persistent left superior vena cava and aorta. C: ligation of descending aorta. D: dilated vein of Galen and straight sinus filled with blue colored gelatin. E: dilated transverse, sagittal and occipital sinuses

Discussion

The majority of reviews publication related the confirmation of VGAM by using ultrasonography with color Doppler and MRI techniques. In this paper we intended to demonstrate the VGAM by conventional autopsy. Due to degradation of the cerebral tissue we couldn't tracing all the cerebral vascular system with colored gelatin in order to exactly establish the arterio-venous communications. However, this method highlighted a dilated straight sinuses, which could be considered an indirect sign of VGAM.

Aneurism of the vein of Galen is a rare condition, with a good prognosis if is isolated [10] and postnatal treatment is applied to viable foetuses [11]. Associated abnormalities, heart failure or increased aneurysm volume strongly affects the outcome [5,12].

Vijayaraghavan et al. [13] described a case of prenatal diagnosis of thrombosed aneurysm of the vein of Galen, where sonographic findings were confirmed by autopsy that evidence the thrombus in dilated dural venous sinuses. The poor outcome of our case was due to the shunt gravity with subsequent early fetal heart failure and hydrops. Besides the differential diagnosis mentioned above, we ruled out a dilated cerebral venous system observed in growth-restricted fetuses [14] and thrombosis of the posterior dural venous sinuses [15].

Gökçe et al. [16] evaluated on 394 patients the superior dural venous sinuses and classified their types and variations using magnetic resonance venography and digital subtraction angiography

10.12865/CHSJ.44.01.13

and found that 15% of patient presents an occipital sinus. In our case a dilated occipital sinus was found.

Further, Saiki et al. [17] measured on 91 cadavers the calibers of the left and right internal jugular veins and sizes of the left and right transverse sinuses and the drainage patterns of the superior sagittal sinus and straight sinus in the torcular Herophili. They reported that superior sagittal sinus draining was predominant in the right transverse sinus, while the draining of straight sinus was in left transverse sinus, and the size of right internal jugular veins in this situation is high. Authors speculated that may be seen in case of disappearance of the left superior vena cava, and in case of persistence of left superior vena cava they concluded the absence of predominance of right internal jugular veins. We also consider that persistent left superior vena cava that we described may influence the diameter of internal jugular veins that was similar in our case.

Conclusions

Our case underlines the poor prognosis of VGAM associated with severe cardiac failure. We consider that the tracing technique is a useful tool that permitted to establish a correlation between morphological and imagistic aspects of this anomaly, by evidence of vascular alteration.

Acknowledgement

The authors declare that they have no conflict of interests.

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