

IMAGERY OF THE ANEURYSM OF GALEN VEIN

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Abstract

The aneurysm of Galen vein is a rare anomaly of intracranial circulation. Galen vein malformations prognosis in children is unfavorable. Antenatal, Galen vein aneurysm is suspected when an ultrasound examination identifies in the brain, a liquidian structure localized behind the third ventricle. Color Doppler ultrasound helps in differentiating aneurysm of Galen vein from other cystic cerebral median lesions. Ultrasound diagnosis of these malformations could facilitate postnatal therapy, greatly improving prognosis.

Key words: Galen vein, aneurysm, ultrasound

Introduction

The aneurysm of Galen vein is a rare anomaly of intracranial circulation, representing 1% of the intracranial circulation malformations, but it represents also 30% of all fetal vascular malformations.(1) It occurs as a result of direct communication between arterial and venous proencephalic median network. Raybaud et al.(2) found that this abnormal development occurs between 6 and 11 weeks of intrauterine life. Although this anomaly appears in this early period of gestation, prenatal diagnosis is usually possible only in the third trimester, but most cases were postnatally diagnosed. Galen vein malformations prognosis in children is unfavorable, with a high risk of postnatal mortality and a 50% increased risk of neurological disabilities (3). Antenatally, Galen vein aneurysm is suspected when an ultrasound antenatal examination identifies in the brain a structure localized behind the third ventricle. Color Doppler ultrasound helps in differentiating aneurysm of Galen vein from other cystic cerebral median lesions. Ultrasound can also diagnose associated complications such as hydrocephaly or heart failure. The aneurysm of Galen vein can lead to cardiac failure due to the size of the arterio-venous shunt that can steal more than 80% of the cardiac input, with large amount of blood under high pressure returning to the right heart and pulmonary circulation, this is the most common cause of death in these patients.(4) Compression by the dilated vessel of the drainage holes from the cerebrospinal fluid circuit (Monro and Magendie) may induce hydrocephaly, another complication of this condition. If a patient presents congestive heart failure and other cardiac causes have been excluded, we should consider also, the possibility of an aneurysm of Galen vein.(5)

Case presentation

A pregnant woman, primigesta, nulipara, of 27 years old, with no significant pathological personal history, was monitored by the obstetrician from 2 months of pregnancy. Previous ultrasounds showed an apparently normal fetus without structural abnormalities and normal development. The ultrasound performed at 34 weeks of pregnancy, showed at the level of the brain, in the midline, an oval fluid structure of about 5/3 cm placed behind the third ventricle and above the thalamus. The other intracranial components were normal, including third and fourth ventricles. Color Doppler ultrasound identified a turbulent blood flow with a speed of 35 to 40 cm / sec. Hence, the suspicion of arterio-venous aneurysm at the level of the Galen vein was formulated. (fig. 1,2,3,4).

The possibility of a cardiomegaly with tricuspid insufficiency was also taken into account. At the ultrasound examination of the heart, a right ventricle hypertrophy and a dilated pulmonary artery was observed. Problems of diagnosis, prognosis and therapeutic possibilities were discussed with an interdisciplinary team including an obstetrician, a sonographer, a neurosurgeon, a neonatologist and with the parents, of course. A caesarean section was performed at 37 weeks of pregnancy, resulting a new born alive, of 48 cm length, head circumference of 33 cm, chest circumference of 31 cm, Apgar = 8. Immediately postpartum, the newborn showed relatively good general condition, with acrocyanosis, the auscultation describes a systolic murmur grade II/VI localised in left parasternal position, subcostal discrete retraction, but with a saturation and a normal pulse. Following clinical examination and paraclinical tests in particular, the antenatal ultrasound performed at 34 weeks of amenorrhea the diagnosis of Galen vein aneurysm was formulated. The newborn was transferred in the neonatal intensive care unit. The transfontanelar ultrasound showed a venous vascular, central cerebral inter-talamic image, confirmed to be a Galen vein aneurysm (fig. 5).

Echocardiography performed after birth describes the following: large, permeable foramen ovale, permeable ductus arteriosus with left-right shunt, increased dilation of coronary sinus, pulmonary hypertension with right heart dilatation, pulmonary artery dilatation and tricuspid reflux, normal systolic function and ventricular contractility disorders (fig. 6,7).

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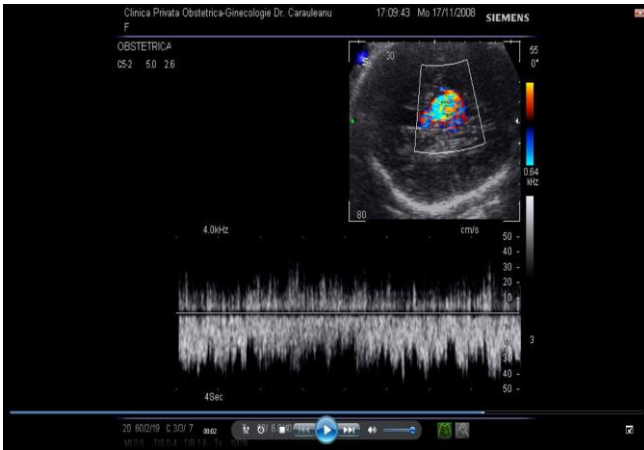


Figure 1. Pulsed Doppler image in the aneurysm.

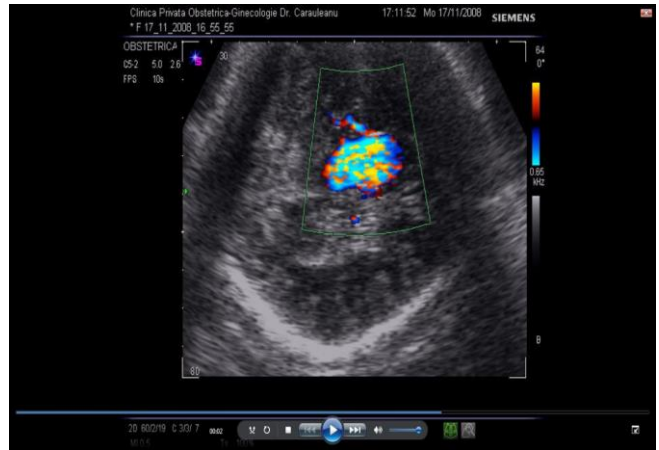


Figure 2. Color Doppler image in the aneurysm.



Figure 3. Color Doppler image in the aneurysm.



Figure 4. 2D ultrasound image in the aneurysm.

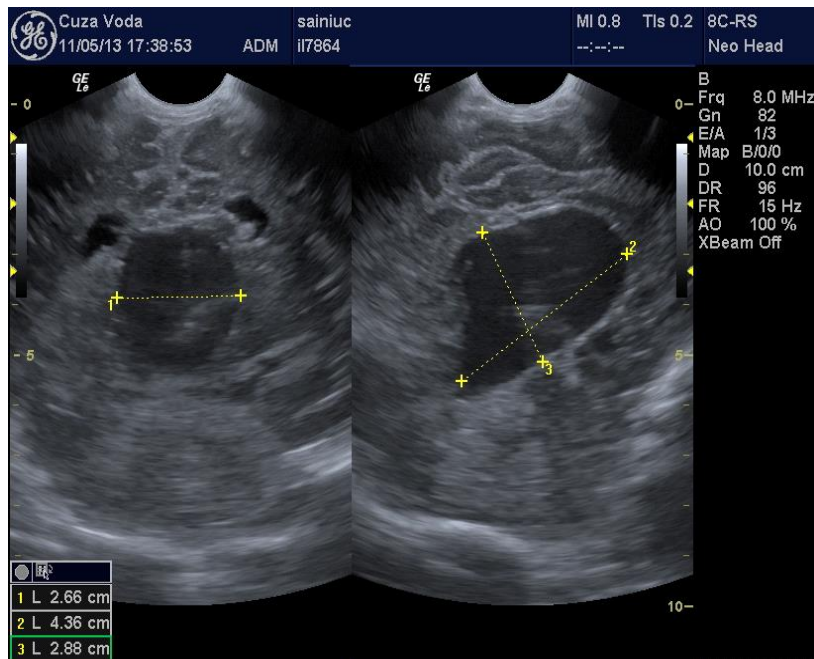


Figure 5. Transfontanelar 2D ultrasound in the newborn.



Figure 6. Doppler echocardiography in the newborn.



Figure 7. Echocardiography in the newborn.

To confirm the diagnosis of Galen vein aneurysm, a CT with contrast is performed showing a giant aneurysmal dilatation of a Galen vein with dimensions of 5,1/2,97/3,5

cm, with homogeneous filling, with nutrient vessels derived from pericallosal artery, polygon of Willis, temporal enlarged fluid spaces, spaced lateral ventricles (fig. 8,9).



Figure 8. Cerebral CT without contrast - Galen vein aneurysm (day 3).

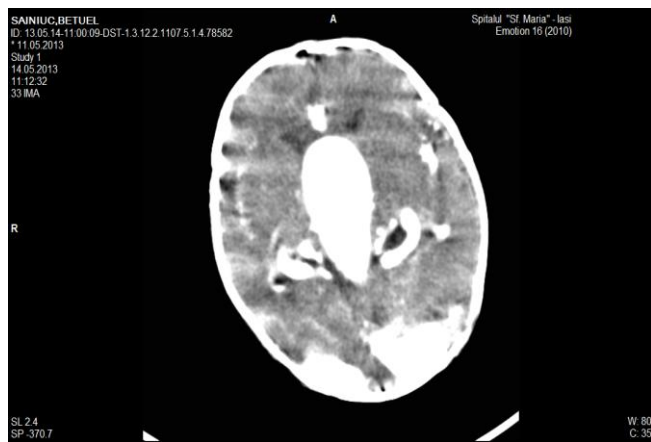


Figure 9. Cerebral CT with contrast - Aneurysm of Galen vein.

During the 2-nd/3-rd day after birth, the newborn's condition deteriorates, on day 5 his condition is mediocre with rapidly progressive edemas, progressive heart failure, hepatomegaly, so that, on day 9, the condition worsens, resulting in newborn death by irreversible cardiac arrest,

resistant to resuscitation. The necropsy found a Galen vein aneurysm complicated with subdural posterior hemorrhage (fig. 10), heart failure, pulmonary multifocal hemorrhage, liver, kidney and spleen with stasis.

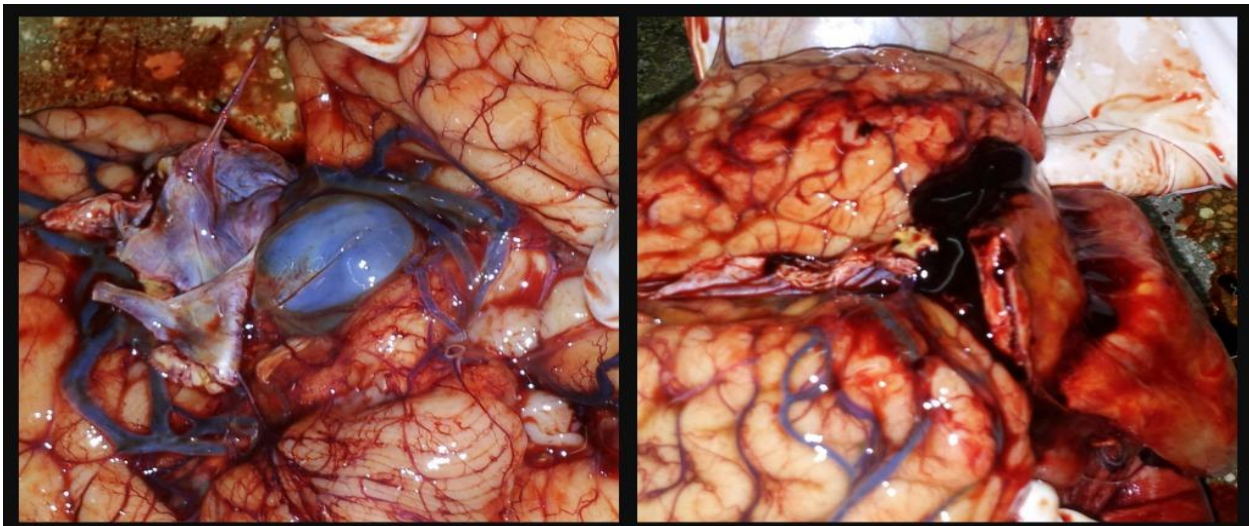


Figure 10. Macroscopy – Rupture of Galen vein aneurysm, hemorrhage.

Discussions

The aneurysm of Galen vein was first described in 1937 by Jager. Until 1984, less than 200 cases have been reported in the literature. Doppler ultrasound has allowed the first prenatal diagnosis. Galen vein aneurysm is a vascular malformation of choroid plexus. Due to the increased flow and turbulence, the vein wall is hypertrophied. Normally, Galen vein venous drainage is directed by sagittal sinus, but in some cases, thrombosis may be missing. The phenomenon of "stealing" is produced because the shunt, with the consequence of cerebral area hypoperfusion. (5,6). The placenta protects the fetus in the fight against the aneurysm, because it has a low resistance, so the amount of blood that reaches the aneurysm is also reduced and congestive heart failure is absent. But if the shunt is major, the placenta cannot compete with aneurysm, and cardiomegaly and fetal hydrops with brain damage may occur. Galen vein aneurysm prognosis depends on many factors, but the size of the aneurysm is not correlated with prognosis. The following factors: structural brain defects, dilated tract drainage, hydrops, dilated jugular vein, superior dilated vena cava, aortic retrograde flow, cardiomegaly, are correlated with a poor prognosis at birth. In utero, few cases are diagnosed Galen vein aneurysm. Forty percent are diagnosed in the neonatal period, the rest being diagnosed much later. (7,8). Antenatal diagnosis is performed after 30 weeks of gestation because malformations develops with increasing age of gestation and therefore pregnancy ultrasound in the third trimester is of utmost importance. In our case, all the ultrasounds examinations performed before 34 weeks of pregnancy couldn't detect the aneurysm. The differential diagnosis is performed with arachnoids cyst and porencephaly cyst. All these entities do not show color Doppler flow inside. Studies have shown that 77% of

untreated cases evolve to death. But even if some cases benefit from surgical treatment, the mortality rate remains also high, around 39.4%. Treatment consists in blocking arterio-venous fistula flowing into the vein of Galen, thus reducing blood flow in the vein. (9,10). Open surgery has a high rate of morbidity and mortality, but recently, the endovascular embolisation is preferred. Unfortunately, in our center this technique is not available, and the case was quickly complicated by heart failure and cerebral hemorrhage and died before we managed to organize a transfer. (11,12).

Conclusions

The widespread use of routine prenatal ultrasound, allowed detection in the third trimester, of several cases of aneurysm of Galen vein. Useful in identifying the anomaly and its differential diagnosis from other lesions with space replacement, Doppler ultrasound is useful in fetal cardiovascular assessment, establishing the gravity of the case. (13).

Ultrasound diagnosis of these malformations facilitated postnatal therapy, greatly improving prognosis. In conclusion, Galen vein aneurysms and its possible complications can be detected by ultrasound and prognostic indices are useful in choosing the best therapeutic alternative.

Prenatal ultrasound identifies an anechogenic formation localized behind the third ventricle, but color Doppler examination is useful in differentiating aneurysm of Galen vein from other cystic median lesions. The same investigation can detect also the complications, such as congestive heart failure or those localized in the brain: cerebral hemorrhage, hydrocephaly.

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