

Nonimmune hydrops fetalis secondary to aneurysm of the vein of Galen

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Ann Saudi Med 2008; 28(1); 45-47

Aneurysm of the vein of Galen is a rare intracranial vascular abnormality associated with severe hemodynamic and central nervous and/or system disturbance leading to intrauterine death and/or early neonatal death.¹ It is associated with a wide range of abnormalities like cardiomegaly, ventriculomegaly, intrauterine growth restriction, hepatomegaly, enlarged neck vessels, polyhydramnios and vascular steal syndrome.² Spontaneous intracranial hemorrhages are not uncommon in these aneurysmic malformations.³ Though nonimmune hydrops has been associated with wide variety of conditions ranging from structural heart disease, arrhythmias, chromosomal anomalies and intrauterine infections, occurrence with aneurysm of the vein of Galen is rare. Diagnosis of this condition is usually made by ultrasound complimented with color Doppler and MRI.³⁻⁵ Though this condition was associated with more than 90% mortality in the past, a favorable outcome has been reported recently with endovascular treatment and intensive neonatal care. However, morbidity cannot be entirely eliminated. We report a case of aneurysm of the vein of Galen that presented with cardiomegaly and hydrops that culminated in intrauterine fetal demise.

CASE

A 26-year-old primigravida was referred to our unit at 30 weeks of gestation for fetal evaluation because of polyhydramnios and hydrops fetalis. There was no family or medical history of note, and ultrasonographic scans done earlier in pregnancy at 18 weeks and 24 weeks had been normal. An ultrasonographic scan repeated in our institution confirmed the presence of hydrops fetalis with generalized skin edema, mild pericardial effusion and ascites. The fetal biometric measurements including biparietal diameter, head circumference, abdominal circumference and femur length corresponded to the period of gestation. The sonographic examination also revealed cardiomegaly with dilatation of all the four chambers and outflow tracts,

tricuspid regurgitation and mild pericardial effusion. The heart, however, was structurally normal. The fetal brain showed a supratentorial anechoic cystic structure in the midcerebral area superior to the third ventricle measuring 25 mm×15 mm with a tubular extension reaching the occipital region. Color Doppler interrogation of this lesion demonstrated a turbulent flow within the cyst while pulsed Doppler analysis showed a bi-directional flow with both arterial and venous flow in the tubular extension (Figure 1). There was no associated ventriculomegaly. A diagnosis of aneurysm of the vein of Galen with hydrops fetalis secondary to cardiac failure was made. The amniotic fluid index was 37 cm, and as the patient was symptomatic, amnioreduction was carried out the next day and the fluid sent for karyotyping. This was subsequently reported normal. The sonographic findings indicated a grave/grim prognosis, but the parents refused any intervention or termination of pregnancy. A day later there was fetal demise. Labor was then induced and a stillborn hydropic male baby weighing 1800 g was delivered vaginally. The couple refused postmortem.

DISCUSSION

Intracranial vascular malformations are rare in the fetus. Aneurysm of the vein of Galen, dural sinus malformations and pial arteriovenous fistulas have been detected antenatally with the vast majority reported being aneurysmal dilatation of the vein of Galen.¹ Galenic vein aneurysm is a potentially disastrous anomaly with a mortality rate exceeding 90% in the past. In the fetal form with severe clinical symptoms of congestive heart failure as in our case, the prognosis is extremely poor. With advances in neonatal intensive care and endovascular embolization, control of cardiac failure and normal neurological function has been reported in 66% cases.²

Aneurysm of the vein of Galen is a complex arteriovenous malformation presumed to result from an abnormal connection between the primitive cho-

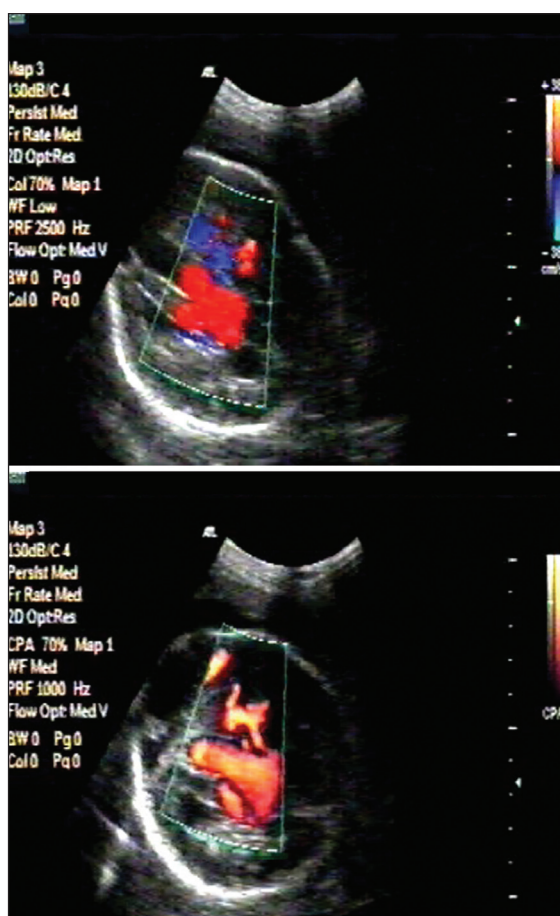


Figure 1. Color and power Doppler images demonstrating highflow and feeder vessels to the aneurysm of the vein of Galen.

roidal vessels and the median prosencephalic vein of Markowski. The afferent vessels include the branches of the vertebrobasilar system and carotid arteries while the draining vessels lead to the straight sinus, falcine sinus. This vascular malformation can lead to hemodynamic decompensation or brain injury during the fetal period or after birth, post natally.³ The vascular “steal” phenomenon leads to increased venous return from the brain resulting in high output cardiac failure and hydrops as in the case reported here. Hydrocephalus is often present and this may be due to compression of the aqueduct of sylvius by the aneurysmal dilatation or due to altered hemodynamics in the reabsorption of the spinal fluid. Brain infarcts and leucomalacia result from pressure or impaired blood flow. Spontaneous in-

tracranial hemorrhages are frequently associated with this malformation.⁴ Rarely it can follow endovascular treatment also.⁴ Prenatal diagnosis of vascular malformation is made essentially by ultrasonography and color Doppler with spectral analysis.^{5,6} Color Doppler will differentiate between a vein of Galen aneurysm and other anechoic intracranial lesions such as arachnoid cyst and porencephalic cysts. Spectral analysis on the other hand will differentiate between aneurysm of the vein of Galen and other malformations like pial arteriovenous fistulas.¹ Complimentary modalities like MRI and three-dimensional color power angiography allow demonstration of the feeding vessels and the venous drainage, may differentiate true Galenic anomalies from pial arteriovenous fistulas, and identify associated brain injury.⁷ Though not essential, these would help in treatment decisions.

The management of pregnancies complicated by fetal intracranial vascular malformations is debatable. Spontaneous thrombosis of the vein of Galen leading to regression in size and even disappearance of the aneurysm has been reported.⁸ However, the presence of congestive cardiac failure, hydrops, ventriculomegaly and other brain pathology are associated with an adverse perinatal outcome. Yuval et al suggested prognostic indices for neonates complicated with Galen’s aneurysm.⁹ The presence of hydrops and brain injury carry a grave prognosis while cardiomegaly puts the fetus at moderate risk. The prognosis could be influenced by the angioarchitecture of the aneurysm. Multiple feeding vessels and an abnormally dilated venous drainage were scored as moderate fetal risks, which are prone to heart failure. The case reported here carried a grim prognosis with abnormally dilated venous drainage (straight sinus), cardiomegaly, congestive heart failure and hydrops fetalis, and therefore, a decision to not intervene was taken.

The prognosis in prenatally detected cases has been uniformly poor, having a much worse prognosis than those detected later in childhood. Though the prognostic indices for fetuses with aneurysm of the vein of Galen have not been established, ultrasonography with color Doppler can identify those fetuses that are likely to be lethal. In others, a prenatal diagnosis would ensure prompt postnatal treatment, thereby reducing the risk of neonatal cardiac failure and increasing the success of postnatal neurosurgical treatment.

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