Case Report

Vein of Galen Aneurysmal Malformation: Antenatal Diagnosis by Ultrasound and MRI

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Abstract: Vein of Galen aneurysmal malformation is a rare intracranial arteriovenous malformation. It accounts for up to 30% of intracranial vascular malformations in pediatric population and 1% of all pediatric congenital anomalies. It causes severe morbidity and mortality in neonates in the form of high output cardiac failure, nonimmune hydrops, hydrocephalus and intracranial hemorrhage. It is often diagnosed in postnatal period. Prenatal diagnosis of VGAMs may be accomplished with prenatal ultrasound (US) and fetal MRI which helps in early diagnosis and effective management.

Key Words: Vein of Galen, Aneurysmal malformation

Introduction:
Vein of Galen aneurysmal malformation is an arterio-venous fistula involving aneurysmal dilatation of the median prosencephalic vein of Markowski. Dilated arteries drain into a large midline venous pouch in the region of the quadrigeminal cistern. It develops during the 6th to 11th weeks of gestation and can be diagnosed prenatally by ultrasound but most often the VGAM is detected in the postnatal period.[1] Various treatment strategies are available ranging from conservative management, elective embolization and surgery.

The prenatal diagnosis is achieved through ultrasound, colour Doppler and MRI. Early detection is useful in planning treatment modality.

Case Report
A 31 years old gravida 2 para 1 presented for the first time for antenatal USG at 31 weeks of gestation with previous history of LSCS. Fetal brain on USG and colour doppler showed a midline tubular anechoic venous vascular structure posterior to the quadrigeminal cistern with multiple anechoic vascular structures in the basal ganglia region bilaterally giving arterial and venous type of flow. There was hydrocephalus seen as dilated lateral ventricles and third ventricle. Fetus had anasarca, hepatomegaly, cardiomegaly and polyhydramnios.

Fetal ECHO showed left ventricular hypertrophy with rightward deviated inter-atrial septum. Lower segment uterine scar was thin and irregular. Fetal MRI showed midline flow void representing VGAM with multiple small flow voids in the bilateral basal ganglia region representing arterio-venous fistulae with non-communication type of hydrocephalus on T1 and T2 Weighted sequences.

Emergency LSCS was done at 34 weeks of gestation as patient went into labour. The fetus showed anasarca with signs of cardiac failure and died within 1 hour of birth.

Fig 1: Axial ultrasound and doppler images showing anechoic midline tubular structure showing colour flow on Doppler
CT demonstrates a well representing the dilated hulla G, Pearl M, et al. of Galen -ires emergent endovascular led assessment of shunt AM most.

Prenatal diagnosis of VGAMs may be undertaken as early as the second trimester of intrauterine life by sonography. Prenatal diagnosis of VGAM may be made as early as the second trimester of intrauterine life by sonography. MRI: Fetal MRI is increasingly used for confirming the diagnosis suggested by ultrasound. MRI and MRA/MRV may illustrate the size and configuration of the venous sac and provide an initial estimate on the number and type of arterial feeders involved. It also provides detailed evaluation of the brain parenchyma and other associated anomalies.

Digital Subtraction Angiography (DSA): DSA remains the gold standard technique for the assessment of the intracranial vasculature. It is the only available modality that provides a detailed evaluation of the VGAM angioarchitecture necessary for endovascular treatment. It also helps in knowing the morphology of the fistulae and detailed assessment of shunt hemodynamics in a neonate[3].

Management

Antenatal management: Cardiac failure and severe cerebral damage in the antenatal period are appropriate indications for termination of the pregnancy[5]. However not all cases are indication of termination of pregnancy. Neonatal management: Ideally the initial treatment of VGAM is conservative[7]. There are 3 management categories for neonates with a VGAM[5].

1. Those who are not offered endovascular treatment: In asymptomatic patients
2. Those in whom endovascular treatment can be deferred: Patients with mild cardiac overload which can be medically managed until embolization is performed at 4 to 5 months of age
3. Those who require emergent endovascular intervention: A subset of patients requires emergent endovascular embolization based on cardiac, cerebral, respiratory, hepatic and renal functions.

References

2. Pearl M, Gregg L, Gandhi D. Cerebral Venous Development in Relation to Developmental Venous Anomalies and Vein of Galen Aneurysmal Malformations. Semin Ultrasound CT MRI 32:252-263

Discussion:
Vein of Galen aneurysmal malformation was first described by Steinhel in 1895. It is a rare congenital cerebral arteriovenous abnormality with an incidence of 1:25000.[2] The hallmark feature of a VGAM is the presence of one or more arteriovenous shunts draining into a dilated median cerebral venous collector. This midline venous structure corresponds to a persistent embryonic channel, the median prosencephalic vein, which normally regresses with the development of the internal cerebral veins.[3]

Classification
Vein of Galen Malformations are divided into following types[4]:

1. Vein of Galen Aneurysmal Malformation (VGAM): VGAM is a choroidal type of AVM supplied by the choroidal arteries. The choroidal shunt drains into dilated midline median prosencephalic vein. Internal cerebral veins do not form and the thalamostrate veins drain into the posterior and inferior thalamic (diencephalic) veins and secondarily join the anterior confluence, a subtemporal vein or the lateral mesencephalic vein to open into the superio.

2. Vein of Galen Aneurysmal Dilatation: Vein of Galen aneurysmal dilatation correspond to AVMs localized in the subpial space supra or infratentorially, which drains into one of the usual tributaries of the vein of Galen and result in its overload and dilatation. The prenatal diagnosis of a vein of Galen dilatation is uncommon as it is found in older children.

3. Vein Of Galen Varix: This lesion refers to a varicose dilatation of the vein of Galen without an underlying arteriovenous shunt and drains the normal brain parenchyma.

4. Dural Vein Of Galen Dilatation: These lesions occur almost exclusively in adults. These constitute a group of acquired dural lesions that develop in the wall of the vein of Galen or at the venous-sinus junction.

Clinical presentation: Patients with a VGAM most commonly present with cardiac and neurologic complications. The clinical presentation depends on the age of presentation[5]. Neonates tend to present with high-output cardiac failure, pulmonary hypertension and in more severe cases multiorgan system failure. Infants commonly present with hydrocephalus, seizures, or neurocognitive delay. Older children and adults present with headaches or intracranial hemorrhage.

Radiological Investigations:

Prenatal USG: Prenatal diagnosis of VGAMs may be accomplished with prenatal US and fetal MRI[3]. Most diagnoses by ultrasound occur in the third trimester as sufficient dilatation of the vein does not occur till that time. Grayscale US imaging detects a midline tubular anechoic structure superior to the thalamus, representing the dilated median prosencephalic vein. Use of color and power Doppler sonography facilitates the diagnosis, documenting the vascular nature of the lesion and differentiating it from other cystic lesions of the fetal brain[3].

Neurosonogram: A transfonatelle ultrasound can be performed to assess VGAM, the brain parenchyma and ventricular size in neonates[5].

Computed tomography (CT): CT demonstrates a well-defined, multilobulated, intensely enhancing lesion, located within the cistern of velum interpositum. Dilatation of the ventricular system, periventricular white matter hypodensities, as well as diffuse cerebral atrophy are the commonly associated findings[6].

MRI: Fetal MRI is increasingly used for confirming the diagnosis suggested by ultrasound. MRI and MRA/MRV may illustrate the size and configuration of the venous sac and provide an initial estimate on the number and type of arterial feeders involved. It also provides detailed evaluation of the brain parenchyma and other associated anomalies.

Radiography: X ray skull may demonstrate a rim of calcification within the wall of the aneurysmal sac. Chest radiographs may reveal features of congestive heart failure[6].

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