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Vein of Galen Malformation- A Unique Congenital Malformation

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Abstract: ***Vein of Galen malformation (VGM) is a congenital arterio-venous fistula in the brain associated with cardiovascular and neurological complications leading to high morbidity and mortality. It may present from the newborn period to late childhood. We report a case of five month old male child who was admitted with increased head size since the age of one month, history of increased work of breathing since two days. Antifailure treatment was given. On correlating history, clinical examination, ultrasound of skull and two dimensional echo cardiography (2D-echo) the diagnosis of VGM with hydrocephalus with Atrial septal defect (ASD) with partial anomalous pulmonary venous drainage (PAPVD) was made. After stabilisation the child, embolisation was planned. This case deals with detection and management of VGM with hydrocephalus patient.***

Keywords: Vein of Galen malformation, hydrocephalus.

Disclaimer: The case was taken after the due consent of patient's mother.

Work attributed to: Department of Pediatrics Dr.D.Y.Patil Medical College Pune, Maharashtra, India.

1. Introduction

VGM is a congenital arterio-venous fistula in the brain associated with cardiovascular and neurological complications leading to high morbidity and mortality. It may present from the newborn period to late childhood.[1] VGM constitute 1% of all intracranial vascular malformation and 30% of vascular malformations presenting in pediatric age group.[2] Malformation occurs due to cerebral arteriovenous fistula of the median prosencephalic vein (MPV) which is a precursor of the vein of Galen occurring at 6-11 weeks gestation. The MPV fails to regress and becomes aneurysmal. It drains via the straight sinus and vein of Galen is not formed. It is the most common antenatally diagnosed intracranial vascular malformation. VGM result in high-output congestive cardiac failure and may present with developmental delay, hydrocephalus and seizures. Main goal of treatment in newborn infants involves initial cardiovascular stabilization together with endovascular embolization of feeding arteries and draining veins.[3] Ventricular shunting may worsen the cerebral venous hypertension and is associated with high risk of complication so it is avoided before elimination of the arteriovenous shunt.[4] Here, we present a five month male child with VGM who was not diagnosed antenatally and first presented at five month of age with hydrocephalus and cardiac failure. He was successfully treated with medical management and after stabilisation embolisation was planned. We have chosen this case to show that late presentation of VGM case have a good prognosis. The treatment policy should be planned by the entire group of physicians caring for these children including paediatrician, pediatric cardiologists, neuroradiologists and neurosurgeons.

2. Case Report

A five month old male child was brought by parents with complain of increased head size since the age of one month and not able to hold head without support. He had history of increased work of breathing since two days. Only one antenatal ultrsonography was done in first trimester which does not revealed any abnormality. The child was delivered at home. On admission, he had tachypnea, tachycardia, oxygen saturation was 94% and hypotension was present. Fluid resuscitation was done and child was started on dopamine. In view of sepsis antibiotic cover was given . Antifailure treatment (Furosemide, Spironolactone, Enalapril) was started. On examination of skull, occipito-frontal diameter was increased and skull veins were found to be dilated. Cranial sutures were widely separated. Trans-illumination test was positive. Acetazolamide was started. Chest radiograph was normal. Cranial ultrasound demonstrated a large vein of Galen arteriovenous malformation. Magnetic resonance venography demonstrated vein of galen malformation having communication with straight sinus with obstructive hydrocephalus and hence the diagnosis was confirmed. Child responded to medical treatment. Two dimensional echocardiography was done which revealed Bidirectional Atrial septal defect (ASD) with partial anomalous pulmonary venous drainage (PAPVD). Neurosurgeon and neuroradiologist were consulted for hydrocephalus with VGM and embolisation was planned.

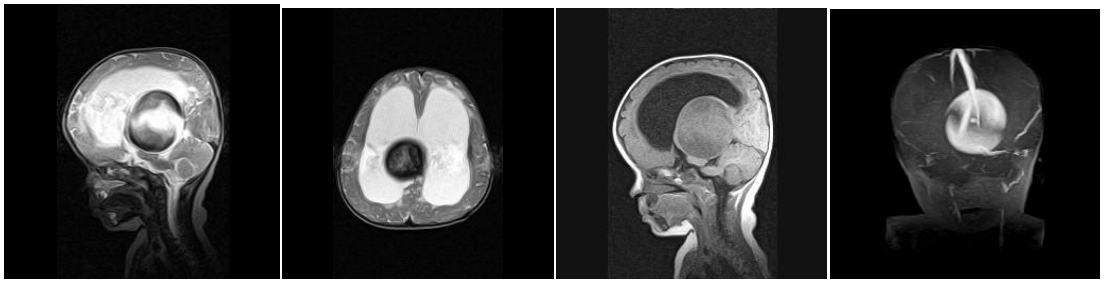

Figure- MR venography images demonstrating large well defined rounded heterogenus lesion showing avid post contrast enhancement, lying posterior to cerebral aqueduct suggestive of vein of galen malformation having communication with straight sinus. Associated obstructive hydrocephalus is also present.

3. Discussion

VGM is a rare congenital vascular malformation located in the midline in choroidal fissure. It consists of multiple feeding arteries, principally the anterior and posterior choroidal arteries and anterior cerebral artery, draining directly into an enlarged venous pouch. Malformation occurs due to cerebral arteriovenous fistula of the MPV which is a precursor of the vein of Galen occurring at 6-11 weeks gestation. The MPV fails to regress and becomes aneurysmal. VGM are occasionally detected on antenatal ultrasonography from about 25 weeks gestation as a cystic midline brain lesions, and colour Doppler then suggests a VGM.[5] More commonly they are diagnosed after birth. Larger shunts show rapid deterioration with cardiac failure leading to multiorgan failure. A smaller shunt present later in life with mild cardiac failure and failure to thrive.[5]

Clinical manifestation of VGM varies according to age group. In neonates, as they have multiple fistulas, upto 25% of their cardiac output passes through the fistulas causing high output cardiac failure. Their manifestations can range from asymptomatic cardiomegaly to severe cardiac failure.[6]

Infants and children usually have single fistula with smaller shunt. So, cardiac manifestations are absent or very mild. They present with hydrocephalus. In long standing cerebral venous hypertension, may present with failure to thrive and delayed milestones.[7]

Older children usually have low-flow fistulae, so they present with headache and seizures. Some may also present with developmental delay, focal neurological deficits, epistaxis.[7]

Foe diagnosis, antenatal ultrasonography should be done which shows cystic midline brain lesions. Colour flow Doppler is also used. Antenatal magnetic resonance imaging (MRI) helps to confirm the diagnosis and assesses any pre-existing damage to the brain. Cardiac evaluation in the form of chest radiograph and 2D-echo is used. If cardiac evaluation is normal, transfontanelle ultrasound will detect the VGM. MRI or computed tomography helps in the assessment of arteriovenous shunt. Angiography is best performed at the time of embolisation.

The initial treatment of VGM is conservative until 5 or 6 months of age with regular outpatient assessment. If the infant have seizures, failure to thrive and worsening cardiac failure, embolisation of feeding vessel can be performed earlier. Surgery has little role in treatment of VGM and they are associated with very high mortality or severe morbidity. Emergency embolization is indicated when cardiac failure is not responding to medical treatment. The goal of therapy in them is to arrest the congestive cardiac failure rather than to achieve complete obliteration of the shunt. Ventricular shunting may worsen the cerebral venous hypertension and is associated with high risk of complication so it is avoided even if hydrocephalus is present before elimination of the arteriovenous shunt.[4] Shunting of the ventricles prior to embolisation may accelerate progressive atrophy of brain (melting brain syndrome).[5] Radiosurgery has been tried, but is limited to patients who are not candidates for other treatment modalities.[8] Complication associated with VGM are intracranial haemorrhages.[9] Untreated VGM have a poor prognosis and are always fatal, but it is equally important to recognise that treatment may be inappropriate in few cases. If there is evidence of pre-existing brain damage, progressive atrophy, parenchymal calcification or severe multiorgan failure, a poor outcome is inevitable.

A team approach with the help of neonatologist, paediatric cardiologist, neurosurgeon and interventional neuroradiologist is important for successful management of VGM. Thus, with advances in imaging technology, cardiac care, interventional neuroradiology and with better post-procedure intensive care, these once non-treatable conditions are now potentially curable, with excellent clinical results, low complication rate and low morbidity and mortality. In the future, with the establishment of more centers with such facilities in the country, the treatment of VGM will be within the reach of all patients.

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