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Paediatrics

DETECTION OF VEIN OF GALEN MALFORMATION IN A NEWBORN- A CASE REPORT

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ABSTRACT Vein of Galen aneurysmal malformation (VGAM) is a rare congenital anomaly and amongst rare cause of congestive heart failure in the neonatal period carrying high mortality rate. The diagnosis may remain elusive as the clinical picture often mimics congenital cardiac malformation. Patients usually undergo high-risk procedures such as cardiac catheterization to establish a diagnosis. Often, these investigations are inconclusive and can delay correct diagnosis. This study highlights the role of antenatal and follow up Doppler USG which can lead to early diagnosis on the basis of characteristic Doppler findings in a resource limited peripheral hospital. Various therapeutic options for a VGAM include no treatment, endovascular treatment, open surgery and stereotactic radiosurgery. Timely diagnosis and appropriate therapy especially endovascular management can result in fairly good outcome in these children.

KEYWORDS : Vein of Galen, Transcranial Ultrasound

CASE REPORT

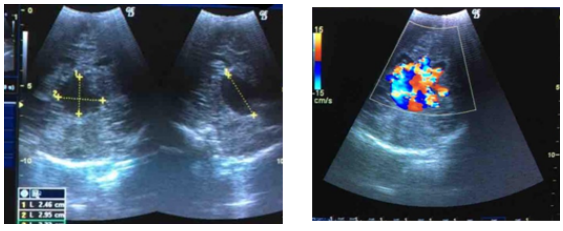
An one day old term baby was brought to this hospital at 24 hrs of life with complaints of poor feeding and repeated vomitings. The baby was delivered by LSCS at another nursing home.

On examination the baby weighed 3.5 kgs, was pale with cold peripheries, tachypnea, subcostal retractions and SPO2 of 86% at room air which improved to 96% with oxygen therapy. Baby was admitted in NICU as a case of respiratory distress with possibility of early onset sepsis and was started empirically on I/V antibiotics and other supportive treatment.

On investigations sepsis screen was negative, ABG showed metabolic acidosis with ph 7.26 and lactate 9.84mmol/l. Chest xray showed cardiomegaly but on examination there was no cardiac murmur. Abdominal examination showed hepatomegaly. Based on above findings a working diagnosis of Congestive Heart Failure was made with acyanotic heart disease and 2D-Echo was contemplated.

On checking antenatal records we found mention of a foetal cyst in midline on antenatal USG. Transcranial USG was done at our hospital which revealed a well defined midline anechoic lesion measuring 2.4.x2.9x.3.3 cm, posterior to 3rd ventricle. Doppler showed significant high velocity, low resistance flow within the lesion with communicating vascular channel posteriorly towards venous sinuses . The diagnosis of vein of Galen aneurysm was made. USG abdomen revealed mild ascitis secondary to CCF. Baby was transferred to higher centre for further imaging workup and management.

This case report shows the classical presentation of VOGM as congestive cardiac failure in neonatal period which was diagnosed at peripheral hospital without advanced imaging facilities within 48 hrs of life and within 24hrs of hospitalization.



(Fig 1) showing anechoic midline lesion post to 3rd ventricle

DISCUSSION

Vein of Galen aneurysmal malformation (VOGAM) is a rare congenital anomaly, accounting for less than 1% of all cerebral vascular malformations and approx. 30% of symptomatic vascular malformations in children. [1-4] It is a rare cause of CHF in the neonatal period and carries a high mortality rate. The diagnosis remains elusive and difficult as the clinical picture often presents with symptoms suggestive of a congenital cardiac disease. [1-3].This often leads to high-risk procedures such as cardiac catheterization to establish a diagnosis causing delay in correct diagnosis.

During the development of the cerebral vascular system, the developing telencephalon is supplied by multiple choroidal arteries that drain via a single transient midline vein in the roof of the diencephalon, the Median Prosencephalic Vein (MPV) of Markowski. Normally as internal cerebral veins develop, the MPV (precursor of the vein of Galen) regresses [5,6]. In a VGAM, a high-flow fistula between deep choroidal arteries and a persistent MPV prevents formation of the definitive vein of Galen. The presence of these AV shunts keeps the MPV patent and causes flow-related aneurysmal dilatation of this primitive vein. This leads to formation of a midline venous pouch behind the third ventricle. The drainage is usually into an enlarged superior sagittal sinus via an embryonic falcine sinus. Reduced outflow causes increased venous hypertension increasing the risk of intracranial hemorrhage. Communicating hydrocephalus occurs as a result of impaired absorption of the CSF by the arachnoid villi.

The VOGAM is classified into mural and choroidal types. In the mural (simple) type, the direct high flow shunt is located within the wall. In the choroidal type (complex) type, there are arterial feeders interpositioned between the venous aneurysm. The mural types of VOGAM present later in infants with macrocephaly or failure to thrive . They may be associated with mild cardiac failure or cardiomegaly. However, the choroidal types usually cause heart failure in newborns. Imaging plays a crucial role in diagnosis of VOGAM. Antenatal ultrasound shows hypoechoic to mildly echogenic midline mass located behind the third ventricle. Color Doppler shows turbulent flow within. In our case the USG mentioned the presence of a midline lesion but no Doppler examination was done leading to a misdiagnosis in antenatal period. Subsequent post natal transcranial USG at our hospital lead to the diagnosis of VOGAM.

Other imaging modalities can further evaluate this anomaly. CT head will show a mildly hyperdense mass at the typical midline location showing uniform intense post contrast enhancement. Ancillary findings such variable encephalomalacia, hemorrhage and/or obstructive hydrocephalus can be assessed.

Fetal MRI shows serpentine "flow voids" corresponding to arterial feeders adjacent to the midline lesion. Thrombus of varying ages may be present lining the VGAM. MR can also identify important secondary complications of VGAM, such as hemispheric white matter injuries and progressive heart failure with development of fetal hydrops.

Angiography delineates the angioarchitecture of the anomaly remains the mainstay of diagnosis as well as pre-interventional workup. Therapeutic options available for a VOGAM include no treatment, endovascular treatment, open surgery, and stereotactic radiosurgery. The appropriate therapeutic option is chosen depending on patient's age, type of the lesion and its angioarchitecture. The ultimate goal is sufficient control of the malformation to allow normal brain maturation and development. Recent advances in the field of

intervention neuroradiology has changed the treatment and prognosis of children with VOGAM.[8-11] Appropriate and timely endovascular intervention can result in a good outcome in these children.

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