ANTENATALLY DIAGNOSED, VEIN OF GALEN MALFORMATION: A CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Published reports are reviewed relating to pediatric patients diagnosed with a vein of Galen malformation (VGM). We report a case of antenatally diagnosed VGM, including five patients from this institution; we reviewed the literature for antenatally diagnosed VGM.

Key words : *Vein of Galen, Pre-natal, case report*

INTRODUCTION:

Vein of Galen aneurysmal malformations (VGAM) are rare congenital vascular malformations characterised by shunting of the arterial flow into an enlarged cerebral vein dorsal to the tectum. Occurring during embryonic development, VGAM's are abnormal connections between arteries and the deep draining veins of the brain. VGAM's do not have capillaries, thus the blood flow can be extremely fast increasing the work of the heart. Incidence is < 1/25000 deliveries, male: female ratio 3:1.

Most of these malformations present in early childhood, often causing intractable CCF in the neonate, which is the most common cause of death (1, 2, 3). In less severe cases, a child may develop hydrocephalus because the enlarged malformation blocks the normal flow or absorption of cerebrospinal fluid. Seizures and other neurological signs are unusual. Tranarterial embolisation with liquid adhesive agents or microcoils is the treatment of choice. Surgery has very little role.

CASE REPORT:

CASE 1 : Antenatal ultrasound of a 26 year old lady at 36 wks of gestation detected VGAM. Baby was delivered by elective LSCS at 39 weeks. Baby did not require any resuscitative measures at birth, but within half an hour baby went into congestive cardiac failure. Hence baby was electively intubated, connected to ventilator and treatment of cardiac failure was started. Echocardiography done showed severe right heart and PA dilatation. Cranial angiogram was performed which showed VGAM – choroidal type with multiple choroidal feeders. In view of innumerable shunt and tortusity of feeders embolisation through arterial route was considered unsuitable and occluding the VGAM through venous route was considered. Treatment modality and prognosis was explained to the parents who declined further management. Baby died of intractable CCF within 24 hours of life.

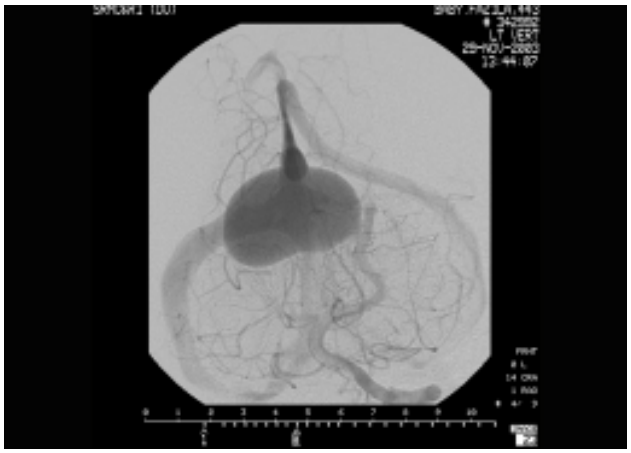

Fig.1. Cranial angiogram showing vain of galen malformation

CASE 2: Antenatally diagnosed VGAM was referred to our institution for further management. After birth baby was clinically stable hence, no intervention was required. Later at the age of 6 months, child came with rapidly increasing head circumference. Child was conscious with no focal neurological deficit, hence was taken up for AV fistula embolisation. Four vessel angiogram revealed VGAM with arterial feeders from numerous posterior choroidal arteries. Arterial feeders were embolised but child died on 2nd post-op day due to intracranial bleed.

CASE 3: Another child with antenatally diagnosed VGAM; no intervention was required at birth as child was clinically stable. Later embolisation was done electively at 11 months. Angiogram done showed mural type of VGAM with simple fistula between left posterior choroidal artery and aneurysmal sac. The child underwent successful embolisation. At follow-up, the child has no developmental delay or any neurological deficit.

DISCUSSION:

There are two basic types of vein of Galen malformation. In the first, single or multiple arteries drain directly into enlarged venous structures of the Galenic system. The most common anomaly is the singular multiple direct arterio-venous fistula between the choroidal and the quadrigeminal arteries and a median venous sac. In

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the second type, a parenchymal AVM is present usually in the thalamus or midbrain and its nidus has deep Galenic drainage. Diagnosis is made by transcranial ultrasound, cerebral angiography, MR and MR angiography (4). Before the era of sophisticated imaging technologies and endovascular treatment, VGAM was fatal in 90% of patients under 1 month of age and half of those between 1 month and 1 year (2). With transcatheter embolisation there is 70-80% survival among neonates and young infants and cure rates of approximately 50% (5, 6).

Neonatal Rating Score (7)

Lasjaunias and ter Brugge score is derived from measures of cardiac, cerebral, hepatic, renal, and respiratory function.

- < 8 results in a decision not to treat.
- 8-12 prompts emergency endovascular intervention.
- 12 recommends medical treatment alone and delayed embolization at 5 months.

NEONATAL RATING SCORE

Score	Cardiac	Cerebral	Respiratory	Renal	Hepatic
5	Normal	Normal	Normal	_____	_____
4	Non-treated overload	Infraclinical EEG anomalies	Tachypnoea Bottle finished	_____	_____
3	CCF stable with treatment	Non-convulsive CNS signs	Tachypnoea Bottle not finished	Normal	Normal
2	CCF unstable with treatment	Isolated convulsion	Assisted ventilation	Transient Anuria	Hepatomegaly normal function
1	Need ventilation	Seizure permanen CNS signs	Assisted ventilation FiO2 > 25%	Unstable diuresis with treatment	Moderate hepatic impairment
0	Resistant to Rx.	Coma	Assisted ventilation	Anuria	Coagulation disorder

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