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Review Article

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CASE SERIES ON VEIN OF GALEN ANEURYSMAL MALFORMATION IN NEONATES

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ABSTRACT

Vein of Galen malformation (VOGM) is one of the rare congenital vascular malformations, presenting in neonatal or early childhood with features of left to right shunt or high cardiac output failure, leading to both mortality and morbidity. VOGM is caused by maldevelopment of median prosencephalic vein of Markowski. We present case series of 4 full term infants (3 males and 1 female).

KEYWORDS: Vein of Galen malformation, congenital anomalies, neonates.

INTRODUCTION

VOGM is an infrequent congenital vascular malformation (1:25,000 births). Gender distribution of this malformation is equal and accounts for less than 1% of cerebral arteriovenous malformations in children.[1] Majority of VOGMs are identified in neonatal period with remainder are identified in early childhood. Steinheil first reported VOGM in 1895.[2] Majority of VOGMs become symptomatic in neonatal period with almost 100% mortality and morbidity.[3]VOGM develops at 6-11 weeks as a result of persistence of of the embryonic median prosencephalic vein of Markowski.[4] First attempt to treat VOGM started in last decade wherein bilateral carotid artery ligation was attempted in an infant. At present endovascular ligation is the preferred mode of treatment.

Case Series: We present case series of total 4 cases with 3 males and 1 female infant(Table 1). The diagnosis was made antenatally in 3 patients(between 30-34 weeks of gestation) and 4 days after birth in one of the patients. The clinical manifestations at birth were macrocephaly and intracranial bruits in all 4 patients, heart murmurs in 3 patients, 2 patients experienced convulsive episodes with subsequent development of Congestive heart failure(CHF) in all 4 cases. Out of 4 patients, 3 underwent multiple episodes of embolization(Fig.1). One of the patients did not undergo any type of endovascular treatment due to overwhelming financial burden.

Table 1: Characteristics of patients.

	Sex	Time of diagnosis	Hear murmur	Intracranial bruits	Macrocephaly and hydrocephalus	Heart Failure	Treatment(timing)
1	Male	Antenatal(30-34weeks)	Yes	Yes	Yes	Yes	Embolization (1 month post birth)
2	Male	Antenatal(30-34weeks)	Yes	Yes	Yes	Yes	Embolization(20 days post birth), 3 episodes
3	Male	Antenatal(30-34weeks)	No	Yes	Yes	Yes	Embolization(3 months post birth)
4	Female	Antenatal(30-34weeks)	Yes	Yes	Yes	Yes	No

VOGM is quite rare, but it should always be included into differential diagnosis of neonates/infants presenting with CHF with unknown etiology. MR angiography is the first line imaging test for diagnosis of VOGM. Embolization is the treatment of choice for VOGM with timing of embolization depends upon the clinical severity of the patients and the main indication for it is the CHF refractory to medical treatment. Size of the aneurysm and age of the child determines prognosis of this condition.

Figure 1: Percutaneous coil embolization of vein of Galen malformation.

DISCUSSION

The vein of Galen is present under the cerebral hemispheres and it drains the anterior and central regions of the brain into the sinuses of the posterior cerebral fossa. The precursor of VOG is median prosencephalic vein of Markowski which usually regresses during the 11th week of gestation, and by 3 month of gestation, its posterior part joins the internal cerebral veins and basal veins to form the VOG. An arteriovenous shunt between this vein and surrounding arteries induces the hemodynamic changes[5]. Hydrocephalus is the most common finding seen associated with VOGM.[5] The development of a VOGM may be an acquired event between the 6th and 11th week of gestation. Based on the complexity, type of supplying arteries, location of fistula and degree of venous ectasia VOGMs have been classified in many ways.[6] Lasjaunias classified VGAMs into two types, namely the choroidal and mural type.[7]

Table 2: Classification of vein of Galen aneurysmal malformations.

Classification system	
Litvak	
Category A	Aneurysms of the great vein of Galen
Category B	Racemorse conglomeration of blood vessels in the cerebral structures
Category C	Transitional types of midline AV shunts
Lasjaunias	
Type I	Choroidal type
Type II	Mural type
Yasargil	
Type I	Pure AVF between leptomeningeal arteries and feeders from P3, segments of posterior cerebral arteries and vein of Galen
Type II	Feeders from the thalamo-perforating vessels and from P1 and P2 segments of the posterior cerebral arteries
Type III	Mixture of type I and II
Type IV	
IV A	Aneurysmal dilation of the vein of Galen resulting from shunting from an adjacent thalamic AVM
IV B	Similar to type IV A with the AVM being mesencephalic instead of thalamic
IV C	Thalamomesencephalic or mesodiencephalic plexiform malformation along with an adjacent and separate cisternal AVF to the vein of Galen
Secondary enlargement of vein of Galen	
Vein of Galen dilation	Malformations that drain pial or dural shunts into the true vein of Galen or its tributary associated with the dilation of the vein of Galen
Vein of Galen varix	Dilation of the vein of Galen in the absence of AV shunt

Because of natural history of disease progression, prenatal diagnosis of VOGM is generally not done before third trimester. Progressive foetal cardiac dysfunction implies that the high flow lesion may not respond to treatment.[8] Advanced stage of this malformation is called as Melting brain syndrome wherein cerebral arterial blood flow is gradually decreased because of increased venous hypertension with progressive loss of white matter. Fetal MRI is now the preferred modality for diagnosing fetal central nervous system (CNS) abnormalities because of its advantages over ultrasonography.[9] Prenatal MRI and MR angiography can identify the prognostic factors (cardiac failure, polyhydramnios, pericardial and pleural effusion, ascites, fetal hydrops, and brain injury) that affect prenatal counselling and aid in delineating the blood supply to the lesion for planning postnatal management.[10] Yuval et al[11], while enumerating the prognostic features for VGAMs have suggested that neonates with cardiomegaly, dilated vena cava or jugular vein, retrograde aortic flow and multiple feeder arteries may have a better prognosis if there is immediate intervention.

In summary we want to conclude that, VOGM is rare but devastating AV malformation if left untreated. Prenatal diagnosis with antenatal ultrasound, fetal MRI, and fetal echocardiography provide an opportunity to plan the delivery of the foetus at a tertiary care centre where immediate and definitive care can be provided by a multidisciplinary team. Endovascular treatment has had a significant impact in outcome.

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