A case of vein of Galen aneurysmal malformation in a preterm baby

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Introduction

Vein of Galen aneurysmal malformation (VGAM), also termed as median prosencephalic arteriovenous fistula is a rare congenital cerebral arteriovenous abnormality with an incidence of 1: 25,000[1]. It is thought to be due to exposure to teratogenic insults during 6-11 weeks of gestation[2]. It usually presents in the neonatal period as macrocephaly, features of left to right shunt and high output cardiac failure. We present a preterm baby presenting at 2 months of age with rapidly rising occipitofrontal circumference (OFC) with widely separated sutures and large fontanelles who is having a VGAM.

Case report

A 55 day old baby boy was reviewed at the multidisciplinary clinic due to marginal prematurity (36 weeks) and low birth weight (1.09kg). He was found to have large anterior and posterior fontanelles communicating with each other with widely separated cranial sutures. His occipito-frontal circumference (OFC) on presentation was 34 cm (on -3SD), which had risen from the birth OFC of 29 cm. Baby's length was far below (-3SD). Tone of the limbs was high with normal limb reflexes. Fixing was present. There was no dependent oedema and no lesions suggestive of cutaneous capillary malformation. His pulse rate was 160 bpm with normal volume. The pulse pressure and the rest of the cardiovascular system examination were normal. Neither features of heart failure nor cranial bruits were detected.

The baby was born to elderly non-consanguineous parents by emergency caesarean section due to absent diastolic flow. Baby did not need any resuscitation at birth, but was treated for presumed sepsis and jaundice. Antenatal period was complicated with pregnancy induced hypertension. Mother denied any exposure to teratogens. Antenatal anomaly scans did not reveal any abnormality. A right sided vitreous hemorrhage was found on routine ophthalmological evaluation for retinopathy of prematurity. No other bleeding manifestations were noted. The initial ultrasound scan (USS) of the brain, done on the 10th day of life, did not reveal any abnormality. ToRCH screening was negative. The baby was breast fed with good weight gain until the current presentation. There was no history of any skin lesions or vascular malformation in the family.

USS of the brain, done during the current presentation, showed a vein of Galen aneurysm without hydrocephalus and midline shift. No intra-cranial bleeding was noted and the rest of the brain was structurally normal. 2D echocardiogram revealed a small patent foramen ovale with good ventricular function. Magnetic resonance imaging (MRI) of brain with angiography showed a well-defined structure in the quadrigerminal cistern in midline measuring 1.4cm (trans) ×1.2cm (AP) × 1.1cm (CC) in size (Figures 1 & 2) with signal voids in both T1W and T2W images corresponding to the site of the expected vein of Galen, in favour of a VGAM- type 1 (Yasargil classification)[3].

The clotting profile was normal. His *Bicêtre*-score was >12/25, which allocates him for a better prognosis[4]. Baby was referred for neurosurgical intervention and planned for digital substraction angiography and embolization.

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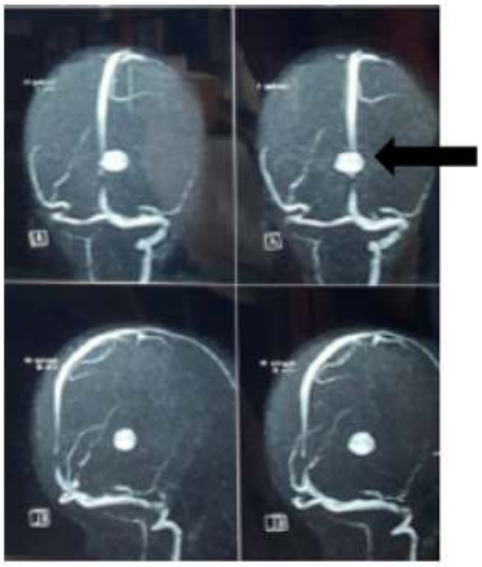

Figure 1: ***MR venogram of the baby (Arrow indicates VGAM)***

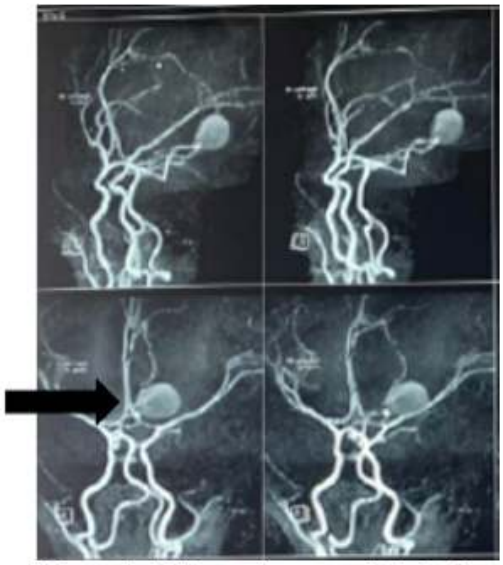

Figure 2: ***MR arteriogram of the baby (Arrow indicates VGAM)***

Discussion

The great vein of Galen (VG) is formed by the union of the two internal cerebral veins at the caudal part of the tela choroidea of the third ventricle and basal veins of Rosenthal. It passes caudally and dorsally to merge with the inferior sagittal sinus to form the straight sinus[5]. Persistence of embryonic vein of Markowski results in aneurysmal dilatation of the VG with arteriovenous shunting. VGAM, first described by Steinhel in 1895, has a quite unclear aetiology[1]. It is the most common antenatally diagnosed vascular malformation and has a male predilection[6].

VGAM mainly presents during the first 28 days of life but can present later as in our patient[7]. A retrospective study by McSweeney *et al* showed that the age of presentation in the non-neonatal period ranged from 11 weeks to 18 months, 3 months being the median age of presentation[7]. VGAM results in high output cardiac failure in about 95% and hydrocephalus, subarachnoid or intraventricular hemorrhage and seizures in 5% of cases, which our patient need to be followed up for[7]. The clinical manifestations are diverse and vary according to the age and the angioarchitecture of the malformation. During the neonatal period, the choroidal type of VGAM, which is the commonest, manifests with volume overload features as high output cardiac failure, pulmonary hypertension, respiratory distress, pulmonary oedema or even multiorgan failure[8]. Infants with mural type of VGAM usually present with hydrocephalus, macrocrania, developmental delay and rarely with epilepsy[8]. Cardiac failure presenting beyond the neonatal period is usually mild to moderate and can be managed medically[8]. Condition can be associated with capillary malformation-arteriovenous malformation syndrome and hereditary haemorrhagic telangiectasia in a subgroup due to *RASA1* and *ENG, ACVRL1* gene mutations respectively[9,10].

Diagnosis can be made antenatally or mostly after birth by USS or MRI of the brain. Since VGAM demonstrates intralesional flow, it can be confidently differentiated from other mimicking intracranial lesions such as arachnoid cysts, porencephalic cysts, cavum septum pellucidum, and quadrigeminal cistern by imaging[3]. Dilated feeding and draining vessels appear as flow-voids on MRI-T2 images[3]. The Yasargil classification and Lasjaunias classification are the two common currently used systems for classifying VGAM[3]. The Yasargil classification consists of type I to IV, whereas the Lasjaunias classification includes choroidal and mural types[3]. Type-I in Yasargil classification, which was found in our patient implies a small cisternal fistula between the VG and either the pericallosal arteries or posterior cerebral artery[3].

Treatment options are embolization and surgical correction of the lesion. Timing and modality of treatment depends on the signs and symptoms. The *Bicêtre*-score, which takes into consideration the cardiac, neurological, respiratory, hepatic and renal state of the patient, is helpful to evaluate the therapeutic management[4]. In neonates with a stable cardiovascular and neurologic status (*Bicêtre*-score>12), it is recommended to postpone intervention until 5-6 months of age. In a minority, especially lesions with a low flow state resolve spontaneously with thrombosis[10]. The safest and

most popular technique is transarterial embolization owing to its low risk profile whereas transvenous embolization and surgical treatment are associated with a higher risk of complications as well as high mortality[7,12,13].

Fetuses with hydrops and neonates with severe cardiac failure usually have a higher mortality[7]. Survivors have a varying prognosis from normality to severe neurodevelopmental sequelae at follow up[4,7,12]. Patients who have undergone endovascular embolization are reported to have a better neurocognitive outcome[3,4,13].

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