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Vein of Galen Malformations- An Insight to Undo the 'Gordian Knot' of Surgeons by Neuro-Interventional Radiologist

Case Report

Avi Agrawal, Chandrika Makker, Kritika Agrawal* and Shankey Garg*

Department of Radiology, MMIMSR, Mullana, India

***Corresponding author:** Kritika Agrawal and Shankey Garg, Department of Radiology, MMIMSR, Mullana, India

Email: doctorkritika3@gmail.com

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Abstract

Vein of Galen malformations are rare and unique congenital malformations of the cerebral vasculature. It is due to direct arterio-venous fistulous connection of misshapen arteries in brain with a persistent embryonic precursor of Vein of Galen (Median Prosencephalic vein of Markowski) instead of connecting with the capillaries. We are reporting 2 cases of VGAM. The first case is of a middle-aged man with intractable occipital headache with features of VOGM. The second case demonstrates antenatal diagnosis of VAGM in association with ventriculomegaly and cardiomegaly in routine antenatal scan at 30 weeks of gestation in a twin pregnancy. Considering the rarity of these lesions, there are very few studies that have been able to adequately diagnose VOGM. Thus, making this study radically essential for continuing developments in the diagnostic aspects of diagnosing and managing these lesions.

Keywords: Vein of Galen Malformation; Arterio-Venous Fistula; Median Prosencephalic vein of Markowski; Middle-aged man; Twin pregnancy; Congenital malformations; Gordian knot; Congestive cardiac failure; Cerebral Venous hypertension; Embolization; Misshapen arteries; Aneurysmal Dilatation; Endovascular therapy; Varix Aneurysm

Introduction

VGAMs are rare arterio-venous fistulas (AVFs) constituting <1% of all cerebral vascular malformations & represent 30% of symptomatic vascular malformations in the pediatric age group. These lesions are characterized by the presence of arterio-venous shunts keeping MPV patent and causing high flow/ pressure related aneurysmally dilated large midline venous pouch just behind the 3rd ventricle [1]. Steinheil in 1895, made the first reference to a Galenic malformation-referring to it as a 'varix aneurysm' [2].

Though these lesions are uncommon, they are of special interest to the interventional neuroradiologist because endovascular therapy [3] has proved itself to be an effective, and often the only safe therapeutic modality.

These lesions have been termed as the 'Gordian knot' of cerebrovascular surgery [4] with major problems being:

A) Deep seated, high-flow shunt

B) Poor myelination of the brain parenchyma

C) Tendency to tear easily on retraction

D) Ventricular shunting may worsen cerebral venous hypertension [5]

Case 1

A 35 year old male patient presented with headache in the occipital region for past five months. Patient was being evaluated for occipital headache when his vitals were within normal range and all routine lab reports were normal. Physical examination evaluated GCS to be 15/15 and the ophthalmological examination was also normal. There was no history of seizures, loss of consciousness, ENT bleed, vomiting or any other weakness.

CEMR brain with MRA scan was performed, which revealed a well-circumscribed T2W hypointense oval midline lesion measuring approximately 30mm X 25mm X 23mm dorsal to the third ventricle (Figure 1,2) in continuity with dilated straight sinus and dilated bilateral transverse sinuses s/o Vein of Galen aneurysmal malformation with mass effect on midbrain causing compression of aqueduct resulting in aqueductal stenosis and upstream hydrocephalus (Figure 3).

Multiple arterial feeders for VGAM originated from bilateral PCAs and basilar artery (Figure 4). It was drained by right internal cerebral vein and right basal vein of Rosenthal with evidence of multiple dilated venous channels from inferior sagittal sinus and superficial cortical veins (Figure 5).

Due to the high rate of morbidity associated with VOGM, the patient and his family members chose to continue on supportive treatment and observation of VOGM.

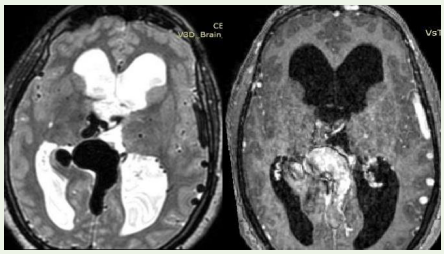

Figure 1: Aneurysmally dilated midline MPV, behind 3rd ventricle on axial A) CEMR T2W B) T1W POST GD

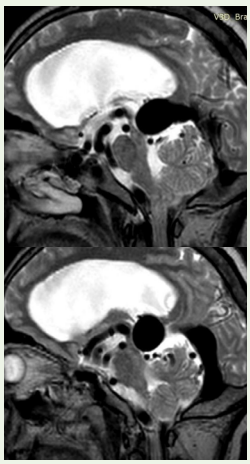

Figure 2: VOGM in midline dorsal to 3rd ventricle showing flow void on Sagittal T2W CEMR with multiple arterial feeders & dilated venous channels and sinuses at 2 different levels.

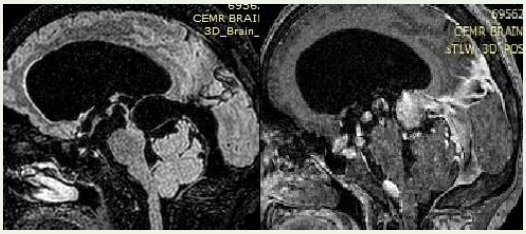

Figure 3: MPV draining into dilated straight sinus & torcula with aqueductal compression on midbrain & upstream hydrocephalus on sagittal A) FLAIR B) T1W POST GD

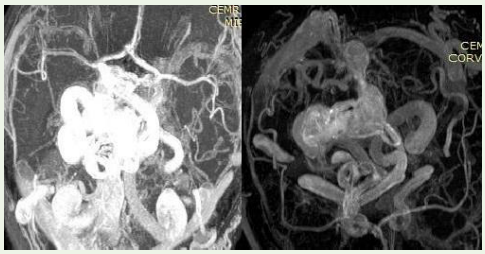

Figure 4: Multiple arterial feeders from B/L PCA & basilar artery attaching to AV aneurysm without any intervening nidus (capillaries) on MRA s3DI_MC A) MIP B) CORV.

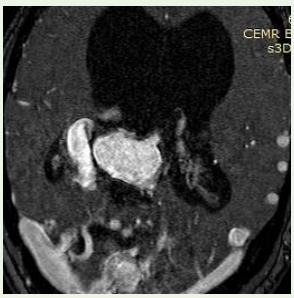

Figure 5: Prominent draining right basal vein of Rosenthal, B/L transverse and straight sinuses.

Case 2

A 28 year old pregnant woman presented for routine PNDT scan at 30 weeks of gestation with twin pregnancy. On transabdominal scan of fetus B, neuro-sonography revealed a tubular dilated anechoic structure coursing from the splenium of the corpus callosum towards the cisterna magna showing pulsatile and turbulent blood flow draining through dilated falcine sinus and torcular herophili into dilated transverse sinuses s/o Vein of Galen aneurysmal malformation with mild cerebral ventriculomegaly (Figure 6). Cross-section of fetal B's chest demonstrated slightly enlarged area of fetal heart in relation to area of chest s/o mild cardiomegaly. These findings of cerebral venous hypertension & cardiomegaly were consistent with VOGM for which counselling & neurosurgical consultation of parents was done.

Discussion

Development of the telencephalic choroid plexus is accompanied by simultaneous differentiation of a transient venous structure which drains the choroid plexuses & has been designated as MPV or the

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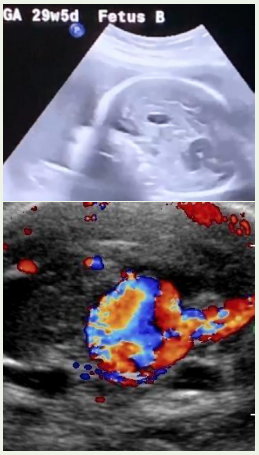


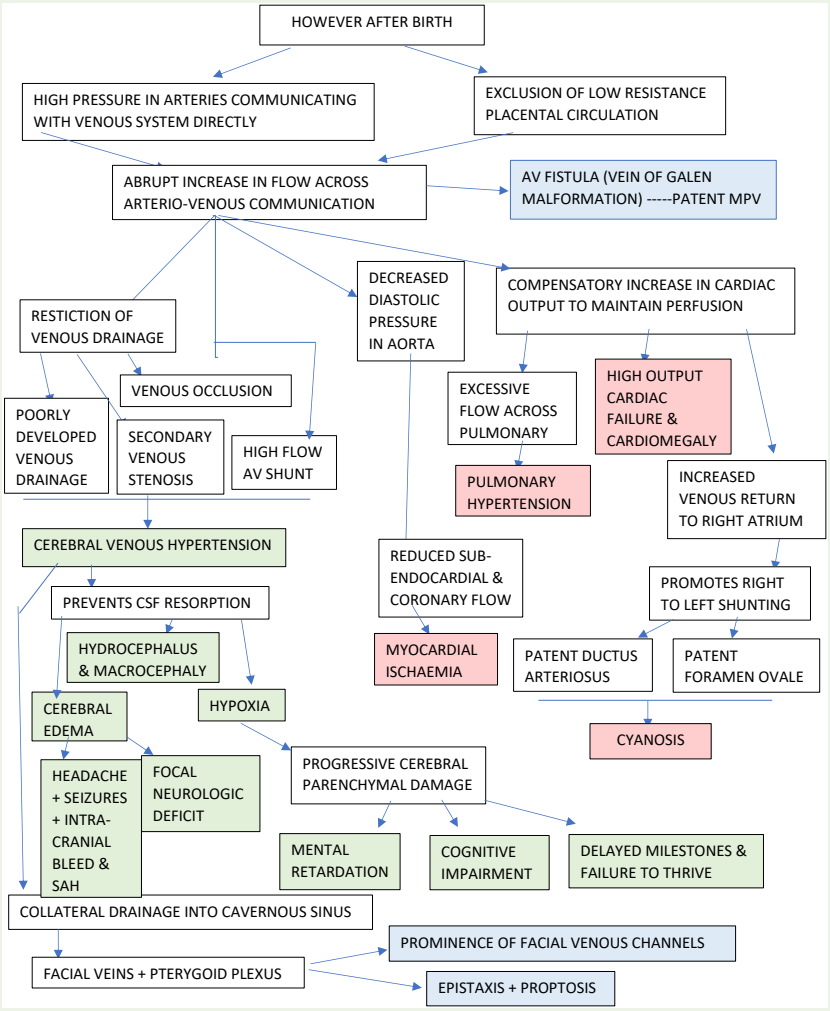
Figure 6: Tubular dilated anechoic structure draining blood from dilated falcine sinus into transverse sinus on PNDT scan A) 2D USG & B) Showing pulsatile & turbulent flow on colour doppler- features consistent with VOGM.

primitive internal cerebral vein. By the 11th week, there is formation of paired internal cerebral veins, which annex venous drainage of choroid plexuses [6].

This results in the regression of the MPV, except for its most caudal part, which joins the internal cerebral veins to form the vein of Galen.

Principal feeders of the malformation are those that normally supply the tela choroidea and the quadrigeminal plate. MPV, which drains the shunt, lacks a fibrous wall and lies free in subarachnoid space; therefore, it balloons out to a large size [7].

Prenatal ultrasound aids in identification of VGAM's usually in third trimester and its differentiation from other nonvascular space- occupying lesions along with assessing the status of the fetal cardiovascular system. It demonstrates the sonolucent venous sac as a mass located posterior to the third ventricle with pulsatile flow differentiating VOGMs from other midline cystic lesions [8].



Flow chart: PATHO-PHYSIOLOGY: Normally, the developing internal cerebral veins annex drainage of fetal choroid plexus and MPV regresses

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Axial Brain CT scan demonstrates a well- defined, multilobulated, intensely enhancing lesion within the cistern of velum interpositum. Diffuse cerebral atrophy, periventricular white matter hypodensities & dilated ventricular system are associated with features of cerebral parenchymal damage.

Angiography remains the gold standard for evaluation of small feeders supplying the fistula, as well as the dynamic aspects of venous drainage of the normal brain and AV shunt [9].

MRI is the modality of choice for VOGMs, demonstrating the location of fistula, presence of any nidus and relationship between the different pathological arterial & venous components. MR angiography is used as a noninvasive alternative to diagnostic angiographic studies [10].

VOGMs are also associated with the Turner syndrome, blue rubber bleb syndrome, supernumerary digits, hypospadias, transposition of great vessels, aortic stenosis and right-sided aortic arch [11].

Aggressive medical management postpones intervention until the child is about 5 - 6 months, when intervention is easier and safer [12]. Congestive cardiac failure (CCF) in a neonate that is refractory to medical treatment is an indication for emergency embolization. In neonates not presenting with cardiac failure, the aim would be to prevent consequences of cerebral venous hypertension and thus promote normal cerebral development [13]. AV fistulas are occluded on the arterial side, using embolic agents such as coils, cyanoacrylates and detachable balloons.

Transvenous and transtorcular coil embolization [14] of the venous sac is used to achieve flow reduction & is the technique of choice in patients with multiple fistulas, as it results in retrograde thrombosis obliterating the fistulas [15].

Conclusion

The varied and life-threatening clinical presentations and distinctive complex angioarchitecture of VOGM makes it essential for their early diagnosis; allowing a caring physician to understand their embryological and pathophysiological aspects. Management of these lesions – both in the prenatal/neonatal period and at the time of definitive intervention, is challenging. Thus, in near future and at current scenario; role of imaging is essential in making these lesions now potentially curable with better prognosis & low morbidity.

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