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Spontaneous Hemorrhagic Complication of Vein of Galen Aneurysmal Malformation Case Report

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Abstract: **Vein of Galen aneurysmal malformation (VGAM) is a rare congenital vascular malformations found in children. Chronic cerebral ischemia in VGAM is manifestation of long-term pial venous congestion secondary to venous outflow stenosis. Spontaneous hemorrhagic complication in a patient with VGAM is rare. We reporting a case of VGAM associated with intracranial hemorrhage (ICH), hydrodynamic disorder and cerebral ischemia, which was treated with multistage transarterial embolization.**

Keywords: **Vein of Galen, hydrodynamic disorder, vascular malformation in pediatric, galenic aneurysmal malformation.**

I. INTRODUCTION

Vein of Galen aneurysmal malformation (VGAM) is a rare intracranial vascular malformations constituting less then 1% of all intracranial vascular malformation. Most of these malformations present in neonatal period with congestive cardiac failure. Infants and older children usually present with hydrodynamic disorder or neurologic symptoms. Spontaneous intracranial hemorrhage is rare complication of VGAM. [1] Here we present our experience in treating VGAM with spontaneous hemorrhagic complications of the rerouted pial vein.

II. SUMMARY

A 4-year-old boy who is a known case of hydrocephalus secondary to cerebral aqueduct stenosis presented to our hospital with complaints of right upper limb jerking movement and uprolling of right eyeball for 1-day duration. He child was lethargic and drowsy. Examination revealed normotonia with hyporeflexea of both upper limbs and power of 3/5. Other systemic examinations as well as biochemical analysis were within normal limits. No evidence of skin lesions to suggest capillary malformation nor any history of skin lesions or vascular malformations in the family.

Brain CT scan showed hydrocephalus with bilateral subdural effusion as well as calcifications at the subcortical white matter in both frontal, left parieto-temporal lobes and striatum Fig. 1. During hospital stay, he had developed status epilepticus treated with phenytoin infusion. A repeated CT brain demonstrated basal ganglia hemorrhage with intraventricular extension, with a rounded hypodense structure within the hematoma, suggesting vascular aneurysm Fig. 2. MRI showed serpegenius flows void at quadrigeminal cistern with dilated median prosencephalic and falcine sinus in keeping with VGAM. The hematoma with dilated venous sac within is located away from malformation site Fig. 3. Cerebral angiogram demonstrated angioarchitectural characteristics of choroidal type VGAM. The arterial supply involves choroidal arteries including posterior choroidal arteries, anterior cerebral artery, (pericallosal) branch and thalamoperforating arteries. Venous drainage is toward dilated median prosencephalic vein into falcine sinus and subsequently into superior sagittal sinus. There was bilateral stenosis of the jugular vein with evidence venous rerouting mainly into pial veins in basal cistern and perimedullary veins Fig. 4. Three series of transarterial embolization were carried out during the first 3 weeks of presentation. In the first procedure we able to embolize via branches of posterior

choroidal artery and pericallosal artery using n-butyl-cynoacrylate (NBCA)mixed with iodized oil (1:1) as embolic agent and bringing a reduction of shunt flow of about 20 % Fig. 5. Stage embolization on the second and third sessions was successfully carried out within 3 weeks to further reduce the shunt. Subsequent transarterial embolization was done to further reduce the volume of arteriovenous shunting at 6 month and 8 month interval. The size of draining vein was significant reduced with reversal of arterial steal Fig. 4 and 5. The general condition of the patient improved and no breakthrough seizures were reported. Neurological assessment revealed improved limbs power and functions and resolving of hyporeflexia.

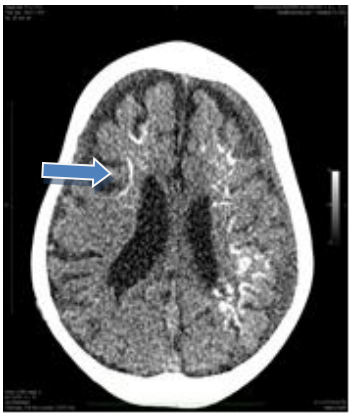


Fig. 1. Axial noncontrasted CT: Rubcortical calcification in both frontal, and left parieto-temporal lobes (arrows)

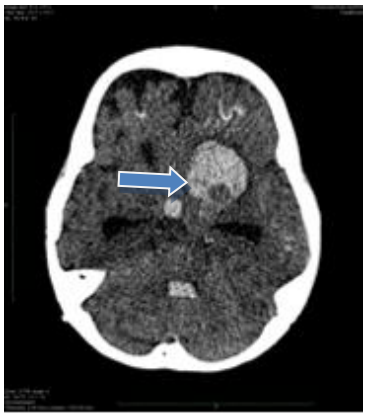


Fig. 2. Axial noncontrasted CT: Rounded hypodensity within intraparenchymal haemorrhage consistent with dilated venous sac (arrow).

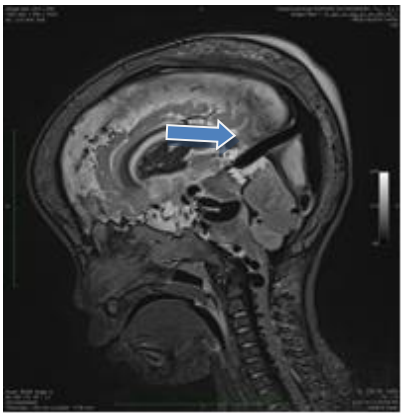
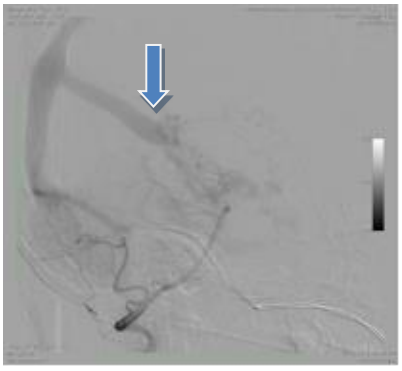


Fig. 3. Sagital T2W MRI: Dilated median prosencephalic vein and falcine sinus (arrows). Note the tortous and congested perimedullary veins (arrowhead)



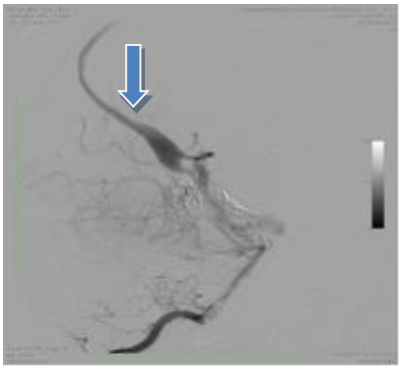


Fig. 4. Pre embolisation angiogram : Dilated median prosenchephalic vein (arrow) and Fig. 5. Post embolisation angiogram : Significant reduction of median prosenchephalic vein size in response to multistage transaretrial embolisation (arrow)

III. DISCUSSION

VGAM are choroidal arteriovenous malformation develops during early embryonic stage (6–11 weeks of fetal life). The vein of Galen is a short vascular trunk formed by the union of the two internal cerebral veins and the basal veins of Rosenthal. This congenital vascular malformation resulting in arteriovenous shunting into median prosencepalic vein of Markowski, which is the precursor of the vein of Galen. [2] There are certain angio-architectural characteristics of VGAM. They typically involve feeding artery of choroidal system and limbic arterial arch. Sump effect of the arteriovenous shunt may possibly recruit secondary supply from the subependymal and transcerebral branch of middle cerebral artery (MCA). [3] VGAM can be divided into mural or choroidal subtypes depending on the characteristics of the nidi. [4] The mural type consists in direct high-flow shunts located within the wall of the venous aneurysm. The choroidal

type, which corresponds to more primitive condition, involves the interposition of a network before opening into the large venous pouch. Hemorrhagic manifestation of ICH in patients with VGAM is rare. Berestein and Lasjaunias, two authors widely regarded as most experienced in the treatment of Galenic malformation found no incidence of ICH in their large series. [5] In contrast, hemorrhage is more common manifestation of the vein of galen aneurysmal dilatation (VGAD). [6] We postulate the cause of ICH in this patient is due to bilateral jugular stenosis causing outflow restriction and led to severe pial venous reflux. Cerebral calcifications in VGAM are objective anatomical evidence of venous hypoxia. They are typically located in subcortical white matter of transcerebral venous watershed area between the superficial and deep venous system. The development of jugular stenosis protects the heart but exposes the brain. It may, however also contribute to enlargement of the vein of Galen, with consequent aqueductal compression. [7] Among the various causes of hydrocephalus in VGAM, such as aqueductal compression or ex vacuo hydrocephalus, impaired CSF reabsorption secondary to venous hypertension is of particular importance.

The therapeutic strategy is to ensure normal development; therefore venous congestion must be reduced. Endovascular treatment is performed either by transarterial approach or the transvenous approach. The transfemoral transarterial embolization is our first treatment choice. We opted for stage transarterial embolization for this particular patient, as the risk of immediate and delayed hemorrhagic complications is significantly less. Transvenous embolization is technically less demanding but is reserved for situations where the transarterial method is not feasible. The venous method has an increased risk of venous infarction and hemorrhage.

IV. CONCLUSION

The prognosis of VGAM has been notably poor in the past but with the advent of endovascular therapy it is being rapidly reversed. Multistage transarterial embolization therapy has made a good outcome for our patient despite presence of ICH.

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