



Galen Vein Aneurysm: Problem of Management in an Under-Equipped Neurosurgical Environment about a Case and Review of the Literature

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Vein of GALEN aneurysmal malformation (VGAM) is a rare congenital disease caused by arteriovenous shunts between the choroidal arteries and the porencephalic ectatic vein. The diagnosis is often made in utero or during infancy, endovascular treatment remains the most suitable therapeutic means in a well-equipped environment. Here we report here the case of a patient complaining of headache for 1 year, and whose brain CT imaging showed the presence of Galen vein aneurysm with associated non-communicating hydrocephalus. In the absence of the appropriate technical platform, the placement of a ventriculoperitoneal shunt relieved our patient's symptoms.

Keywords: *Aneurysm; vein of galen aneurysm; ventriculoperitoneal shunt.*

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ABBREVIATIONS

VGAM : Vein of Galen Aneurysmal malformation
ETV : Endoscopic Third Ventriculostomy
CHF : Congestive Heart Failure
RVP : Rapid Ventricular Pacing.

1. INTRODUCTION

Veins of Galen aneurysmal malformation (VGAM) are congenital arteriovenous fistulas characterized by direct arterial drainage in a persistent midbrain vein of Markowski, the embryonic precursor of Galen's vein draining the primitive choroid plexus, phenomenon which generally occurs between the seventh and twelfth week of pregnancy [1] Steinheil is [2] often cited as the first to describe VGAM, in 1895, it is now believed to have a false vein of Galen malformation characterized by a parenchymal arteriovenous malformation flowing in a mature and dilated Galien vein.

We report the case of a 15-year-old patient complaining of chronic headaches increasing over the past year, CT scan showed a sacciform dilation of the Galen vein. The child was taken care of in the neurosurgery department of Fann University Hospital by using a ventriculoperitoneal shunt with favorable postoperative and improvement of symptoms up to now adays.

2. OBSERVATION

A 15-year-old adolescent with no particular history who was referred to us from a clinic in the city of Dakar for the management of gradually increasing chronic headaches over the past year, refractory to against the usual analgesics associated with vomiting.

Our review basically noted:

1. A normal state of consciousness with the Glasgow coma scale of 15, isocore and photoreactive pupils;
2. Signs of Intra-cranial hypertension syndrome (headache, vomiting);
3. Non-febrile meningeal syndrome;
4. None cognetive trouble (none trouble of memory, none language's trouble);
5. Hyper pulsatility of the carotid artery on cervical palpation with a systolic murmur on auscultation;

6. Constants including blood pressure at 100/70 mmHg; Oxygen saturation 99% on open air; Respiratory rate at 22 cycles per minute; Heart rate at 102 beats per minute;

The CT scan showed a dilation of the vein of Galen measuring 38 X 58 mm with a collar measuring 9.7 mm responsible for tri ventricular hydrocephalus (Fig. 1).

Transthoracic Ultrasound showed a dilation of the left cardiac chambers without significant valvuloplasties, with a good bi ventricular function (Fig. 2).

A Cytobacteriological examination of the Cerebrospinal Fluid was done and returned sterile.

The appropriate treatment in such case is endovascular by embolization, difficult to achieve in our environment due to the lack of the appropriate technical platform, we decide to place a ventriculo-peritoneal shunt to treat hydrocephalus in order to increase intracranial hypertension. A control CT scan requested remotely (1 month postoperative) showed a slight dilation of the third ventricle and temporal horns of the lateral ventricles; a predominant hydroma on the left (Fig. 3), with a normal clinical examination.

3. DISCUSSION

VGAM is a rare congenital disease caused by arteriovenous shunts between the choroidal arteries and the porencephalic ectatic vein of Galen, vein embryonic precursor [2,3,4].

It develops during weeks 6-11 of gestation, result of persistence of the embryonic vein Markowski [5].

The clinical manifestations are very variable, depending on the age of clinical presentation and size of the malformation. Amacher and Shillito [5] suggest a schematic classification of symptoms according to age at diagnosis. Often manifested in neonatal age by cranial respiration or congestive heart failure (CHF). In childhood, it is early expressed by macrocephaly, hydrocephalus, CHF (congestive heart failure) and in older children vomiting during headache, syncope of stress and subarachnoid hemorrhage are the aims symptoms. Without forgetting that, headache in children's medical milieu is

considered as a common symptom of infectious diseases. however it can hide on a several cerebral disease such as the VAGM. The

Diagnosis can be made before the birth period during the second quarter of pregnancy by making angioMRI [5].

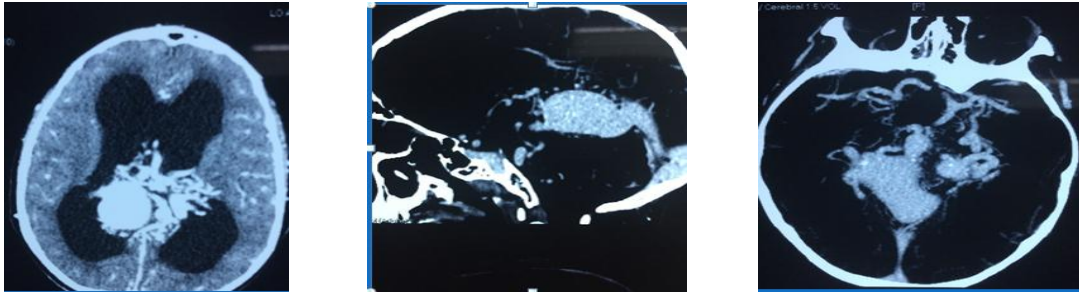


Fig. 1. Sacciform dilation of the Galen vein measuring 38 X 58 mm with a collar measuring 9.7 mm responsible for tri ventricular hydrocephalus

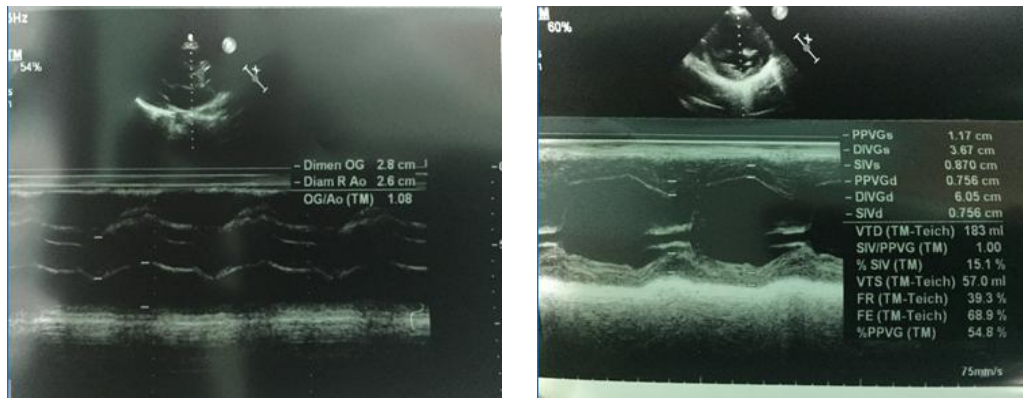


Fig. 2. Dilation of the left heart chambers without significant valvuloplasty and good biventricular function

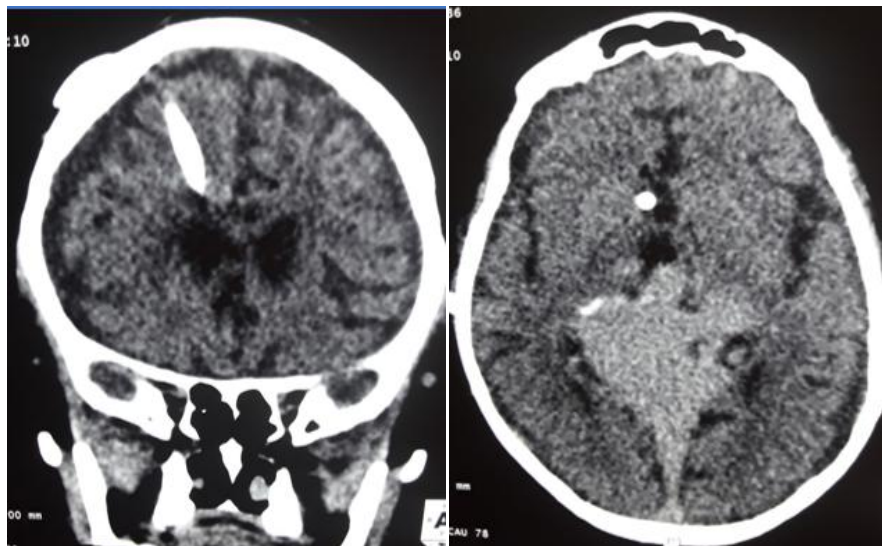


Fig. 3. Slight dilation of the third ventricle and temporal horns of the lateral ventricles; a predominant hydrooma on the left

In our reported case, 15 years old was the patient's age manifestation, with cranial hypertension syndrome as the aim symptom. The CT scan showed a dilation of the Galen's vein with tri ventricular hydrocephalus as details by data from literature [2,5] that's allowed us to confirm the diagnosis of VGAM.

There are several options for endovascular treatment, the most important is embolization aimed at controlling blood flow, which involves being able to occlude the fistula without moving the embolization material. The therapeutic approach should rather focus on stabilizing the cardiopulmonary status of decompensated patients; Appropriate selection of patients who will benefit from endovascular therapy and the course of treatment that promotes the progressive development of bleeding and hemodynamic instability.

Such an approach achieves acceptable cognitive and functional results in the long term.

Rapid ventricular pacing (PVR) reduces cardiac output and blood pressure while the amplitude of blood pressure is reduced without cardiac arrest. However, PVR is often used as an interventional catheterization procedure for congenital aortic stenosis 4 and is a well-known method for balloon aortic valvuloplasty [2,6,7,8].

All the mentioned options were not done in our case, due to the lack of the adequate technical platform; Consequently, we considered the installation of a ventriculoperitoneal bypass system which seemed to relieve our patient, however the literature mentions that DVP would be one of the factors increasing the bleeding and exacerbating of symptoms in Galen vein aneurysm [2].

Some authors [9] advocate ETV as a fairly recent alternative to shunt implantation, mainly used in obstructive hydrocephalus with a global success rate of around 75% and a low complication rate of less than 5%.

Overdrainage is another disadvantage of shunts which can cause symptoms afterwards like a subdural hematoma, a hygroma and split ventricles. Such complications can be avoided with ETV, as it would not change the balance between supratentorial and infratentorial pressures [9]. Without ignoring that even in environments with a suitable technical platform,

embolization which is a major treatment has a success rate of around 60% as reported by certain studies [2,6,8].

4. CONCLUSION

Galen vein malformation remains rare, the taking care of is difficult and focused on embolization and symptomatic treatment. The embolization technique is not always available in all countries that's forcing some authors to do the symptomatic treatment as in our case we placed a DVP.

CONSENT

Informed and written consent have been obtained from the participant and preserved by the author.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist

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