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Vein of Galen Aneurysmal Malformation Presenting with Obstructive Hydrocephalus: A Case Report

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ABSTRACT

The prognosis for children harboring vein of Galen aneurysmal malformation (VGAM) has significantly improved over the last three decades due to refinements in the interventional endovascular techniques. It is now possible to obliterate the malformation with better results. Endovascular approaches evolved as the gold standard treatment option for this disorder. A few reports were published warning about the high rate of complications associated with ventriculoperitoneal shunting (VPS) in this setting. We present a pediatric case with VGAM where an emergency ventriculoperitoneal shunt procedure was required for symptomatic obstructive hydrocephalus (HCP) and management of the subsequent complications that followed it.

Keywords: *Complications, obstructive hydrocephalus, vein of Galen aneurysmal malformation, ventriculoperitoneal shunt*

INTRODUCTION

Vein of Galen aneurysmal malformations (VGAM), which are caused by the maldevelopment of embryonic precursor, the median prosencephalic vein, are rare vascular malformations of the brain. They are most commonly diagnosed in the neonatal period or early childhood.[1] These malformations are most frequently associated with congestive heart failure and hydrocephalus in infants and children. The incidence of VGAM is about one in three million population; and they represent less than 1% of the cerebral arteriovenous malformation.[2] Implication of venous hypertension and/or acquired aqueductal stenosis in VGAM leading to hydrocephalus (HCP) has rendered many arguments amongst the authors.[3-5] A recently published literature review outlined the high risks of ventriculoperitoneal (VP) shunt procedures for HCP associated with VGAM in childhood.[6] Because of the rarity of VGAMs, most publications are in the form of case reports.

Here, we present a pediatric case where an emergency VP shunt procedure was performed for symptomatic obstructive HCP which seemed to be have contributed to the abrupt neurological deterioration maybe due to pathophysiology of VGAM.

CASE PRESENTATION

A 2-year-old male child was referred to our center with a history of progressive swelling of left frontal scalp for past 8 months with increase in head size. It was associated with vomiting for one week and multiple episodes of fever. His neurological status was progressively deteriorating with irritability and altered sensorium. On examination, there was no cardiomegaly or cardiac murmur. Neurologically, Glasgow Coma Scale (GCS) was 9/15 (E2M4V3) and pupils were bilaterally round and reactive to light. He was moving all limbs equally. There was no nuchal rigidity. On fundoscopy, there was frank papilledema. Obvious infective pathology was ruled out following lumbar puncture and cerebrospinal fluid (CSF) analysis and culture. MRI brain showed

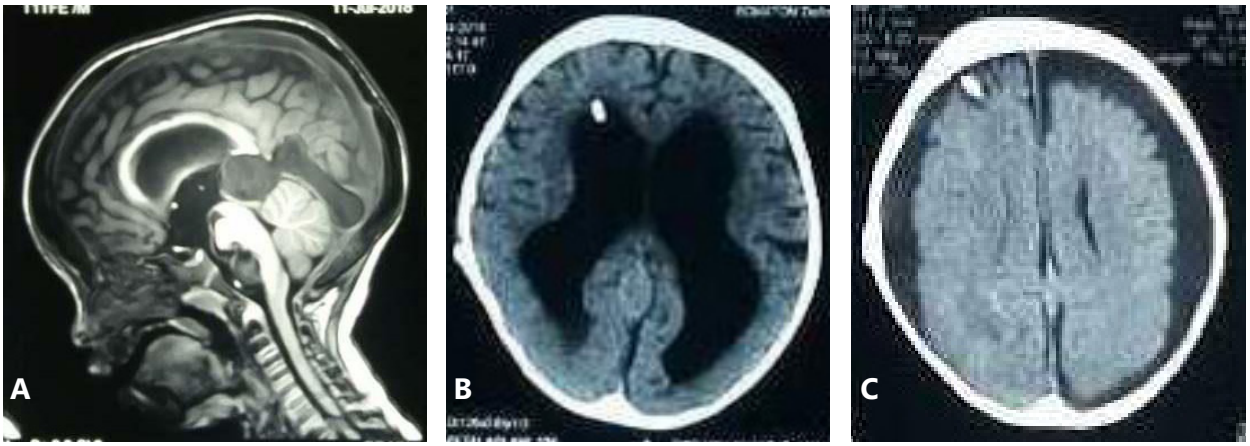

Figure 1. (A) T1-weighted MRI showing dilated Vein of Galen obstructing the aqueduct and proximal dilations of the ventricles. (B) Plain CT head axial view showing tip of shunt in frontal horn of the a right lateral ventricle. (C) CT head axial view showing cresentric hypodense area in bilateral convexity. Note the white tubular structure is the ventricular catheter.

VGAM with compression of the tectal plate in posterior third ventricle, resulting in obstructive hydrocephalus (Figure 1-A).

Diagnosis of obstructive HCP caused by huge VGAM, with low GCS prompted emergency placement of a a medium-pressure VP shunt (Chhabra slit and spring valve) via right anterior approach as a initial management. (Figure1-B) Following surgery, the patient had an episode of seizure on first postoperative day (POD), which was controlled by administrating Phenytoin sodium. He was discharged at POD 4 without any other major event.

However, the patient again presented to Emergency Department on POD 14 with multiple episodes of vomiting for two days and associated one episode of relapse of generalized tonic-clinic seizure. Computed tomography (CT) scan revealed bilateral subdural hygroma (Figure 1-C). Subsequently, the patient underwent evacuation of bilateral subdural hygroma via bilateral frontal burrholes; and, in addition, upgrading of shunt from medium to high pressure shunt was done at the same setting. The surgical intervention was uneventful; however, he developed status epilepticus which was not controlled by use of multiple intravenous antiepileptic drugs (AEDs). The patient was eventually intubated and managed in Pediatric ICU with intravenous Midazolam and Propofol. The seizure was finally controlled, and the patient was extubated on POD 4 and discharged on POD 10 with multiple AEDs (including Leveracetam, Phenytoin and Clobazam). The family has been advised to seek advanced endovascular treatment for definitive treatment of VGAM.

DISCUSSION

Different pathophysiological basis have been postulated that leads to HCP in children with VGAM. Venous hypertension, mechanical obstruction of cerebral aqueduct, post-hemorrhagic HCP and HCP ex vacuo have been implicated. The most accepted explanation is based on the hydrodynamic concept of HCP.[7] Any increase in the venous pressure due to the VGAM results in an impaired CSF reabsorption and alters CSF pressure, which in turn causes increased ventricular volume. In most of the cases, this imbalance in hydrodynamics is considered to be the main cause of HCP in VGAM.[7] Evidence provided by literature illustrates that VP shunting of high venous pressure related HCP in children with incomplete embolized VGAM was associated with poor outcome. CSF diversion does not deal with the problem resulting from the hydrodynamic disorders and only transiently and incompletely resolves the emergency situation at the ventricular level. The reported complications are as high as 70%. Complications include the following: status epilepticus, intraventricular hemorrhage, subdural fluid accumulation and new neurological deficit.[8,9] In our patient with VGAM-associated HCP, post VP shunting presented with both seizures and subdural fluid collection which was evacuation. The management strategy for HCP in VGAM over the last few decades has evolved. It was detected that treatment of choice is early embolization with complete closure or at least significant AV-shunt reduction (> 90%) of the VGAM. It decreases high venous pressure and subsequently, leads to regression of VGAM-associated HCP. Hence, early endovascular embolization may prevent the development of VGAM-associated HCP.[10] In our case, the long-standing, untreated huge VGAM

caused HCP which caused abrupt deterioration of the patient's neurological status, requiring emergency CSF diversion; even though the literature recommends endovascular embolization or coiling in the early phase of VGAM as choice of treatment when the HCP has not developed or to prevent it. However, our patient presented to us in an advanced stage with obstructive HCP. As there is no advanced endovascular facility in the institute and within the country, the only possible alternative was to perform VP shunt on an emergency basis. Ultimately, the patient will require for sure a definitive treatment with endovascular approach to prevent further complications.

CONCLUSION

We present a child harboring VGAM presently with only progressive obstructive HCP and without any cardiac symptoms who required CSF diversion, namely VPS, for abrupt neurological deterioration. Initially, patient did better but went on to have complications (i.e. status epilepticus and subdural fluid collection) which were treated successfully. These complications seemed to be have been caused by shunting in VGAM-associated HCP. The treatment of choice for VGAM is undoubtedly endovascular approach; and literature has reiterated the complications related to shunting alone for those with VGAM and HCP. However, when there is no advanced endovascular facility and in emergency situations, CSF diversion may be the only treatment option one may have.

CONFLICT OF INTEREST

None declared.

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