CASE REPORT

MASSIVE EPISTAXIS IN A NEONATE: A SYMPTOM OF VEIN OF GALEN MALFORMATION!

Shagufta Wahab¹, Rizwan Ahmad Khan², Manjari Thapa Manger³

1. Radiodiagnosis, Aligarh Muslim University, Aligarh, India
2. Pediatric Surgery, Aligarh Muslim University, Aligarh, India
3. PGI Chandigarh, India

Abstract

Vein of Galen Malformation is a rare and intriguing congenital cerebro-vascular anomaly having varied presentation. We present a case of a neonate who presented with one episode of massive epistaxis and was eventually found to have Vein of Galen Malformation with associated complex and unique vascular malformations including AMVs, Carotico-cavernous fistula and Duplicate MCA artery.

Keywords: Epistaxis, Vein of Galen Malformation, Neonate

Corresponding Author: Rizwan Ahmad Khan, Radiodiagnosis, Aligarh Muslim University, Aligarh, India
Email: drrizwanahmadkhan@yahoo.co.in

Introduction

The congenital malformations of Vein of Galen affect the cerebral vasculature leading to complex anatomy of the vein. The presentation is wide-ranging. Imaging involves technical know-how on the part of the radiologist and the management is difficult.

Case Report

A 24-day-old female baby presented in emergency with massive epistaxis, which was eventually controlled by compression and medical management. There was history of enlargement of head birth. However, there was no history of seizures. On physical examination, the head size was increased with prominent superficial veins on face and neck and pulsating proptosis of left eye. Based on the signs and symptoms of the patient some intracranial vascular mass lesion was suspected. We examined the baby with USG skull and color Doppler. USG showed noncommunicating hydrocephalus with massively dilated bilateral lateral and third ventricle. Color Doppler Imaging showed a large rounded anechoic structure of size about 5x3.5cm posterior to the third ventricle showing turbulent flow draining posteriorly into the dilated straight sinus. Based on the Doppler findings the preliminary diagnosis of Vein of Galen malformation was made and MRI was advised. MRI study of the brain revealed a well-defined supratentorial midline cystic mass lesion of approximate size 5.2x3.7cm posterior to the third ventricle showing signal void on spin echo sequences (FIGURE 1).
Figure 1. T2 weighted spin echo MR image showing supratentorial midline cystic mass lesion showing signal void on SE sequences and communicating with the dilated straight sinus.

The lesion was communicating with the dilated straight sinus. On contrast study, there was strong and homogenous enhancement of the central patent lumen with peripheral mural thrombus. Multiple intracranial vascular malformations in form of arterio-venous malformations and arterio-venous fistulas were seen. AVMs with nidus were seen in bilateral lateral and anterior aspect of the Vein of Galen malformation. Duplicate left MCA artery was seen arising from cavernous part of the ICA and draining into sagittal straight sinus with dilated left sided superior ophthalmic vein suggestive of carotico-cavernous fistula, most probably the cause of pulsating proptosis in the patient. Dilated anomalous and tortuous vessels were noted on the anterior surface of the brain stem extending into the cervical spinal cord (FIGURE 2).

Figure 2. Dilated anomalous and tortuous vessels on the anterior surface of the brain stem and cervical spinal cord.
Based on MRI and MR angiography (FIGURE 3) the diagnosis of Vein of Galen Malformation (VOGM) with multiple arteriovenous fistula and malformation was made and the patient was advised endovascular embolisation.

Discussion

Vein of Galen malformation (VOGM) is a rare intracerebral vascular malformation, which accounts for approximately 1% of all intracranial vascular malformations but is responsible for a much larger percentage of cases in pediatric age group.² The malformations involve the deep midline venous channels and has been proved to be the dilated embryonic median prosencephalic vein of Markowaski instead of the vein of Galen itself.³ The VOGM occurs due to direct arteriovenous connections between the arterial network and the median prosencephalic vein. The main feeders of the malformation are the anterior cerebral, middle cerebral, choroidal, thalamoperforating and superior cerebellar arteries.⁵ Due to the high blood flow across the fistula the fetal pattern of venous drainage may persist, such as persistent falcine sinus. Yasargil classified VOGM into four types:

Type 1 is a small cisternal fistula between the VOGM and the pericallosal arteries or posterior cerebral artery.

Type 2 has multiple fistulous communications between the VOGM and the thalamoperforating vessels.

Type 3 has characteristics of both type 1 and type 2.

Type 4 is parenchymal AVM with drainage into the VOGM.⁷

Lasjaunias et al classified VOGM into two types – choroidal and mural types Berenstein et al (1992).¹ The choroidal type is characterized by multiple fistulous communications between the anterior end of the median prosencephalic vein and choroidal, subforniceal or pericallosal
arteries or subependymal branches of thalamoperforators. In mural type, the fistula is situated in the wall of the median prosencephalic vein and fed by collicular and posterior choroidal arteries. VOGM mainly manifests with cardiac or neurologic disturbances. The shunting of blood through the fistula results in high output cardiac failure and right to left shunting of blood causes cyanosis. If fistula remains untreated, pulmonary hypertension and even myocardial ischemia may ensue. The neurological manifestations are due to high cerebral venous pressure which is transmitted to the medullary veins preventing the resorption of fluid and thus results in hydrocephalus, cerebral edema and hypoxia. Thus hydrocephalus is secondary to impaired resorption of CSF due to venous hypertension and not due to Aqueductal compression. The chronic hypoxia results in parenchymal injury resulting in cognitive impairment – delayed milestones to mental retardation. The excessive intravenous pressure leads to opening of alternative channels for drainage of blood like facial veins or basilar or pterygoid plexus. These collaterals produce prominent facial veins and also may cause epistaxis, as in our case.

Neonates usually present with CHF and cyanosis. Infants and children usually present with macrocephaly or delayed milestones or failure to thrive. Older children present with headache and seizures or some present with developmental delay, focal neurological deficits, proptosis and epistaxis. SAH or intracranial hemorrhage may occur due to rerouting of blood into the pial veins. Most cases of VOGMs are detected on routine prenatal USG. Calcification can be seen in approximately 50% cases of thrombosed VOGM on plane radiograph; however calcification is seen only in 14% cases without thrombosis. Sonography and color doppler shows a cystic mass lesion posterior to 3rd ventricle at supracerebellar location with pulsatile blood flow within it and surrounding dilated feeding arteries. Sonography may also show hydrocephalus. USG is also very helpful in antenatal diagnosis and follow-up of patients for assessment of residual flow after endovascular therapy.

Contrast enhanced CT can show an intensely enhancing rounded lesion or “Target sign” in a thrombosed VOGM with associated non communicating hydrocephalus and periventricular ooze. Occasionally focal infarcts, diffuse cerebral atrophy and parenchymal calcification may also be seen. Spontaneous VOGM thrombosis has been described in many cases. Multislice CT angiography is very useful for detailed evaluation of the malformation and complete scan can be performed in a very short time compared to other modalities.

MRI is the investigation of choice for VOGM. It is used both in prenatal and postnatal periods. It can differentiate an AVM that drains into the prominent vein of Galen from true VOGM. It clearly depicts the vascular malformation apart from a detailed evaluation of cerebral parenchymal damage because of its superior soft tissue contrast. This is important for treatment planning and prognosis. MR angiography is used commonly for delineating the feeding vessels and venous drainage. Angiography however is the gold standard investigation for VOGM evaluation prior to any endovascular intervention. It depicts even small feeders supplying the aneurysm, as well as the dynamic aspect of the venous drainage.

To conclude VOGM has a very poor prognosis if not treated early. Early diagnosis coupled with endovascular embolisation of the fistula and...
multidisciplinary management is however capable of significantly improving the dismal prognosis of such patients.

References


