Neonatal Aneurysm Vein Of Galen (case report)

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Abstract

Vein of Galen aneurysm is a rare pathology, representing less than 1% of intracranial vascular malformations [1]. The presentation depends on the age and amount of the blood shunted to the malformation. A neonate or a child may present with cardiac failure, macrocephaly, headache, focal neurological signs or subarachnoid hemorrhage[2]. We report a case of 7-days aged girl who presented with macrocephaly and diagnosed to have vein of Galen aneurismal malformation by brain imaging that showed a large expanding mass in the pineal region.

Key words: vein of Galen aneurysm, vein of Galen malformation, cerebral vein, congenital vascular malformation.

Introduction:

The unpaired great cerebral vein of Galen is formed by the fusion of two internal cerebral veins at the caudal part of the tela choroidea of the third ventricle. It passes caudally and dorsally to merge with the inferior sagittal sinus forming the straight sinus. An aneurysm involving the vein of Galen is a rare sporadic entity of unknown aetiology. The typical diagnostic features are of a midline irregular dilated vascular channel posterosuperior to the third ventricle and directed to the occipital region [2],[3],[4] (fig.1). The common clinical features in the neonatal period are cardiomegaly with congestive heart failure[2-3], and increased intracranial pressure with hydrocephaly or cranial bruit[5]. in older patients, a variety of symptoms have been reported, that include headache[6], syncope, subarachnoid hemorrhage, seizures, and mental retardation[2,7].

Case presentation

A 7-days old girl was brought to the clinic complaining of repeated vomiting and macrocephaly. She was born to a primigravida at 39wks of gestation. Antenatal ultrasound fetal assessment was done and showed arterovenous malformation at 30wks of gestation. As precaution, the mother underwent a cesarean section after 39th week. The weight was 3.2kg, her parents noted that she had enlargement of the head, they also noted prominent dilated vessels at the upper eyelids. There was no history of seizures, jaundice, cyanosis and feeding problems. On examination there was macrocephaly, dilated vein over the skull and forehead, there was no cranial bruit. The rest of the neurological examination was normal. An examination was done with a Siemens Versa Pro, with pulsed Doppler and convex 5 MHz transducer. It revealed, in the axial section of the cranium, the presence of a well defined fluid filled oval structure measuring 45 x 19 mm, located posteriorly above the thalamus. Behind the splenium of corpus callosum (fig.2). The lateral, third ventricles appeared mildly dilated and no other intracranial abnormalities were present. Pulsed Doppler of the cystic lesion and its elongation throughout all its extension demonstrated a high velocity venous flow (fig.2). This type of flow was not seen elsewhere. Ultrasound imaging suggested the possibility of an arteriovenous aneurysm, and the location strongly suggested an aneurysm of the vein of Galen. A CT scan confirmed a big aneurysm of the vein of Galen, with marked contrast medium enhancement (fig.3). Angiography not done and was postponed to after 1 year age. Echocardiograph was normal. The child had no signs of cardiac decompensation, or liver enlargement. These facts and the absence of hypertensive hydrocephalus warranted postponing surgical treatment.

DISCUSSION:

Aneurysm of the vein of Galen was first described by Jager in 1937[8]. Less than
200 postnatal cases had been reported in the literature up to 1984[9]. The use of high resolution echography and pulsed Doppler sonography allowed the first prenatal diagnosis[9]. There are only 10 prenatal diagnoses of aneurysm of the vein of Galen by ultrasound and/or pulsed Doppler. VGA is a vascular abnormality typically found in children[10]. Raybaut et al [11] believe it is a reminiscent of fetal anatomy produced by frequent occlusions of posterior fossa dural sinus, especially at the sigmoid sinus. Typically diagnosed during the neonatal period or in childhood with heart failure signs, macrocephaly and/or cranial murmurs[12]. Children with slow flow fistulas have a better extra uterine adaptation[10]. Subarachnoid and intracerebral hemorrhages can occur because of blood flow reorganization to pial veins[13].

**Prognosis:**
Amacher in 1973 identified three groups (neonatal, infantile and juvenile) based on the seriousness of the lesion and the age of the patient at the onset of symptoms[14]. The severity of cardiomegaly and cardiac decompensation depends on the size and complexity of the vein of Galen aneurysm. If the aneurysm is small (less than 1 mm), the child may be asymptomatic at birth and the aneurysm may cause no relevant consequences for a long period. Later on, during infancy, adolescence or juvenile age, symptoms may occur such as headache, seizure, visual disturbances, due to chronic hydrocephalus and/or subarachnoid or cerebral hemorrhages. On the contrary, with a large aneurysm (greater than 20 mm), the great amount of blood circulating in the high flow fistula induces an overload of the venous circulation that can cause cardiomegaly, decompression at early childhood. Even considering recent micro-neurosurgery advances, the lesion resection usually is not possible. The indications of these treatments are based on case-specific clinical manifestations [13]. Sasidharan et al[15] suggested conservative conduct and patient monitoring for those with severe heart failure, absence of signs or symptoms of intracranial hypertension, and pharmacological control of seizures, elderly with co-morbidities, and neurological stable patients without severe neurological signs or symptoms. Nowadays, endovascular techniques are the procedures of choice considering interventional treatment.

**References:**
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**Figure 1:** Schematic illustration of the venous drainage of the brain. Left: normal; right: pathologic. 1: right internal cerebral vein, 2: left internal cerebral vein, 3-4: right & left basilar veins; 5-6: right & left medial occipital veins; 7: tentorium; 8: left transverse sinus; 9: torcular Herophilus; 10: inferior sagittal sinus; 11: superior sagittal sinus; 12: falx;
Figure 2: Pulsed Doppler in draining vessel documents continuous venous flow a midline supratentorial cystic lesion with draining vessel that extends posteriorly in the direction of the straight sinus is visible.

Figure 3: CT scan with contrast medium. Note the enlarged lateral ventricles and the large well-defined globular mass in the pineal region. Contrast enhancement emphasises the venous drainages and herophylus turcularis.