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Atypical Clinical Manifestation in a Child with Vein of Galen Malformation: Case Report

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Abstract

Vein of Galen aneurysmal malformation (VGAM) is a pediatric intracranial vascular abnormality that is characterized by multiple arteriovenous shunts draining into a dilated median prosencephalic vein of Markowski.[1] The varied clinical presentations and their distinctive and complex angio-architecture make it important for the caring physician to understand the correlation. Most cases present in neonatal life with congestive cardiac failure. We report a rare case of aneurysm of vein of Galen diagnosed in a toddler with initial manifestation mimicking neuro-regressive disorder.

Keywords: Vein of galen malformation; Neuro-regression; CT angiography.

Case Report

An 18 months old male child was apparently alright with normal milestones till 9 months of age when he had an episode of complex partial seizure involving right side of body for which treatment was taken, after which patient stopped sitting on his own. At 11months of age patient went into status epilepticus for 12 hours. seizures started as focal convulsion on right side of body with secondary generalization associated with loss of consciousness for 1 day, for which patient was admitted and managed. Over next 6-7 months patient had lost all the achieved milestones (lost turning over at 13months of age, the ability to pick up objects, waving bye-bye ,and recognizing mother and father at 14months of age). He also stopped responding to sound and stopped talking around 15months of age. Now the patient lies in supine position with stiffening of both upper and lower limbs in flexeded position, cries intermittently, with poor oral intake.

On examination patient was afebrile, vitals were normal, , head was abnormal in shape with parietal bossing, head circumference was 49cms within the 2 standard deviation for that age and sex, anterior fontanel was closed. Visible dilated veins were seen in the neck & forehead, cranial bruit was present. Continuous venous hum could be felt over the dilated veins. On CNS examination, pupils were bilaterally equal normal size reacting to light, patient did not follow light, fundus showed dilated veins with no evidence of papilloedema, gag reflex was absent , but had no cough reflex. Doll's eye reflex was present, Tone was increased in all four limbs with no involuntary movements. Pain sensation was intact, superficial reflexes were present, Babinski sign was positive. Deep reflexes were exaggerated, ankle and patellar clonus were

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present. Rest of the systems was normal.

MRI brain was suggestive of aneurysmal dilatation of vein of Galen - Choroidal type with microangiopathy predominantly in right cerebral hemisphere with obstructive hydrocephalus. Angiography was suggestive of high flow in vein of galen malformation with feeders from posterior cerebral communicating arteries which was seen in all four injections. Embolization was planned.

Discussion

The term "vein of Galen malformation," is a misnomer, because the ectactic vein has been identified as the median prosencephalic vein of Markowski—the embryonic precursor of the vein of Galen itself, which is normally absent at the adult stage.[2] Although it is a relatively rare malformation, VGAM accounts for an estimated 30% of all pediatric vascular anomalies.[3] The vein of Galen aneurysmal malformations are subdivided into two types, choroidal and mural. The choroidal type is where there are multiple shunts communicating with the anterior dilatation of the median prosencephalic vein. The mural type is where the arteriovenous shunt is in the wall of the dilated vein. The choroidal type is more common than the mural type. The presentation differs depending on the size and age at presentation. In the newborn with a large shunt, severe cardiac failure and cranial bruit are the typical signs. In an infant, an enlarging head due to obstructive hydrocephalus at the aqueduct is a common presentation. In older children, subarachnoid hemorrhage and focal neurological signs may occur.[4]

Neonates characteristically have multiple fistulas. Up to 25% of their cardiac output passes through the fistulas causing high-output congestive cardiac failure. Depending on the size of the shunt, adequacy of venous drainage, complexity of arterial supply and the host response, the cardiac manifestations can range from asymptomatic cardiomegaly to severe cardiac failure that is refractory to medical management.[4-6] Infants typically presents with milder cardiac symptoms and smaller shunts. The chief symptom associated with a diagnosis of VGAM during infancy is hydrocephalus. Non communicating hydrocephalus typically results from direct compression of the aqueduct or posterior third ventricle by a venous aneurysm, whereas communicating hydrocephalus is thought to be due to impaired CSF reabsorption by subarachnoid blood.[7] Increased head circumference and seizures are also associated with diagnosis during infancy. Older children and adults usually have low-flow fistulae. These patients usually present with headache and seizures. A small number of patients may also present with developmental delay, focal neurological deficits, proptosis and epistaxis. Subarachnoid hemorrhage and intracerebral hemorrhage can occur in this age group due to rerouting of blood into the pial veins.[3]

The differential diagnosis includes conditions such as cavum vergae, arachnoid cyst, and porencephalic cyst.[8] Left untreated, the mortality of newborns with severe cardiac insufficiency amounts to 100%, in infancy 72%. [9] The safest targeted treatment of a VGAM and also therapy of choice is the endovascular embolization—primarily transarterial using a special kind of glue.[10-12] Higher rates of complications and mortality are described for other therapeutical methods (transvenous embolization, surgical treatment).[13] Time and method of endovascular embolization depends on clinical signs and symptoms of the patient.[14,15]

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