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Conservative Management of Cerebral Venous Angioma - A Difficult Alternative For Patient And Physician

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ABSTRACT

Cerebral venous angioma, also referred to as a developmental venous anomaly is a congenital anomaly of the medullary vein, the vessel that drains into the transparenchymal venous stem. We describe a 7 year old male child presenting with left sided focal seizures in status followed by left hemiplegia. On examination child had altered sensorium and left hemiplegia. His baseline blood, metabolic and CSF studies were normal. MRI brain and MR angiography detected venous angioma involving the deep white matter of Right frontoparietal lobe. Currently child is being managed conservatively on medical therapy as mentioned in the literature. This report describes the clinical and radiologic findings for large venous angioma that caused seizures in a child..

Keywords- cerebral venous angioma, refractory seizures

INTRODUCTION

Cerebral venous angioma, also called developmental venous anomaly (DVA) is a congenital anomaly of medullary vein, the vessel that drains into the transparenchymal venous system. It is one of the causes of seizures in children. Radiological and autopsy studies have demonstrated that DVAs occur in 2.5 to 3% of the general population and constitute approximately 60% of all vascular malformations of the central nervous system [1-3]. Patients with venous angioma are

managed on medical therapy. Surgical intervention is indicated specifically in the presence of an associated lesion like cavernoma which must be undertaken with the objective of preserving the DVA and collecting veins. Surgical intervention carries the risk of catastrophic venous ischemic and hemorrhagic complications. [4]

Case history: A seven year old male child presented in emergency hours with multiple episodes of focal convulsions involving left upper and lower limb since two hours. Parents noticed altered sensorium and inability to move left upper and lower limb following convulsions. Patient had headache prior to onset of convulsions. There was no fever, vomiting, diplopia, or irritability. In past child had febrile seizures at age of five years, EEG at that time showed generalised slowing and seizure activity. Elder sibling had seizures secondary to neurocysticercosis one year back. Child was developmentally normal, immunised appropriately for his age and birth history was clinically uneventful. On admission child had altered sensorium, was arousable but drowsy. Pupils were bilaterally and equally reacting to light and had no papilledema. Cranial nerves on examination were normal. Power in left upper and lower limb was I/V and > IV/V in right upper and lower limbs. All deep tendon reflexes were brisk with left plantar extensor response. A clinical diagnosis of space occupying lesion like neurocysticercosis, tuberculoma was made. Meanwhile on baseline investigations his Hb was 10.3 gm/dl, WBC- 10300/cumm P58,L26, E10,M6. Platelets- 3.69 lacs/cumm. RBS was 124 mg/dl. Malarial antigen and peripheral smear for malaria parasite was negative. BUN was 29mg/dl, Creatinine was 0.8 mg/dl, Sr.Na was 149 meq/L, K 4.5 meq/L and Cl 104 meq/L. X- ray chest was normal. Urgent CT scan Brain was non-conclusive showing prominent enhancement along the cortical sulci and sylvian fissure with more significant involvement of right side. CSF protein was 12 mg /dl, sugar 78mg/dl, and one lymphocyte. CSF C/S detected no growth. Mantoux test was negative after 48 hours.

Clinically child continued to be drowsy till next 48 hours and developed multiple episodes of convulsions on two parenteral anticonvulsant drugs. Convulsions gradually became left focal onset with secondary generalisation, occured more frequently and persisted for longer duration and progressively became less responsive to anticonvulsants, which indicated the possibility of an organic brain lesion. MRI brain was done which showed multiple stellate tangles of medullary vein in deep white matter with convergence draining into the inferior saggital sinus. MR angiography detected venous angioma involving the deep white matter of right frontoparietal lobe. Convulsions were controlled with parenteral anticonvulsant drugs, gradually changed to and controlled with two oral antiepileptic drugs. Over one week the power in affected limb gradually became normal. Child is currently managed on oral antiepileptic drugs yet continues to have intermittent episodes of seizures even on compliant therapy.

Discussion:

Cerebral venous angioma is a congenital anomaly of the medullary vein, the vessel that drains into the transparenchymal venous stem. Developmental venous anomaly (DVA) is a widely used synonym

for venous angioma or cerebral venous malformation. DVAs are encountered in both the pediatric and adult populations, with a slight predominance in males. [5] They are low flow congenital brain vascular malformations solely of venous origin. [6-8] DVAs are the most commonly encountered cerebral vascular malformation and are frequently reported as incidental findings in computed tomography and magnetic resonance imaging. DVAs are considered extreme anatomic variations of the normal transmedullary cerebral vasculature necessary for the drainage of white and gray matter of the brain tissue as the typical pial or subependymal venous drainage of their territory is absent. DVAs may drain into both the superficial and deep venous systems. [7-9] An associated impaired brain perfusion attributed to venous congestion in areas drained by DVAs has also been documented. Though brain parenchyma drained by a DVA has historically been considered to be normal, recent reports have found abnormalities like white matter lesions locoregional cerebral atrophy, dystrophic calcification, cavernous malformations and arteriovenous malformations in a significant proportion of patients. [6]

DVAs are characterized by a cluster of venous radicles that converge into a collecting vein, resulting in the typical caput medusae appearance thereby affecting a variable volume of brain parenchyma, ranging from a few sulci to even a whole hemisphere. DVAs occur more frequently at the supratentorial compartment, with frontal lobe predominance. [10] DVAs occur most often in the frontal lobe (36% to 56%) followed by the parietal (12% to 24%), occipital (4%), and the temporal lobes (2% to 19%); the cerebellum (14% to 29%); the basal ganglia (6%); the thalamus, the ventricles (11%); and the brainstem (less than 5%). [7-9]

A few reports in the literature have documented developmental venous anomaly—related clinical features like tinnitus, epilepsy, neurologic deficits, and intracranial hemorrhage.[6,7,11] Epilepsies may occur due to associated cortical dysplasias,[6]and pseudotumoral effects can be secondary to associated lymphatic malformations.[12]

The current understanding of DVA is based primarily on adult studies. There are no published studies for the evaluation of DVAs in the pediatric population. The type and frequency of associated features in the pediatric population may be different [13].

DVAs are usually asymptomatic and follow a benign clinical course in the vast majority of cases [4,14-16] However, 18–40% of DVAs are associated with one or more cavernous malformations [17], which are at higher risk of bleeding. DVAs usually present with headache, seizures or dizziness. In these cases, management decisions are usually focused on the cavernoma rather than on the DVA. While surgery is not required in the majority of patients with DVA alone, individuals diagnosed with a DVA should undergo neurosurgical assessment and more thorough radiological evaluation to exclude the presence of a coexisting lesion. Surgical procedure is indicated in the presence of a related lesion and shall be undertaken with the objective of preserving the DVA and collecting veins. DVA alone is not an indication for surgical intervention. Catastrophic

venous ischemic and hemorrhagic complications may result from the surgical removal of a DVA or damage to a collecting vein, due to its important contribution in the normal cerebral venous drainage. [4] While removing cavernous angiomas the associated venous angioma needs to be preserved.[18] Non-invasive neuroradiological examinations such as CT and MRI generally permit diagnosis of DVAs and detection of associated cavernoma. Digital subtraction angiography is reserved for cases presenting with ischemic or hemorrhagic infarction, or whenever an associated vascular malformation is suspected.

MRI is superior to CT in demonstrating associated parenchymal abnormalities such as white matter lesions, and cavernous malformations [5]. Some authors recommend that venous angiomas to be managed conservatively as these lesions are deep seated and removal of angioma without resection of interspersed normal brain parenchyma is difficult. [19,20]

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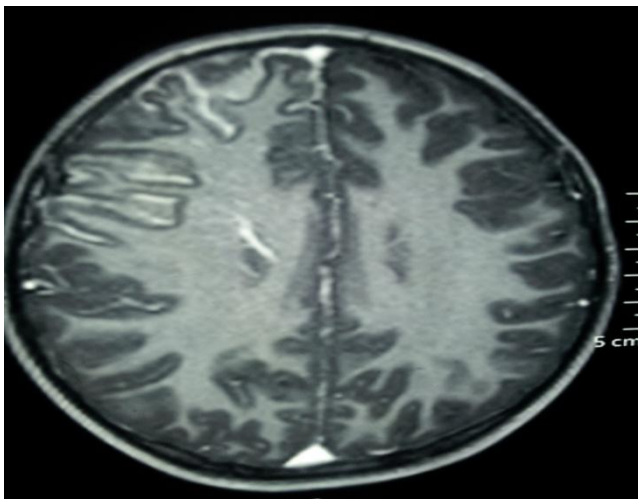

Figure-1 MRI Brain Axial image showing multiple medullary flow voids in deep mater of right frontoparietal areas draining into transcortical vein.

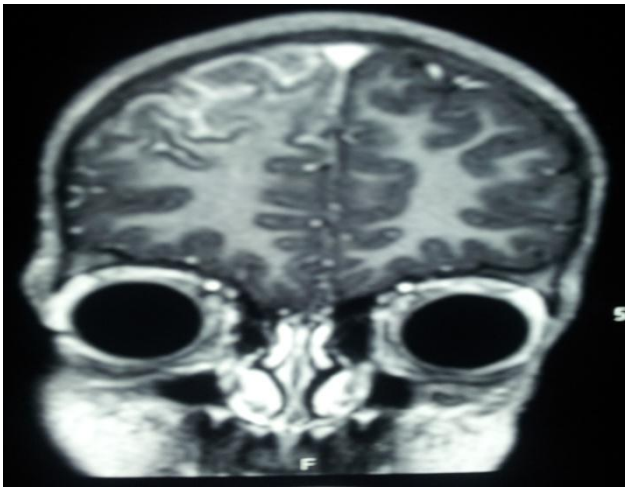

Figure-2 MRI Brain Coronal image showing increased intensity signals right frontoparietal area.

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