VEIN OF GALEN ANEURYSMAL MALFORMATION PRESENTING AS MELTING BRAIN SYNDROME

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ABSTRACT

Vein of Galen aneurysmal malformations (VGAM) are congenital arterio-venous malformations in infants and fetuses which are rare and characterized by shunting of arterial flow into an enlarged cerebral vein dorsal to the tectum. These malformations present in early childhood and often present as congestive heart failure in the neonate. It is of choroidal and mural type. It can present as congestive cardiac failure in early life due to high cardiac output or show neurological manifestations due to cerebral venous hypertension. Untreated VGAM lead to chronic venous ischemia manifested by the development of subependymal atrophy with ventricular dilatation and calcification in subcortical white matter. These calcifications reflect deep hydrovenous watershed failure. These occur when the compliance of medullary veins loses its normal ventriculo-cortical gradient. We report a case of 16 month old female child presenting with vomiting, convulsions with neuroregression. MRI Brain showed prominent flow void inferior to the splenium of the corpus callosum and posterior to the third ventricle. Diffuse T2 hyperintense areas were noted involving the bilateral cerebral white matter involving fronto-temporo-parieto-occipital region with multiple foci of blooming in bilateral thalamus and lentiform nuclei and in bilateral fronto-parieto-occipital subcortical and deep white matter on the GRE suggestive of chronic venous ischemia with dystrophic calcification and mild generalized cerebral atrophy suggestive of melting brain syndrome. CT Brain with cerebral angiography confirmed white matter ischemia with dystrophic calcification, VGAM and its feeders.

Keywords: Vein of Galen Aneurysmal Malformation, Melting brain syndrome, Chronic Venous ischemia, Venous hypertension.

Introduction

Vein of Galen malformations (VGAM) constitutes 1% of intracranial vascular malformations and 30% of vascular malformations in paediatric age group. In 1895, Steinheil referred it as a Varix aneurysm and made the first reference to Galenic malformation. It has an incidence of 1:25000 and is a rare congenital cerebral arterio-venous malformation. It develops during 6th-11th weeks of gestation and can be diagnosed antenatally during ultrasound.1 Depending on anatomy and Angio-architecture of the malformation, age of the patient symptoms varies. In a neonatal/choroidal type of VGAM, volume overload leading to high output cardiac failure occurs with resultant tachycardia, cardiomegaly, pulmonary hypertension, pulmonary edema and multi-organ failure. In an infant/mural type of VGAM, AV fistula occurs within the wall of the median prosencephalic vein of Markowski with resultant hydrocephalus, microcrania and developmental retardation. Epileptic seizures can occur rarely due to brain damage.2

Case Report

A 16 month old female child presented with neuro-
regression with repeated episodes of convulsions with vomiting. She was referred to the MRI department for MRI Brain. MRI BRAIN showed mild generalized cerebral atrophy- disproportionate to age, ill-defined hyperintense areas in bilateral cerebral white matter on T2WI and FLAIR appearing hypointense on T1WI with no restricted diffusion on DWI (Fig. 1, 2, 3b). Multiple small foci of blooming s/o calcification were noted on GRE T2 (Fig. 3a) in bilateral thalami, lentiform nuclei, bilateral fronto-parieto-occipital subcortical and deep white matter suggestive of chronic venous ischemia. A prominent flow void was noted posterior to the third ventricle inferior to the splenium of corpus callosum suggestive of VGAM (Fig. 2).

MR Angiography and Venography (Fig. 4a, b) showed prominent feeders from left posterior choroidal artery with drainage in straight sinus which was prominent. CT Brain showed ill-defined hypodense areas in bilateral fronto-parieto-occipital white matter with multiple small calcific foci in bilateral thalami, lentiform nuclei, bilateral fronto-parieto-occipital subcortical

Discussion

Vein of Galen malformations occur due to direct communication between arterial network and median deep white matter s/o Chronic venous ischemia (Fig. 5a-d). CT Brain Angiography showed a densely enhancing vascular structure in the midline posterior to the third ventricle and inferior to splenium suggestive of VGAM. Multiple tortuous ectatic arterial feeders (left lateral choroidal artery, anterior choroidal, posterior choroidal) were reaching and opening VGAM. The left lateral choroidal artery was the largest feeder. Subependymal arteries, posterior tectal perforators and right thalamic perforators were hypertrrophied and tortuous. VGAM was communicating with straight sinus through a short stenotic segment. Straight sinus was hypertrophied. Reflux was noted in other dural venous sinuses and spinal epidural veins suggestive of venous reflux in retrograde fashion. (Fig. 6a-b)
These arterio-venous communication occurs in the cistern of velum interpositum and the quadrigeminal cistern. Common feeders are prosencephalic or anterior group (the anterior cerebral, anterior choroidal, middle cerebral and the posterolateral choroidal arteries), and the mesencephalic or posterior group (the posteromedial choroidal, posterior thalamoperforating, quadrigeminal and superior cerebellar arteries).1

VGAM-a rare vascular malformation, is neither an aneurysm nor a vein of Galen, according to embryogenesis of the malformation. Normal deep veins are absent and venous drainage is through embryonic vessels. The Galenic malformation in fetal life is due to the persistence of the single median prosencephalic vein of Markowski resulting in a dilated venous sac in the midline.3 At the end of the 10th week temporary vein drains the choroid plexus from both sides. With the
development of the thalamus, basal ganglia, and cerebral vascularization, paired internal cerebral veins progressively annex the venous drainage of the midline structures. Posterior end of the median prosencephalic vein is joined by bilateral internal cerebral veins, the future vein of Galen; the remaining segment of midline solitary embryonic vein of Markowski progressively attenuates. The vein of Galen is completely in the subarachnoid space.

VGAM are of two types- (a) Choroidal type- Multiple feeders from choroidal, thalamostriate and pericalosal arteries empty into the vein of Galen; usually present in neonates with congestive heart failure and/or hydrocephalus. Usually show stenosis of draining venous sinus and persistence of falx sinus. (b) Mural type- Few feeders from collicular or posterior choroidal arteries supplying periphery of dilated vein of Galen.3

Imaging: CT depicts cerebral parenchymal damage in the form of generalized cerebral atrophy, ventriculomegaly, chronic ischemic changes, parenchymal calcifications and focal parenchymal infarcts. CECT demonstrates VGAM as a well-defined, multiloculated, intensely enhancing lesion in the cistern of velum interpositum, thrombosis in aneurysmal sac and can detect crescentic rim of calcification in thrombosed VGOM. Collateral parenchymal veins are dilated. Thrombosed VGOMs show crescentic rim of calcification. Complete calcification of the sac is extremely rare and calcification is rarely seen before the age of 15 years.4 MRI can demonstrate the location of the fistula, arterial components, presence of any nidus and status of venous drainage. It can identify major arterial trunks, primary and secondary branches feeding the fistula, draining veins venous anomalies. Superior soft tissue contrast resolution is useful in detection of cerebral parenchymal changes and ventriculomegaly. MRI can detect draining veins and venous anomalies. MR Angiography and MR Venography are being increasingly used in the initial evaluation of these lesions as a noninvasive alternative to diagnostic angiographic studies.4

VGAM clinically presents either due to high output cardiac failure or cerebral venous congestion and abnormal CSF flow. In untreated VGAM, venous congestion in watershed areas leads to venous ischemia with the resultant development of subcortical white matter calcification and subependymal atrophy with ventriculomegaly. Chronic venous ischemia leads to cerebral atrophy and parenchymal calcification known as melting brain syndrome. Calcification is dystrophic and usually bilateral and symmetrical. Subependymal atrophy is commonly seen in occipital regions. These calcifications reflect deep hydrovenous watershed failure. These occur when the compliance of medullary veins loses its normal ventriculo-cortical gradient.5,6 Neurological manifestations in VGOM occur primarily due to cerebral venous hypertension. These lesions are associated with venous anomalies like poorly developed venous drainage, secondary venous stenosis and occlusion. Due to restriction of venous drainage and high flow in an arterio-venous shunt there is an increase in cerebral venous pressure. In infants arachnoid granulations are not fully matured. Hence, most of ventricular CSF is reabsorbed across ventricular ependyma into brain parenchyma, which is subsequently drained by medullary veins. In VGOM high venous pressure is transmitted to medullary veins which prevent reabsorption of fluid (CSF) with resultant hydrocephalus, cerebral edema and hydrocephalus.7,8 Hydrocephalus in VGOM is not due to aqueductal compression, but is due to impaired resorption of CSF due to venous hypertension. Venous hypertension leads to chronic hypoxia with resultant progressive parenchymal cerebral damage. This presents as cognitive impairment ranging from delayed milestones to mental retardation.9 Ventriculomegaly in VGAM is multi-factorial obstruction at the aqueduct of sylvius with resultant hydrocephalus, resorptive blocks, abnormal transependymal resorption of CSF and ex-vacuo hydrocephalus (due to encephalomalacia and cerebral atrophy). Resorptive blocks occur due to increased pressure within sagittal sinus. Developmental retardation occurring in vein of Galen aneurysm is due to arterial steal, chronic venous hypertension with venous ischemia and subsequent venous infarction due to spontaneous partial or complete thrombosis of VGAM.7 Venous haemodynamic balance is necessary for myelogenesis, synaptogenesis and maturation of granulations which are needed for normal brain growth. As VGAM is active from birth, it leads to
rapid brain atrophy due to apoptosis and cause the brain to melt. The melting brain syndrome is seen only in the severe neonate form of VGAM with patent sinuses. Calcifications represent slow and relentless brain insult (dystrophic calcification). Calcifications in VGAM are seen in 3 different locations - mural (within the lesion - due to complete or partial thrombosis), striatum - Bilateral and symmetrical due to striatal congestion. (venous ischemia), subcortical white matter - due to watershed failure. Chronic venous hypertension leads to impaired oxygenation (venous ischemia), delayed myelination, subependymal atrophy and ventriculomegaly leading to melting brain syndrome. 5, 6, 10

Conclusion

Early diagnosis and treatment of VGAM is essential as untreated VGAM lead to chronic venous ischemia manifested by the development of subependymal atrophy, ventricular dilatation and calcification in subcortical white matter. It is a cause of convulsions and neuroregression.

References


