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Rare case presentation of vein of Galen malformation with melting brain syndrome

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Abstract

Introduction: The Vein of Galen aneurysmal malformations (VGAM) is a rare form of embryonic arteriovenous shunt (AVS) in the choroidal fissure [1].

Case Report: 18 months female child brought with C/o Regression of milestones and Vomiting since 2 months. Echocardiography showed mildly dilated left atrium and ventricles. Fundus examination showed temporal pallor & later optic atrophy. MRI brain: S/O Vein of Galen malformation with melting brain syndrome.

Conclusion: Early and appropriate diagnostic tests permit the proper management.

Keywords: Vein of Galen malformation, melting brain syndrome, cardiac failure, embolization

Introduction

The Vein of Galen aneurysmal malformations (VGAM) is a rare form of embryonic arteriovenous shunt (AVS) in the choroidal fissure [1]. It consists of anterior and posterior choroidal arteries and anterior cerebral which develops before the formation of the vein of Galen and straight sinus, and the pouch drains via falcine sinus to the superior sagittal sinus [2].

Case report

18 months female child brought with C/o Regression of milestones and Vomiting since 2 months. Past history and Birth history being uneventful. Family history was also not significant. Child was immunized till 9 months of age.

O/e Conscious and alert. No dysmorphism. Anterior fontanelle was closed. Vitals- HR-114/min, RR-28/min, CRT<3 sec, HC-46.5 cm, CC-46cm, LT- 75cm, WT- 7.8 Kgs, MAC-14 cms

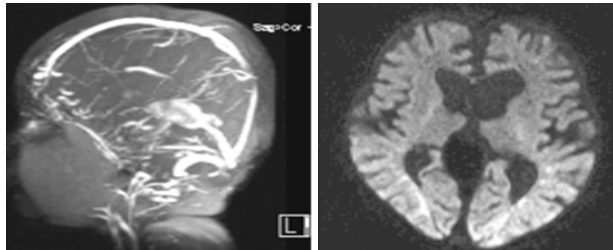
S/e- (CNS) - Cranial Nerves normal. All four limbs showed hypotonia, All group of muscles of upper and lower limbs had power of 2/5, Plantar – up going, deep tendon reflexes were diminished and rest were normal.

CBC

Hb- 10.5 Gm%
TLC-8400/Cumm
P-44%, L-42%, E-2%, M-12%
ESR- 29
P/C- 3.4 Lakhs/Cumm
AEC- 168 Cells/Mm³
PCV- 37.5
MCV-76.7, MCH-24, MCHC-31.3
Pbs-Microcytic Hypochromic Rbcs.
Serum Bilirubin (T) - 0.4 Mg%
Serum Bilirubin (D) -0.2 Mg%
SGPT-23 IU/L
SGOT-41 IU/L
Serum ALP-130 Iu/L
Blood Urea- 13 Mg%

Serum Creatinine- 0.3 Mg%
Serum. Protein- 5.6 Gm%
Serum. Albumin – 4.4 Gm%
Globulin – 1.2 Gm %
Serum. Calcium-9.6 Mg%
Serum.Phosphorus-4.2 Mg%
Serum.Magnesium-2.2 Mg%
Serum. Sodium- 136 Mmol/L
Serum. Potassium-4.7 Mmol/L

Echocardiography showed mildly dilated left atrium and ventricles. Fundus examination showed temporal pallor & later optic atrophy. MRI brain: S/O Vein of Galen malformation with melting brain syndrome. Further management couldn't be done as the child was discharged against medical advice.



MRI Angiography

DWI

Discussion

Vein of Galen aneurysmal malformations (VGAM) and Vein of Galen aneurysmal dilations (VGAD) are the most frequent arteriovenous malformations in infants and fetuses. VGAM consisted of a tangled mass of dilated vessels supplied by an enlarged artery. The malformation increases greatly in size with age, although the mechanism of the increase is unknown. The right sided cardiac chambers and pulmonary arteries also develop mild to severe dilation.

There are 5 patterns of malformation

Many vessels, including anterior cerebral arteries, thalamic perforating arteries, and superior cerebellar arteries discharge into the vein of Galen.

1. A single posterior choroidal artery drains into the vein of Galen.
2. One or both posterior choroidal and one or both anterior cerebral arteries drain directly into the Galenic system.
3. An angiomatous network of posterior choroidal and thalamic perforating arteries enter the Vein of Galen directly.
4. A high flow arteriovenous malformation in the right inferior frontal lobe drains via the inferior sagittal sinus and pericallosal vein into the Vein of Galen

Malformations may lead to

1. Cardiac failure
2. Cranial bruits (pattern 1)
3. Hydrocephalous
4. Subarachnoid hemorrhage in neonates.

The heart failure is due to the size of the arteriovenous shunt that can steal 80% or more of the cardiac output, with large volumes of blood under high pressure returning to the right heart and pulmonary circulation and sinus venosus atrial

septal defects. It is also the most common cause of death in such patients.

Such large AVS may raise venous sinus pressure, which is transmitted to the cortical and the medullary veins. This results in water congestion of brain parenchyma, leading to sub ependymal atrophy and progressive “melting brain syndrome” [3]. The treatment is conservative i.e embolisation and surgery has little role.

Conclusion

Management of AVS of the brain is among the most challenging areas in modern medicine. Therefore early and appropriate diagnostic tests permit the proper management.

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