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A VEIN OF GALEN MALFORMATION - A RARE CAUSE OF CONGESTIVE CARDIAC FAILURE IN NEONATE

Dr.Bela Verma	M.D. Pediatrics, Professor and Head of Department, Department of Pediatrics,GGMC and Sir J.J.Hospital, Mumbai
Dr.Poonam Mane*	JuniorResident,M.D. Pediatrics,Department of Pediatrics,GGMC and Sir J.J.Hospital Mumbai*Corresponding Author
Dr.Deepa Ganpathi	JuniorResident,M.D. Pediatrics,Department of Pediatrics, GGMC and Sir J.J.Hospital, Mumbai
Dr.Kaif	JuniorResident,M.D. Pediatrics,Department of Pediatrics,GGMC and Sir J.J.Hospital, Mumbai

ABSTRACT Vein of Galen aneurysmal malformation (VGAM) is a rare congenital vascular malformation caused by the maldevelopment of its embryonic precursor,the median prosencephalic vein of Markowski.Vein of Galen aneurysmal malformation (VGAM) is a rare cause of congestive heart failure in the neonatal period and carries a high mortality rate. It is a difficult diagnosis because the clinical picture often presents with symptoms suggesting that the patient has a congenital cardiac malformation.1,2 Patients are subjected to high-risk procedures such as cardiac catheterization to establish a diagnosis. These investigations often have negative findings and can delay correct diagnosis. Here we report a case of Four days old neonate with c/o Increased respiratory activity since 2 days with tachycardia,tachypnea and oliguria.Continuous murmur was noted with features suggestive of Congestive cardiac failure i.e.Tachycardia,tachypnoea,hepatomegaly. Bruit + over Anterior Fontanelle.USG skull revealed the presence of Vein of Gallen Aneurysmal Malformation. 2D ECHO was done which suggestive of Congenital Heart Disease;moderate OS ASD with left to right shunt Dilated cardiac chambers;No PAH;Normal biventricular function .CECT Brain showed the presence of Aneurysmal deep venous structure measuring 20 x 25 mm noted in the supratentorial region of posterior fossa (in quadrigeminal cistern) which drained to vein of galen (VGAM) and the vessel feeding the aneurysm was arising from basilar artery.Hence diagnosis was made.Child initially treated symptomatically for cardiac failure then Endovascular treatment i.e. embolization was performed on 30th day of life.The VGAM was partially occluded.Then symptoms got relieved after Embolization.

KEYWORDS :

INTRODUCTION

Vein of Galen aneurysmal malformation (VGAM) is a rare congenital vascular malformation caused by the maldevelopment of its embryonic precursor,the median prosencephalic vein of Markowski.Vein of Galen arteriovenous malformation (VGAM) is a severe and complex vascular malformation characterized by multiple arteriovenous shunts between the vein of Galen and the choroidal arteries. Because of these shunts, vascular steal occurs and this determines the increase in blood returning to the heart at the level of the cerebral cortex, resulting in overload of the right heart and progressive heart failure [1].

The majority of VGAMs become symptomatic in the neonatal period and if left untreated have an almost 100% morbidity and mortality [2].

CASE REPORT

Four days old Full term/Appropriate for gestational age/ male child, 1stBirth by order born out of consanginous marriage (3rd degree), with c/o Increased respiratory activity since 2 days.soon Child developed tachycardia,tachypnea and oliguria. On examination- Continuous murmur was noted with features suggestive of Congestive cardiac failure i.e.Tachycardia,tachypnoea,hepatomegaly. Bruit + over Anterior Fontanelle (Pulsatile AF), Investigations were done as follows ;Chest X ray –Increased CT ratio>0.5 suggestive of cardiomegaly. USG skull revealed the presence of Vein of Gallen Aneurysmal Malformation. 2D ECHO was done which suggestive of Congenital Heart Disease;moderate OS ASD with left to right shunt Dilated cardiac chambers;No PAH;Normal biventricular function

.CECT Brain showed the presence of Aneurysmal deep venous structure measuring 20 x 25 mm noted in the supratentorial region of posterior fossa (in quadrigeminal cistern) which drained to vein of galen (VGAM) and the vessel feeding the aneurysm was arising from basilar artery

TREATMENT

Endovascular treatment i.e. embolization was performed on 30th day of life.The VGAM was partially occluded.After embolization there was symptomatic relief and the patient was discharged.

DISCUSSION

It is a case that illustrates the dramatic outcome of a neonate presenting with heart failure and being diagnosed with a vein of Galen aneurysmal malformation 6 The size of the fistulas determine the amount of arteriovenous shunting and consequently the time course to the development of cardiac failure.1 Heart failure develops because of volume overload in the right side of the heart. Due to a low vascular resistance in the head, the majority of left ventricular output is directed toward the head. These 2 mechanisms lead to reduced systemic blood flow, severe lactic acidosis, potentially ischemic multiple organ failure, and persistent pulmonary hypertension of the newborn.7 During diastole, the systemic perfusion is furtherreduced by circulatory “steal” to the vein of Galen aneurysmal malformation, which produces the characteristic reversed diastolic flow in the descending aorta.

Usually, signs of severe heart failure develop shortly after birth. The clinical picture is dominated by findings suggestive of a congenital cardiac disease such as cyanosis, compromised peripheral pulses, and a cardiac murmur. Often cyanosis is present suggestive of persistent pulmonary hypertension of the newborn with signs of right-to-left arterial shunting at the ductal level.8 A continuous murmur can be heard over the scalp, but this is not routinely checked as our case illustrates.

Echocardiography often shows a structural normal heart and great vessels but a dilated and noncompliant right ventricle with signs of suprasystemic pulmonary arterial pressure and right-to-left ductal shunting. Computer tomography imaging give a clear demonstration of the intracranial arteriovenous malformation. The treatment of vein of Galen aneurysmal malformation involves initial cardiovascular stabilization directed toward improvement in the non cerebral systemic cardiac output. In most situations, a vasodilator agent, either alone or in combination with low to moderate dose inotropic agents, is needed. Milrinone and inhaled nitric oxide have been found suitable for use in vein of Galen aneurysmal malformation.1,6.

In the modern treatment of vein of Galen aneurysmal malformation, surgery has little role. Embolization by endovascular treatment is the preferred treatment modality. A good outcome is to be expected when

treatment is performed before significant brain injury has occurred in patients who have been selected carefully.Vein of Galen aneurysmal malformation presents with signs of congestive heart failure.

CONCLUSION

Vein of Galen aneurysmal malformation is a rare disorder and is often diagnosed late. Considering vein of Galen aneurysmal malformation in the differential diagnosis of neonatal congestive heart failure is important in minimizing diagnostic delay Initial treatment is targeted at improving noncerebral systemic circulation and reduction in pulmonary vascular resistance. When circulatory stabilization can be achieved, endovascular treatment of the arteriovenous malformation is the preferred method of intervention. However, despite optimal management, the disorder still carries a high mortality risk.

REFERENCES

1) Kojmane W, Hmami F, Atmani S. An unusual cause of cardiac failure in a neonate. Arch Pediatr. 2019 Apr;26(3):176-178. doi: 10.1016/j.arcped.2019.02.004. Epub 2019 Mar 1. PMID: 30827772.
2) Madhuban A, van den Heuvel F, van Stuijvenberg M. Vein of Galen Aneurysmal Malformation in Neonates Presenting With Congestive Heart Failure. Child Neurol Open. 2016 Apr 4;3:2329048X15624704. doi: 10.1177/2329048X15624704. PMID: 28503603; PMCID: PMC5417289.
3) Spada C, Pietrella E, Caramaschi E, Bergonzini P, Berardi A, Lucaccioni L, Iughetti L. Heart failure caused by VGAM: a lesson for diagnosis and treatment from a case and literature review. J Matern Fetal Neonatal Med. 2021 Jul;34(14):2384-2390. doi: 10.1080/14767058.2019.1665013. Epub 2019 Sep 18. PMID: 31488018.
4) Chen MY, Liu HM, Weng WC, Peng SF, Wu ET, Chiu SN. Neonate with severe heart failure related to vein of Galen malformation. Pediatr Neonatol. 2010 Aug;51(4):245-248. doi: 10.1016/S1875-9572(10)60047-2. PMID: 20713291.
5) De Rosa G, De Carolis MP, Tempera A, Pedicelli A, Rollo M, Morena TC, Luca E, De Luca D, Conti G, Piastra M. Outcome of Neonates with Vein of Galen Malformation Presenting with Severe Heart Failure: A Case Series. Am J Perinatol. 2019 Jan;36(2):169-175. doi: 10.1055/s-0038-1666813. Epub 2018 Jul 10. PMID: 29991070.
6) Cory MJ, Durand P, Sillero R, Morin L, Savani R, Chalak L, Angelis D. Vein of Galen aneurysmal malformation: rationalizing medical management of neonatal heart failure. Pediatr Res. 2023 Jan;93(1):39-48. doi: 10.1038/s41390-022-02064-1. Epub 2022 Apr 14. PMID: 35422084.
7) Lasjaunias, P. L. et al. The management of vein of Galen aneurysmal malformations. Neurosurgery 59, S184–S194 (2006).
8) Chevret, L. et al. Severe cardiac failure in newborns with vgam. prognosis significance of hemodynamic parameters in neonates presenting with severe heart failure owing to vein of Galen arteriovenous malformation. Intensive Care Med. 28, 1126–1130 (2002). - DOI