An Unusual Cause of Heart Failure in a Newborn: Your Diagnosis?: A Case Report

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Abstract

Introduction: Arteriovenous malformations (AVMs) are abnormal connections between an artery and a vein whereby the interconnecting capillary network is missed. Vein of Galen is a short midline venous structure formed by the confluence of the two internal cerebral veins and the basal veins of Rosenthal and the aneurismal malformation of this vein is a congenital vascular anomaly that comprises 30% of the pediatric vascular malformations. The pathogenesis of the vein of Galen aneurysmal malformation (VGAM) is unknown. Here, we report a neonate patient with complications related to vein of Galen aneurysmal malformation.

Case Presentation: A 9-day-old male neonate referred with icter, poor feeding, tachypnea, and tachycardia and was admitted to the neonatal intensive care unit (NICU) of Vali-e-Asr hospital in Birjand city, Iran, on 5th July 2016. In his chest X-ray (C-X-ray), a cardiomegaly was shown. Suspecting to congenital heart disease, a transthoracic echocardiography was performed. In echocardiography, a patent foramen ovale (PFO) with the right to left shunt, small ventricular septal defect (VSD), and tiny patent ductus arteriosus (PDA) were observed, which are the signs of heart failure. The observed heart defects in echocardiography did not justify heart failure in this patient; therefore, we decided to do brain and liver sonography for rolled/out AVM. In brain sonography, an echo-free zone in midbrain with very high turbulence flow vessel was observed that confirmed A-V malformation.

Conclusions: In the infancy, VGAM is usually presented with high cardiac output failure that can be associated with multiorgan dysfunction. Since the prenatal diagnosis of VGAM and its treatment will be associated with better prognosis, intrauterine detection of this anomaly and its treatment are recommended.

Keywords: Galen Vein, Arteriovenous Connection, Anomaly, Neonate, Heart Failure

1. Introduction

In congestive heart failure (CHF) with normal cardiac echocardiographic findings, the differential diagnosis should be considered, including infection, severe anemia, arrhythmias, peripheral shunt, arteriovenous malformations (AVM), etc.

Arteriovenous malformations (AVM) are abnormal connections between an artery and a vein whereby the interconnecting capillary network is missed (1). They frequently occur in midline regions of the brain (2). The incidence of cerebral arteriovenous malformations (CAVMs) amounts to 1.21 per 100,000 persons per year (3).

Vein of Galen is a short midline venous structure formed by the confluence of the two internal cerebral veins and the basal veins of Rosenthal; it is the caudal remnant of the median prosencephalic vein; a centrally located vessel that drains the choroid plexus. The Galenic vein is believed to be formed from the cerebral vasculature between 6 and 11 weeks of gestation (4).

The aneurismal malformation of the Galenic vein is a rare congenital cerebral arteriovenous anomaly with an incidence of 1: 25000 that comprises 30% of the pediatric vascular and 1% of all pediatric congenital anomalies (5, 6).

Its pathogenesis is unknown but findings suggest that an endoglin mutation might be linked with VGAM (6).

VGAM is developed between the 6 and 11 weeks of gestation and can be diagnosed in the prenatal period by ultrasonography (7). Moreover, it is accompanied by high output heart failure and sometimes it can present with an audible cranial bruit (6, 8).

The main presentations of these patients are accompanied with CHF. It is important to highlight the fact that not
all cases of cardiac failure may be due to a primary cardiac cause. An early diagnosis with a prompt referral is crucial in the management of this life-threatening condition.

Here, we report a patient with complications related to VGAM.

2. Case Presentation

A 9-day-old male neonate referred with icter, poor feeding, tachypnea, and tachycardia on 5th July 2016 and was admitted to the NICU of Vali-e-Asr specialized and subspecialized referral teaching university hospital, Birjand, Iran. The birth weight was 3650 grams. He was the first child, full term, normal vaginal delivery (NVD) with no family history of the disease, and the parents were not relatives.

In physical examination, the infant was icteric that spread out to thighs, tachypneic with respiratory distress, and subcostal retraction. His respiratory rate was 73 per minute and the heart rate was 160 per minute and blood pressure of the right hand was 79/55 mmHg, left hand 76/60 mmHg, right foot 75/48 mmHg, and left foot 79/46 mmHg measured with cardiac monitoring. A holo systolic murmur (grade 4/6) was heard in cardiac auscultation. Furthermore, a hepatomegaly was observed in the abdominal examination.

He was admitted to the NICU and his C-X-ray showed right-sided cardiomegaly (Figure 1). The findings of laboratory test showed increased bilirubin, alanine transferase (ALT), asparagine transferase (AST), and decreased serum albumin (ALB), indicating liver dysfunction (Table 1).

Table 1. Laboratory Data

| Variables | Value |
|-----------|-------|
| CBC       | WBC: 10.5, Hb: 10.3, Plt: 250 |
| Bilirubin | Total: 17, Direct: 0.5 |
| ABG       | PH: 7.3, Ptc2: 39.2, Po2: 98, HCo3: 23.2 |
| LFT       | ALT: 67, AST: 60 |
| Biochemistry | Blood suger: 95, Urea: 14, Creatinin: 1, Ca: 10.4, Na: 140, K: 5.2 |
| UA        | PH: 5, Nitrite: -, WBC: 0 - 1, RBC: 0 - 1, EP.cells: 2 - 3 |
| Serum protein | Total protein: 5, ALB: 2.1 |

Abbreviations: ABG, Arterial Blood Gas; Bilirubin, Total Bilirubin; CBC, Cell Blood Count; Hb, Hemoglobin; Hct, Hematocrit; LFT, Liver Function Test; Plt, Platelets; UA, Urine Analysis; WBC, White Blood Cells.

In echocardiography, a patent foramen oval (PFO) with the right to left shunt, small ventricular septal defect (VSD), and tiny patent ductus arteriosus (PDA) were observed, which are the signs of heart failure (Figure 2). The observed heart defects in echocardiography did not justify heart failure in this patient. Therefore, we decided to do brain and liver sonography and CT-angiography with contrast for rolled/out AVM.

In brain sonography, an echo-free zone in midbrain with very high turbulence flow vessel was observed that confirmed A-V malformation.

The contrast CT-angiography of the brain showed a hyperdense lesion in posterior cranial fossa in accordance with central vein whose differential diagnosis is VGAM (Figure 3).
On his 14th day of life, a neurointerventionist (neurosurgeon) visited the patient. The surgeon inserted a sheath in the right femoral artery under general anesthesia (GA) and intubation. The angiography was done by No. 3, 5 right Judkins catheter size of 4 French that showed fistulae of Galen vein supplied by both posterior choroidal arteries and drained directly into the vein of Galen (Figure 4). Then, after estimating the exact size of the fistula, a detachable coil accompanied with Onyx was deployed successfully. Control angiogram at straight lateral view showed a tiny residual shunt. By this technique, the neurosurgeon immobilized the VGAM.

Following up the patient at 3rd and 11th months after discharge in serial echocardiography revealed closed PDA, PFO, and VSD. Furthermore, in the 11th month, normal growth of head circumference (HC = 46 cm), weight (8.5 kg), height (75 cm) and good neural development were observed.

3. Discussion

Development of arteriovenous connection between primitive choroidal vessels and the median prosencephalic vein of Markowski caused the vein of Galen aneurysmal malformations as a rare congenital vascular anomaly (9).

It is estimated that 3% - 5% of intramedullary cavernous angioma is seen in central nervous system lesions (10). In the infancy, VGAM is usually presented with high cardiac output failure, which can be associated with multi-organ dysfunction. This anomaly can also cause space occupational lesion (SOE) that may lead to progressive neurological impairment and can present with cerebral ischemic changes (6). A systolic murmur, as the first clinical manifestation of high cardiac output failure induced by CAVM after birth, has been reported by Barajas-Gamboa et al. in Mexico (11). Similarly, in our study, cardiac murmur with CHF and elevated liver enzyme and creatinine were detected 9 days after birth.

A 2-day-old female neonate with VGAM like ours was reported by Shah Farhat et al. (2016) who had systolic ejection murmur (grade 3/6), cranial bruit accompanied by congestive heart failure and cyanosis but in contrast to our case, she had VSD and normal ABG (8).

In another study, Ramgren et al. (2016) reported a case of VGAM who was born full term with no symptoms at birth, but after 12 hours, the patient became tachypneic with a cardiac murmur. Cardiac ultrasonography revealed that the neonate patient had tricuspid regurgitation and a suspected cardiomyopathy. The VGAM of this patient was closed by coil similar to our case (12).

Congenital anomaly of Galen’s vein has a negative in-
The angiogram shows that injection of contrast media into right internal carotid artery drains into cerebral veins before endovascular treatment. (CV, Cerebral Vein; TS, Transverse Sinus; SS, Sigmoid Sinus; RICA, Right Internal Carotid Artery; RJC, Right Judkins Catheter and RJV, Right Jugular Vein).

fluence on the prenatal and postnatal development of the brain of newborns (13), but our case had a good weight gain and head circumference.

Merritt et al. reported the case of a 4-day-old girl with CHF and CAVM who died 9 days after birth (14). In another study, two cases of intrauterine CHF due to CAVM were reported by Cerbo et al. (15). Therefore, in the cases that have murmur and CHF with normal echocardiography findings, we must consider other differential diagnoses including sepsis, severe anemia, arrhythmia, and peripheral shunt such as CAVM.

In our case, the coexistence of ASD, VSD, and CAVM was observed, which is similar to a case reported by Hortobagy et al. concerning a 38-week-old neonate with CAVM (2).

Although the mortality of a fetus or neonate with VGAM is very high, the prognosis is dependent on the size of the malformation, age at diagnosis, and successful neurological outcome (9). Studies show that poor outcome is
associated with an early presentation in the neonatal period. Neonatal mortality following treatment ranges from 8% to 63% (11).

In McConnell et al.’s study, seven neonates were diagnosed with CAVMs all of whom presented with CHF. Three of them died immediately after birth because of severe CHF and the others died after endovascular embolization (16). However, in our case, endovascular embolization was done as soon as possible within 5 days after diagnosis and the infant currently is under the good general condition and gaining weight.

In spite of several sonographic anomaly scans, intrarteriuean VGAM and cardiomegaly had been missed.

Santo et al. have suggested that prenatal diagnosis must be done by color Doppler ultrasonography at 32 weeks of gestation for more precise diagnosis (17).

3.1. Conclusion

The findings suggest that an endothlin mutation might be linked with VGAM. Therefore, we must pay attention to familial arteriovascular anomalies such as the history of hemangioma in the family of neonates.

Now a day, if prenatal VGAM can be diagnosed by color Doppler ultrasonography during thirty-second weeks of gestation, and embolization is performed intrarteriueanly as soon as possible, the prognosis will be very good.

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Footnote

Conflict of Interest: The author of this article declares that there is no conflict of interest.

References

1. Riasi HR, Salehi F, Rajabpour Sanati A. Fluctuating neurologic symptoms in a patient with brain lesion. J Med Case. 2017;8(2):67–9. doi: 10.4240/jmcase.2706v.

2. Horrobagyi T, Szücs A, Csenki M, Harkany T, Zador Z, Katona M, et al. Vein of Galen malformation combined with atrial septal defect in a neonate. Clin Neuropathol. 2003;22(4):193–8. [PubMed: 12908756].

3. Daroff RB, Jankovic J, Mazzotti JA, Pomeroy SL. Bradley’s neurology in clinical practice. Elsevier Health Sciences; 2015.

4. Fayyaz A, Qureshi IA. Vein of Galen aneurysm: antenatal diagnosis: a case report. J Pak Med Assoc. 2005;55(10):455–6. [PubMed: 16304588].

5. Wagner MW, Vaught AJ, Poretti A, Blakmore KJ, Huisman TA. Vein of galen aneurysmal malformation: prognostic markers depicted on fetal MRI. Neuroimaging. 2015;20(1):72–5. doi: 10.15274/NRI-2014-01096. [PubMed: 25924777].

6. Tsutsui Y, Kosaki R, Itoyoh Y, Tsukamoto K, Matsuoka R, Shintani M, et al. Vein of Galen aneurysmal malformation associated with an endoglin gene mutation. Pediatrics. 2011;128(5):e1207–10. doi: 10.1542/peds.2010-10961. [PubMed: 21987708].

7. Raybaud CA, Strother CM, Hald JK. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology. 1989;31(2):165–28. doi: 10.1007/BF00698838. [PubMed: 2664571].

8. Shah Farhat A, Alizadeh Kasebi A, Khorakian F, Mohammadzadeh A, Saedi R. Vein of Galen aneurysmal malformation in a neonate: A case report. Iran J Neonatol. 2016;7(2):44–8.

9. De Beritto T, Khan OA, Hageman JR, Schreiber M. Vein of Galen arteriovenous malformation in a Neonate. Pediatr Ann. 2015;44(10):e243–6. doi: 10.3928/00904481-20151008-08. [PubMed: 26473426].

10. Shuhui G, Jiagang L, Siqing H, Haifeng C, Qingrong T, Bohao Z. Rare familial arteriovenous anomalies such as the history of postnatal arteriovenous malformation combined with atrial septal defect in a neonate. Clin Neuropathol. 2017;26(8):771–5. doi: 10.1007/s00009-017-0515-0. [PubMed: 28737112].

11. Barajas-Gamboa JS, Diaz-Perez JA, Leon-Camargo Y, Gonzalez-Gomez CA, Sandoval-Gomez C. Systolic heart murmur as first manifestation of high output heart failure due to the vein of galen malformation. Ann Pediatr Cardiol. 2012;5(3):214–7. doi: 10.1016/j.acmx.2012.04.002. [PubMed: 22936047].

12. Rasmussen B, Rask O, Gelberg J, Liuba P, Undren P, Wesslens J. Endovascular treatment of vein of Galen aneurysmal malformation using rapid ventricular pacing: A case report. Interv Neuroradiol. 2017;23(1):97–101. doi: 10.1177/1078680716676624. [PubMed: 27837112].

13. Brucknerova I, Kaldarova M, Gajdos M, Mach M, Dubovicky M, Ujha E. The influence of malformation of Galen vein on the cardiovascular system in a newborn. Neuro Endocrinol Lett. 2011;32 Suppl 1:S5–7. [PubMed: 22877233].

14. Merritt C, Feit JR, Valente JH. A neonate with high-outflow congestive heart failure and pulmonary hypertension due to an intracranial arteriovenous malformation. Pediatr Emerg Care. 2011;27(7):645–8. doi: 10.1097/PEC.0b013e3182225679. [PubMed: 21730802].

15. Cerbo RM, Cabano R, Radicioni M, Stronati M. Cerebral arteriovenous shunts regression in two preterm newborns with heart failure in utero. Am J Perinatol. 2009;26(9):637–9. doi: 10.1055/s-0029-1220795. [PubMed: 1939084].

16. McConnell ME, Aronin P, Virol J. Congestive heart failure in neonates due to intracranial arteriovenous malformations: endovascular treatment. Pediatr Cardiol. 1993;14(2):302–6. doi: 10.1007/BF00796988. [PubMed: 8469626].

17. Santo S, Pinto L, Clode N, Cardoso E, Marques JP, Melo A, et al. Prenatal ultrasonographic diagnosis of vein of Galen aneurysms-report of two cases. J Matern Fetal Neonatal Med. 2008;21(5):269–71. doi: 10.1080/14767050801924357. [PubMed: 18297576].