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Case Report

Vein of Galen Aneurysmal Malformation and High-output Cardiac Failure in a Newborn

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ABSTRACT

Background: Vein of Galen aneurysm (VGA) is the most common form of symptomatic cerebrovascular malformation in neonates. It develops in a fetus in the first trimester of pregnancy due to unknown reasons, but it is likely to have a genetic etiology. The prognosis of VGA is usually poor, particularly in newborns with heart failure due to high-flow intracerebral shunt. Surgery and endovascular embolization are partially successful treatments for controlling congestive heart failure and pulmonary hypertension.

Case report: In this article, we present the case of a 3600-gram, full-term, female neonate hospitalized with respiratory distress and severe heart failure. The neonate was the second child of a 28-year-old mother. After echocardiography and brain sonography, she was diagnosed with the vein of Galen malformation. The case had severe and persistent congestive heart failure and refractory pulmonary hypertension.

Conclusion: It is important to perform the auscultation of fontanel in newborns and consider VGA as a differential diagnosis in the neonates with congestive heart failure.

Keywords: Heart failure, Neonate, Vein of Galen aneurysm

Introduction

The vein of Galen is located under the cerebral hemispheres and drains the anterior and central regions of the brain into the sinuses of the posterior cerebral fossa. A vein of Galen malformation (VOGM) develops in a fetus in the first trimester of pregnancy due to unknown reasons, but it is likely to have a genetic etiology. The VOGM is a rare disorder characterized by abnormal connections between the vein of Galen and cerebral arteries (1) (Figure 1).

The VOGM is the most common form of symptomatic cerebrovascular malformation in neonates (2). This malformation can cause high-flow congestive heart failure and pulmonary hypertension, followed by multiorgan failure, by making high-flow intracerebral shunts.

Chest X-ray confirmed cardiomegaly, and echocardiography showed normal cardiac anatomy with dilated right atrium. Cranial sonography revealed a midline cystic mass

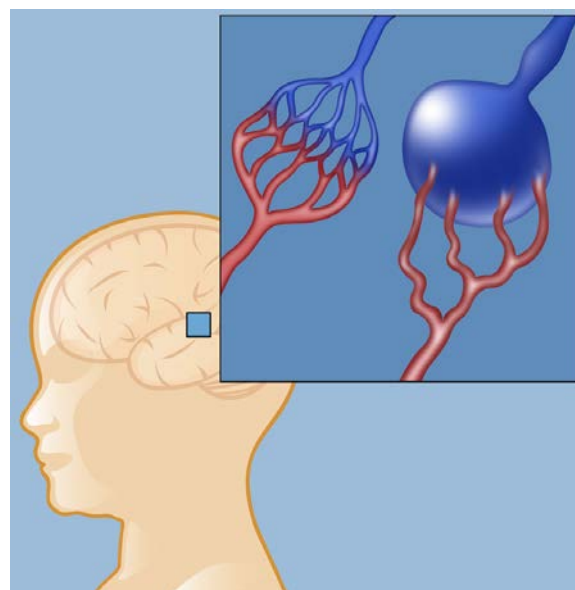


Figure 1. Normal and aneurysmal vein of Galen

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posterior to the third ventricle, and Doppler ultrasound showed a pulsatile cystic mass, suggestive of the arteriovenous malformation of the vein of Galen. Magnetic resonance imaging of the brain confirmed the diagnosis.

Over 90% of patients will die during the neonatal period without intervention (2). The management of these patients needs a multidisciplinary approach with the involvement of neurosurgeons, cardiologists, and neonatologists. In the past decades, these neonates were successfully treated with coil embolization. But surgery and arterial remobilization have been partially successful in controlling the intracerebral shunts in these newborns (3).

According to the literature, the early presentation of VOGM in the neonatal period is associated with poor outcomes and a neonatal mortality rate of 8-63% in case of receiving treatment. However, the mortality rate of this condition in children with a late presentation is 10% (4). In this article, we present a newborn hospitalized with respiratory distress, cardiomegaly, and cardiac murmur that was finally diagnosed with the vein of Galen aneurysm.

Case report

Our case was a 3600-gram, full-term, female neonate admitted to our Neonatal Intensive Care Unit with the diagnosis of respiratory distress when he was 5 days old. The newborn was the second child of a 28-year-old mother. On admission, the newborn had tachypnea, tachycardia, and cardiac murmur. She had a systemic blood pressure of 75/45 mmHg, a heart rate of 175 beats per min, a respiratory rate of 75 breaths/min, and oxygen saturation of 90% (while receiving 5 lit/min oxygen with a hood).

Her liver was palpable 2 cm below the rib cage, and she was hypotonic. But there were no dimorphic features. Her blood gas analysis revealed mixed acidosis. Significant cardiomegaly could be seen on her chest radiography. Echocardiography revealed ventricular septal defect, patent foramen ovale, and severe pulmonary hypertension with pulmonary arterial pressure of 80 mmHg, and insufficiency of the tricuspid and mitral valves.

Dopamine and Lasix were administered intravenously. Furthermore, sildenafil and milrinone were started for pulmonary hypertension, and antibiotic (e.g., ampicillin and gentamycin) were administered. However, respiratory distress got worsened, and oxygen saturation gradually decreased; therefore, the

patient was intubated and ventilated.

In spite of all treatments, her tachycardia continued, and the patient's condition deteriorated.

Therefore, she was subjected to another echocardiography, as well as transfontanelle and abdominal ultrasonographic (USG) examinations. The brain sonography demonstrated intracerebral shunt and a dilated, varicose, and persistent vein of Galen. Congestive heart failure was worsened rapidly, and she was referred to Razavi Hospital, Mashhad, Iran, to undergo an endovascular embolization.

There were many intracerebral fistulas, and the interventionist could embolize some of them. After remobilization, the vein of Galen was still fed by some fistulas.

Echocardiography revealed severe pulmonary hypertension, insufficiency of the tricuspid and mitral valves, and right atrial and ventricular dilation. Despite the strong supportive therapy, the patient developed a rapid multiorgan failure and was ventilated. However, the neonate died 10 days after embolization.

Discussion

Congenital heart disease accounts for one-third of all major congenital anomalies (5). The VOGM is a venous ectasia secondary to an arteriovenous fistula that can cause congestive heart failure (4). Although this malformation is a rare congenital disorder, it is the most common form of symptomatic cerebrovascular malformation in neonates and constitutes about 1% of all intracranial vascular malformations (6).

The VOGM develops between 6th-11th weeks of gestation (7) as a result of arteriovenous connections between the primitive choroidal vessels and the median prosencephalic vein of Markowski (8). Low systemic resistance in fetus can decrease the flow through the fistula and minimize the cardiac decompensation.

This condition can be diagnosed prenatally by antenatal Doppler USG. Evidence of progressive cardiac dysfunction in utero indicates a high-flow lesion (9). The most common presentation in the neonatal period is high-output cardiac failure and multiorgan dysfunction (10). The other presentations include high-flow congestive cardiac failure, intracranial hemorrhage, seizures, and hydrocephaly (11, 12).

The VOGM can sometimes cause a mass effect leading to progressive neurological impairments. It can also present with cerebral ischemic changes and seizure or result in hydrocephalus through

the obstruction of the cerebrospinal fluid flow. Early presentation of VOGM in the neonatal period is associated with a poor outcome and neonatal mortality rates of 8-63% and > 90% in case of the provision of treatment and lack of treatment, respectively. However, this rate is 10% in children with late presentation (4).

Neonatal diagnosis of VOGM is made by transfontanelle Doppler USG, computed tomography angiography, and magnetic resonance angiography. The cranial computed tomography and magnetic resonance angiography performed later confirmed the initial diagnosis of VOGM (12). Our patient could not be diagnosed prenatally because her prenatal screening was not complete.

The patient was referred to our hospital with severe respiratory distress and cardiomegaly indicated in the chest X-ray. At first, she was suspected of congenital heart disease, and a cardiac consultation was requested. But there was no cardiac defect, except cardiac failure, in echocardiography. Cardiologist noticed pulsation and fontanel murmur and observed several intracranial shunts and aneurysmal vein of Galen in the brain assessment.

Conclusion

In conclusion, clinical examination and auscultation of the anterior fontanel are simple and important examinations in neonates. The VOGM could be considered in infants with congestive heart failure. However, the main goal should be early even antenatal diagnosis and prompt management of this life-threatening condition.

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Conflicts of interests

The authors declared no conflicts of interest. No financial support was received for this article.

References

1. Porzionato A, Macchi V, Parenti A, De Caro R. Vein of Galen aneurysm: anatomical study of an adult autopsy case. *Clin Anat*. 2004; 17(6):458-62.
2. Patel N, Mills JF, Cheung MM, Loughnan PM. Systemic haemodynamics in infants with vein of Galen malformation: assessment and basis for therapy. *J Perinatol*. 2007; 27(7):460-3.
3. Berenstein A, Fifi JT, Niimi Y, Presti S, Ortiz R, Ghatan S, et al. Vein of Galen malformations in neonates: new management paradigms for improving outcomes. *Neurosurgery*. 2012; 70(5):1207-13.
4. Lasjaunias P, Ter Brugge K, Lopez Ibor L, Chiu M, Flodmark O, Chuang S, et al. The role of dural anomalies in vein of Galen aneurysms: report of six cases and review of the literature. *AJNR Am J Neuroradiol*. 1987; 8(2):185-92.
5. Saeidi R, Mohammadzadeh A, Farhat A, Naghibi MR, Birjandi H, Lotfi SR. Partial ectopia cordis: a case report. *Iran J Neonatol*. 2017; 8(3):78-81.
6. Kothari SS, Naik N, Juneja R, Saxena A. Aneurysm of the vein of Galen in neonates: report of four cases. *Indian Heart J*. 2001; 53(4):499-502.
7. Raybaud CA, Strother CM, Hald JK. Aneurysm of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. *Neuroradiology*. 1989; 31(2): 109-28.
8. Jones BV, Ball WS, Tomsick TA, Millard J, Crone KR. Vein of Galen aneurysmal malformation: diagnosis and treatment of 13 children with extended clinical follow-up. *AJNR Am J Neuroradiol*. 2002; 23(10): 1717-24.
9. McSweeney N, Brew S, Bhate S, Cox T, Roebuck DJ, Ganesan V. Management and outcome of vein of Galen malformation. *Arch Dis Child*. 2010; 95(11): 903-9.
10. Garcia-Monaco R, De Victor D, Mann C, Hannedouche A, Terbrugge K, Lasjaunias P. Congestive cardiac manifestations from cerebrocranial arteriovenous shunts. *Endovascular management in 30 children*. *Childs Nerv Syst*. 1991; 7(1):48-52.
11. Brunelle F. Arteriovenous malformation of the vein of Galen in children. *Pediatr Radiol*. 1997; 27(6):501-13.
12. Patton DJ, Fouron JC. Cerebral arteriovenous malformation: prenatal and postnatal central blood flow dynamics. *Pediatr Cardiol*. 1995; 16(3):141-4.