

Vein of Galen Aneurysmal Malformation in a Neonate: A Case Report

Ahmad Shah Farhat^{1*}, Abbas Alizadeh Kaseb², Fatemeh Khorakian³, Ashraf Mohammadzadeh⁴, Reza Saeidi⁵

1. Assistant Professor of Neonatology, Neonatal Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

2. Neonatologist, Hashemi Nejad Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

3. Assistant Professor of Pediatric Dentistry, Dental Research Center, School of Dentistry, Mashhad University of Medical Sciences, Mashhad, Iran

4. Professor of Neonatology, Neonatal Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

5. Associate Professor of Neonatology, Neonatal Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

ABSTRACT

Background: Vein of Galen aneurysmal malformation (VGAM) is a rare congenital malformation, accounting for less than 1% of cerebrovascular abnormalities. The majority of reported cases have been associated with congestive heart failure (CHF) in the neonatal period. Herein, we present a case of VGAM, diagnosed at 37 weeks of gestation during the intrauterine life

Case report: A full-term female newborn presented with severe CHF at two days of age. The patient's peripheral pulses were bounding in all four extremities. The first heart sound appeared to be normal, while the second had an accentuated pulmonic component. A systolic murmur (grade 3/6), best heard in the pulmonary area, was reported, and a cranial bruit was sought and found. The echocardiography showed evidence of right ventricular hypertrophy, while the chest X-ray indicated cardiomegaly with increased pulmonary vascularity. Moreover, echocardiography revealed dilation of the right heart chambers, a patent foramen ovale, severe tricuspid regurgitation, as well as a moderate-sized secundum atrial septal defect and a patent ductus arteriosus with right-to-left shunting. Transcranial ultrasound and contrast-enhanced CT scan of the brain detected a vein of Galen malformation. Magnetic resonance venography confirmed VGAM and identified the vessels feeding the aneurysm. Postnatal management included aggressive medical treatment of CHF. Transarterial embolization of the vessels feeding the aneurysm was suggested. However, the newborn succumbed to her disease on the following day.

Conclusion: VGAM, which is a rare cause of cyanosis and heart failure in newborns, can be clinically diagnosed via proper strategies. However, extensive distribution of aneurysm usually precludes surgical management and endovascular treatment.

Keywords: Aneurysm, Heart failure, Neonates

Introduction

Vein of Galen aneurysmal malformation (VGAM) was first described by Steinhel in 1895 (1). VGAM is a rare congenital, cerebral, arteriovenous abnormality with an incidence of 1:25000. This vascular malformation arises from multiple arterial feeders, establishing direct or indirect shunts with a large median venous collector. VGAM is developed between the sixth and eleventh week of gestation and can be diagnosed prenatally via ultrasonography (2); however, this malformation is most often detected in the postnatal period (3).

VGAM classically presents in the neonatal period and is accompanied with high-output heart

failure and sometimes an audible cranial bruit; severe pulmonary hypertension may also complicate the heart failure. In the past, mortality of neonates with VGAM, presenting with heart failure, was estimated at 100%. However, today, endovascular treatment has been introduced as the treatment of choice for patients with neonatal VGAM, presenting with heart failure.

Embolization of both feeding arteries and draining veins can result in a decline in blood flow and a higher survival rate in infants with VGAM. In fact, reduced neuroembolism results in the termination of excessive blood flow through VGAM, which is the key to improved cardiac

* Corresponding author: Ahmad Shah Farhat, Neonatal Research Center, Imam Reza Hospital, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran, E-mail: farhata@mums.ac.ir

function and brain injury prevention.

Multiple shunts of brain arteries contribute to the aneurysmatic enlargement of the great unpaired cerebral vein, also known as the vein of Galen. The symptoms of VGAM vary, depending on the patient's age and anatomy and angioarchitectural features of the malformation. In newborns (choroidal VGAM), volume overload is often dominating, suggesting raised cardiac output per minute, tachycardia, cardiomegaly, cardiac insufficiency (high-output heart failure), pulmonary hypertension, respiratory distress syndrome, pulmonary edema, and multiple organ failure.

Mural VGAM is commonly reported during infancy and consists of fewer but larger shunts as direct arteriovenous fistulas in the wall of the median prosencephalic vein of Markowski. The symptoms include hydrocephalus, macrocrania, and developmental retardation. On the other hand, epileptic seizures causing damage to the brain are rarely seen in VGAM.

In different regions of the world, clinical presentations of VGAM may deviate from the general descriptions in the literature (4). If left untreated, mortality of newborns and infants with severe cardiac insufficiency can amount to 100% and 72%, respectively (5). Endovascular embolization, which primarily includes transarterial glue embolization, is regarded as the safest and most efficient targeted treatment for VGAM (6-8). On the other hand, in other therapeutic methods, e.g., transvenous embolization and surgical intervention, higher rates of complications and mortality are described (9). Overall, time and method of endovascular embolization depend on the patient's clinical signs and symptoms (3, 10).

Transfontanelar Doppler sonography is a non-invasive technique, enabling bedside evaluation of the cerebrovascular system in neonates. Once a diagnosis is established, further evaluations such as



Figure 1. Contrast-enhanced CT scan of the brain

Computed tomography (CT), CT angiography, magnetic resonance imaging (MRI), and angiography may be also helpful if the neonate is stable. CT scan is usually performed to identify ventricular enlargement, brain atrophy, and parenchymal calcifications, associated with severe intracranial hypertension and brain injury. Newborns with large VGAM shunts may also suffer from early ischemic brain injury; consequently, they are not eligible candidates for repair.

Heart failure in newborns may originate from the uterus. Several prenatal echocardiographic studies have revealed an increased flow through the pulmonary artery and a retrograde diastolic flow through the aortic isthmus. In some case reports, infants with concomitant pulmonary hypertension showed signs of pulmonary hypertension on prenatal echocardiography. Excessive pulmonary blood flow occurs as the pulmonary flow far exceeds the dilatory capacity of the ductus arteriosus. Fetal ultrasound imaging has now enabled the identification of VGAM in the fetus, allowing expectant delivery in tertiary care centers to treat neonatal arteriovenous malformations.

Case report

A full-term female neonate (born at 38 weeks of gestation) was born through cesarean section. The patient's Apgar score was calculated to be 8-9 and her birth weight was 3,340 g. The mother was 28 years old and had experienced a normal pregnancy.

Physical examination and chest X-ray results of the newborn were normal after birth. However, she presented with tachypnea, severe congestive heart failure (CHF), and mild cyanosis at two days of age. On the first venous blood gasometry,

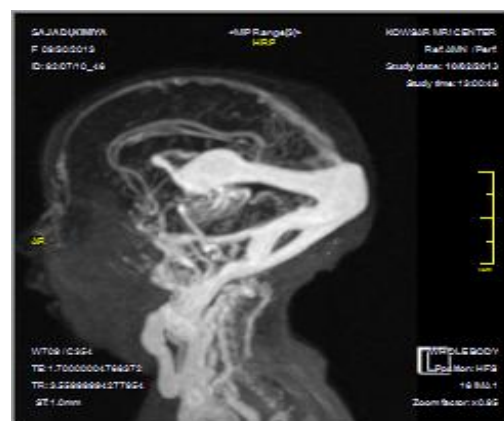


Figure 2. Magnetic resonance venography of the brain

pH=7.31, PCO₂=42, HCO₃=21, PO₂=35, and O₂sat=62 were reported. The patient's peripheral pulses were bounding in all four extremities. The first heart sound was normal, while the second had an accentuated pulmonic component.

A systolic murmur (grade 3/6), which was best heard in the pulmonary area, was reported, and a cranial bruit was sought and found. Echocardiography showed evidence of right ventricular hypertrophy, while the chest X-ray showed cardiomegaly with increased pulmonary vascularity.

Transthoracic echocardiography revealed severe pulmonary hypertension, dilated right heart chambers, a patent foramen ovale, severe tricuspid regurgitation, as well as a moderate-sized secundum atrial septal defect (ASD2) and a patent ductus arteriosus (PDA) with right-to-left shunting. Transcranial ultrasound and contrast-enhanced CT scan of the brain revealed a vein of Galen malformation (Figure 1). Magnetic resonance venography confirmed VGAM and demonstrated the vessels feeding the aneurysm (Figure 2).

Postnatal management included aggressive medical treatment of CHD. Transarterial embolization of the vessels feeding the aneurysm was suggested. CHD, unresponsive to dopamine and dobutamine (up to a dose of 10 µg/kg/min), was treated with a phosphodiesterase inhibitor (0.4–0.75 µg/kg/min of milrinone). The patient required mechanical ventilation to improve respiratory acidosis and cyanosis. However, she succumbed to her disease on the following day.

Discussion

In this case report, the patient was diagnosed at 37 weeks of gestation, based on the prenatal sonography. The findings were as follows: amniotic fluid index (AFI): 20 cm, fetal heart rate (FHR): 152 bpm, weight: 3450±300 g, VGAM with a dilated straight sinus (diameter: 7.7 mm), dilated lateral ventricle, cardiomegaly (with right ventricular and atrial dilation), and mild tricuspid regurgitation (without heart failure); it should be noted that fetal ultrasound images were not presented by the parents.

The patient was referred to a pediatric cardiologist, and prenatal echocardiography showed an increased cardiothoracic ratio, PFO, ASD2, regular rhythm, and dilation of the right ventricle and right atrium, indicating right atrial and right ventricular pressure or volume overload. However, fetal echocardiographic images were not provided in the present case.

The patient was referred to Shiraz Neurosurgery Hospital for further treatment and vascular embolization. However, the patient's transfer to the center failed due to premature labor, and she was born at Imam Reza Hospital of Mashhad, Iran. The newborn was admitted to the hospital for further evaluations. On the first day, she presented with mild tachypnea. On the second day, she appeared sick and was intubated due to cyanosis and respiratory acidosis.

Brain CT scan and magnetic resonance venography were performed on the patient. On the third day, symptoms of right-sided heart failure appeared, followed by the exacerbation of left-sided heart failure, which showed poor response to treatment with dopamine, dobutamine, or milrinone. Given the patient's poor condition, inadequacy of time, and long distance, patient transfer to a specialized surgical center was impossible. By the end of the day, the patient died due to heart failure.

During the neonatal period, symptomatic infants with VGAM present with severe cardiorespiratory alterations at birth or shortly after it; the majority of cases (94%) have high-output cardiac failure (11, 12). Moreover, severe pulmonary hypertension of the newborn (PPHN) may be a complicating factor. Since newborns classically develop CHF as a result of a significant left-to-right shunting through the low-resistance vascular bed of VGAM, symptomatic infants often appear cyanotic due to right ventricular volume and pressure overload (13).

Echocardiography plays an essential role in estimating the ventricular function, shunting across PDA and ASD2, and pressure of the right ventricle and pulmonary artery. In addition, it helps identify the associated cardiac conditions in infants with VGAM and reveal the diastolic reverse flow, which is also known as the "steal" phenomenon, reducing peripheral perfusion (11, 13).

CHD and pulmonary hypertension are the most remarkable and challenging concerns in medical stabilization of symptomatic neonates. Nitric oxide therapy, as the most effective treatment for pulmonary hypertension, has limited success in infants with VGAM and PPHN (13). While optimal strategies are yet to be clearly defined, diuretic and inotropic therapies may be successful in deferring endovascular treatment for these patients. However, management of patients diagnosed with concomitant pulmonary hypertension is demanding and warrants serial brain natriuretic peptide (BNP) tests to monitor the clinical course.

Without interventional neuroradiology or embolization, the prognosis is poor for neonates whose primary presentation is early and severe heart failure, secondary to VGAM. Severe heart failure rapidly progresses to multiple organ failure and death (11). It has been suggested that VGAM-related heart failure, associated with multiple organ failure in the neonatal period, is linked with a high rate of cerebral infarction and thus poor prognosis due to long-term neurodevelopmental outcomes (14, 15); accordingly, endovascular treatment is withheld for such newborns. In the present case, intervention was not discarded for the neonate solely on the basis of severe CHF.

From our perspective, severe cerebral injury may occur unless the excess flow through the arteriovenous malformation is reduced by early neurointervention and aggressive management of CHF. The endovascular treatment of VGAM often requires several successive procedures. Staged embolization sessions initially aim at controlling heart failure in order to avoid the occurrence of parenchymal bleeding or massive venous thrombosis, which potentially compromises the normal venous drainage. Monitoring beyond the neonatal period includes checking for increased head circumference, hydrocephalus, seizures, and developmental delays.

The associated psychomotor disabilities often arise from the cerebral steal phenomenon (13). Advances in endovascular techniques and perinatal management have led to more favorable long-term outcomes (13, 16). Based a 30-month follow-up study, 55% of patients were functionally normal (16). Although the mortality rate of this population has been previously estimated at 100% (17), more recent studies have indicated a mortality rate of 9-15% and little to no neuromorbidity in 61-66% of survivors (18, 19).

Rapid diagnosis and treatment of infants with complex conditions are essential for accelerating appropriate treatments in order to prevent long-term adverse outcomes. Incorporation of serial BNP measurement in patient assessment may aid neonatal management and promote the outcomes in patients with complex conditions.

It is recommended that neonates with VGAM and heart failure be accurately and safely diagnosed and classified, using prenatal ultrasonography and MRI. When heart failure cannot be controlled by medical therapy, various endovascular techniques can allow safe and successful treatment for the infants. In fact, with proper classification of patients, favorable clinical outcomes can be achieved. Overall, detailed

ultrasonography in the third trimester of pregnancy, aneurysm detection, and timely referral of patients to surgical centers are highly recommended.

Conclusion

Aneurysm of the vein of Galen, which is a rare cause of cyanosis and heart failure in newborns, can be diagnosed via proper strategies. However, the extensive distribution of aneurysm usually precludes surgical management or endovascular therapy

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