Neonate With Severe Heart Failure Related to Vein of Galen Malformation

Meng-Yu Chen¹, Hon-Man Liu², Wen-Chin Weng¹, Shinn-Forng Peng², En-Ting Wu¹, Shuenn-Nan Chiu¹*

¹Department of Pediatrics, National Taiwan University Hospital and Medical College, National Taiwan University, Taipei, Taiwan
²Department of Medical Imaging, National Taiwan University Hospital and Medical College, National Taiwan University, Taipei, Taiwan

Received: Jun 17, 2009
Revised: Aug 13, 2009
Accepted: Sep 4, 2009

KEY WORDS:
heart failure; neonates; therapeutic embolization; vein of Galen malformation

1. Introduction

Vein of Galen aneurysmal malformation (VGAM) is a rare congenital abnormality, with a prevalence of less than 1/25,000 deliveries.¹,² It is characterized by a midline intracranial vascular fistula with aneurysmal dilation of the vein of Galen. The typical presenting features in neonates usually include signs of high-output heart failure. In the past, this might have progressed rapidly to multiple organ failure and death. However, aggressive medical treatment of heart failure and urgent endovascular intervention has produced encouraging results, although early diagnosis remains a challenge. We report a neonate with severe heart failure secondary to VGAM successfully treated with aggressive intensive care and urgent embolization.

2. Case Report

This full-term female baby was born to a 26-year-old mother by cesarean section, because of macrosomia; the birth body weight was 3850g. Her mother had experienced an uncomplicated pregnancy, except for an attack of cholecystitis 2 weeks before delivery, which was treated with antimicrobial therapy. The delivery was uneventful, but the baby was noted to have respiratory distress, feeding intolerance and an ill-looking appearance 1 day after birth at a local hospital. A chest radiograph showed significant cardiomegaly. Transthoracic echocardiography revealed dilation of all four chambers, patent ductus arteriosus (PDA) and a moderate degree of tricuspid and mitral regurgitation. Heart failure secondary to perinatal infection was initially suspected.
The patient was treated with inotropes, diuretics, and empirical antibiotics, in combination with nasal continuous positive airway ventilation for respiratory support. Brain ultrasound revealed a large hypoechogenic cystic lesion communicating with the right quadrigeminal cistern, and an arachnoid cyst was tentatively diagnosed. Progressive respiratory distress with cyanosis was noted on the 10th day of life. The patient was intubated and then transferred to the neonatal intensive care unit at the National Taiwan University Hospital.

Upon admission, physical examination revealed a heart rate of 150 beats/min, respiratory rate of 35 breaths/min under mechanical ventilation, and stable blood pressure under inotropic agents. Other findings included acrocyanosis, hyperdynamic precordium with grade II/VI systolic murmur at the left sternal border, and hepatomegaly (3 cm below the right costal margin at the midclavicular line). An electrocardiogram displayed sinus tachycardia without significant ST-T segment change. Laboratory data showed hemoglobin 115 g/L, white cell count $9.980 \times 10^9/L$ and platelet count $62 \times 10^9/L$. Biochemical studies showed impaired hepatic and renal function (alanine transaminase, $5.56 \mu$kat/L (333 U/L); total bilirubin, $303.2 \mu$mol/L; direct bilirubin, $140.7 \mu$mol/L; and serum creatinine, $88.4 \mu$mol/L (1.0 mg/dL)), elevated cardiac enzymes (CK-MB, 104.1 U/L; troponin-I, 8.25 ng/mL) and coagulopathy (prothrombin time, 18.5 seconds). Serum lactate was 10.1 mmol/L, and B-type natriuretic peptide (BNP) was 15,036 pg/mL. Serology tests for Coxsackie virus antibody and pan-enterovirus polymerase chain reaction analysis of throat swabs were negative. Echocardiography demonstrated dilation of all four chambers with normal left ventricular function (estimated ejection fraction, 67%). There were also left-to-right shunts through the PDA and foramen ovale, and a moderate degree of tricuspid regurgitation (flow velocity, 3.28 m/s). The infant was weaned from mechanical ventilation and inotropic agents on the 17th day of life, but tachypnea was still present. Repeat echocardiography showed an almost closed PDA, but aortic runoff with reversal of flow during diastole in the proximal descending thoracic aorta persisted. Continuous and prominent carotid flow was also noted (Figure 1A), with prominent superior vena cava flow. Extracardiac shunting because of a vascular lesion in the head or neck region was strongly suspected. Repeat brain ultrasound with color Doppler revealed flow voids within a large cystic lesion posterior to the right lateral ventricle (Figure 1B), and cranial bruit could be auscultated over the whole skull. Brain magnetic resonance imaging and angiography showed aneurysmal enlargement of the vein of Galen with multiple feeding arteries and engorged posterior draining venous sinuses (Figures 2A and 2B). The right lateral ventricular trigone was compressed, but the cerebral parenchyma was otherwise normal. This was consistent with a large VGAM. Transarterial embolization was performed at 24 days of age, and angiography demonstrated multiple choroidal feeders (Figure 3A). Under general anesthesia, a 0.018-inch guidewire was positioned via a femoral approach, within a 4-Fr guiding catheter (Optitorque Hinck Headhunter 1; Terumo Co., Somerset, NJ, USA). A 0.014-inch Transend Ex Floppy microguidewire (Boston Scientific, Fremont, CA, USA) was then navigated through a 1.7-Fr Excelsior SL-10 microcatheter (Boston Scientific) to the branches from the bilateral anterior choroidal arteries. A total of 33 Guglielmi detachable coils (GDC; Boston Scientific) were placed in the parent arteries adjoining the fistula (Figure 3B). Postembolization angiograms showed a small residual shunt with smaller venous sac and blood flow, and 80% obliteration was achieved (Figure 3C). Follow-up laboratory tests after coiling, including liver enzymes and BNP, showed normal results (BNP, 89.48 pg/mL). A chest radiograph showed normal cardiac size, while echocardiography showed

**Figure 1** (A) Suprasternal notch view with color Doppler shows flow reversal (arrow) in the proximal descending thoracic aorta during diastolic phase. (B) Head sonography in sagittal plane shows a large cystic lesion ($2.5 \times 1.8$ cm) connecting to the quadrigeminal cistern, corresponding to a vein of Galen malformation (G).
Newborn with heart failure

Figure 2  (A,B) Transverse (T1) and sagittal (T2) magnetic resonance images before embolization show a markedly dilated vein of Galen (G) near the right lateral ventricle connecting to the quadrigeminal cistern with lateral ventricle trigone compression. (C) Coronal angiography shows multiple dilated choroidal artery (CA) feeders and a large dilated vein of Galen. Engorged posterior draining venous sinus (S) is also shown.

Figure 3  (A) From transcutaneous femoral artery access, the left carotid angiogram with lateral projection before embolization demonstrates a venous sac with multiple choroidal feeders. (B) The right carotid angiogram with lateral projection after embolization shows coil mass within the aneurysmal component. Residual shunt with smaller venous sac and relative decrease in blood flow from feeding artery is also shown. LCA=left carotid artery; G=vein of Galen malformation; P=platinum coil.

resolution of aortic runoff. Brain sonography demonstrated reduced blood void in the aneurysm, with no evidence of hemorrhagic lesions. Neurological examination revealed only mild hypertonia of the lower extremities. The infant was discharged with oral diuretics on her 35th day of life.

3. Discussion

The most common causes of neonatal congestive heart failure are structural anomalies, such as large left-to-right shunting or left-sided obstructive lesions. However, diagnosis in patients with heart failure and a structurally normal heart remains a medical challenge. The common etiologies include myocardial dysfunction, incessant arrhythmia, or high-output failure. Myocardial dysfunction is commonly related to perinatal viral infection or hypoxic insult. Regarding arrhythmias, both tachyarrhythmia and bradyarrhythmia can compromise cardiac output and cause heart failure. Simple electrocardiography and echocardiography can quickly differentiate among these three conditions. In cases of high-output failure, cardiac output is normal or increased, but oxygen delivery is inadequate for the whole-body needs. This is commonly seen in anemia and large systemic arteriovenous fistulas. In the
latter, the reduced afterload due to abnormal vascularity, along with high preload from the increased venous return from the shunt, results in increased cardiac loading. The two most common lesion sites for systemic arteriovenous malformations are the liver and the brain. Hepatic arteriovenous malformations are hard to miss, because they can be easily palpated and auscultated on physical examination. Although brain sonography is a common screening tool in sick neonates, the results of cranial auscultation can be missed, as in the current patient; the large cystic lesion was initially mistaken for an arachnoid cyst, though the retrograde diastolic “aortic runoff” flow demonstrated on echocardiography suggested an intracranial vascular abnormality.

VGAM is a choroidal type of arteriovenous malformation that develops from the embryonic median vein of the prosencephalon. In the neonatal stage, VGAM has been reported to manifest initially as congestive heart failure, with or without pulmonary hypertension. Without intervention, severe heart failure develops and rapidly progresses to multiple organ failure and death. Poor hemodynamic prognostic factors include severe suprasystemic pulmonary artery hypertension, cardiogenic shock, PDA with a significant right-to-left shunt, a descending aortic diastolic reverse flow, and other parameters mimicking persistent fetal circulation. The need for mechanical ventilation is also a predictor of poor neurodevelopmental outcome. Lasjaunias et al developed a scoring system, which provides a useful guide for treatment strategies; our patient scored 9 out of 21, indicating the need for urgent endovascular intervention.

Serial changes in plasma BNP level have been reported to mirror the clinical congestive heart failure status. BNP is a useful adjunct to the assessment of treatment effect and further follow-up. In the current case, the patient’s plasma BNP level fell markedly from 15,036 pg/mL to 89.48 pg/mL after endovascular embolization. Although several poor prognostic factors for survival and neurodevelopment were present in our patient, the first endovascular treatment produced encouraging results.

VGAM is a rare anomaly, and only two previous cases have been reported in Taiwan. These two cases presented with similar symptoms to our patient, including intractable heart failure, pulmonary artery hypertension, and respiratory distress soon after birth. One received conservative treatment because the family refused endovascular intervention. Coil embolization of the aneurysm was attempted in another case, as early as 6 days of life, but failed to reduce the pulmonary arterial pressure and reverse the heart failure. It has been previously documented that successful embolization does not necessarily affect the pulmonary arterial pressure, which only improved in the survivors. Thus, the disappearance of pulmonary hypertension after embolization could be a major predictor of survival. Moreover, endovascular correction of the anatomic anomaly does not ensure normal neurodevelopment. Because complete occlusion was not achieved in our patient, repeat endovascular embolization will be required if cardiac or respiratory failure recurs, and assessment of the long-term neurodevelopmental outcome awaits further clinical follow-up.

In conclusion, the diagnosis of VGAM should be considered in neonates with congestive heart failure, especially in cases with high-output heart failure and a structurally normal heart. Aggressive medical treatment can stabilize heart failure symptoms, and urgent transarterial embolization leads to hemodynamic recovery and survival.

References