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Case report / Olgu Sunumu

Vein of Galen aneurysm in a newborn baby

Yenidoğanda Galen ven anevrizması

Şebnem Çalkavur,[1] Özgür Olukman,[1] Nuh Yılmaz,[1] Vedide Tavlı,[2] Mustafa Parıldar,[3] Füsun Atlıhan,[1] Nilgün Kültürsay[4]

[1]Department of Neonatology, Dr. Behçet Uz Children's Hospital, İzmir, Turkey;
[2]Department of Pediatric Cardiology, Dr. Behçet Uz Children's Hospital, İzmir, Turkey;
[3]Department of Interventional Radiology, Medicine Faculty of Ege University, İzmir, Turkey;
[4]Department of Neonatology, Medicine Faculty of Ege University, İzmir, Turkey

Although vein of Galen aneurysm is a rare pathology, it is the most common form of symptomatic cerebrovascular malformation in neonates and infants. Advanced high flow intracerebral shunt may cause high-flow congestive heart failure depending on an increase in the volume accompanied by different levels of pulmonary hypertension. Unfortunately, the prognosis is poor particularly in newborns with heart failure findings. Surgery and arterial embolization are only partially successful in controlling flow through Galenic fistulas. However, in parallel to the improvements in endovascular embolization techniques and the innovations in the newborn intensive care practices, major developments in controlling congestive heart failure and pulmonary hypertension have been achieved. In this article, we report a newborn case who presented with severe high-flow congestive heart failure and was diagnosed with the vein of Galen malformation. The case was found worth presenting to remind clinicians the vein of Galen malformation in the differential diagnosis of severe congestive heart failure and persistent, treatment refractory pulmonary hypertension at systemic/suprasystemic level during neonatal period. It is also of utmost importance to emphasize the role of embolization into abnormal feeding arteries as an alternative treatment of choice.

Key words: Congestive heart failure; newborn; vein of Galen aneurysm.

Galen ven anevrizması nadir bir patoloji olmasına karşın, yenidoğan ve infantlardaki semptomatik serebrovasküler malformasyonların en sık formudur. Aşırı yüksek akımlı intraserebral şant, pulmoner hipertansiyonun farklı dereceleri ile beraber olan hacim artışına bağlı yüksek debili kalp yetmezliğine neden olabilir. Prognoz, ne yazık ki özellikle kalp yetmezliği bulgusu ile gelen yenidoğanlarda kötüdür. Cerrahi ve arteriyel embolizasyon Galenik fistüller yoluyla olan akımı kontrol etmede yalnızca kısmen başarılıdır. Bununla birlikte, endovasküler embolizasyon tekniklerindeki gelişmeler ve yenidoğan yoğun bakım uygulamalarında yeniliklere paralel olarak, pulmoner hipertansiyon ve konjestif kalp yetmezliğini kontrol etmede önemli gelişmeler kaydedilmektedir. Bu makalede, şiddetli yüksek debili konjestif kalp yetmezliği tablosu ile başvuran ve Galen ven anevrizması tanısı konulan bir yenidoğan olgusu sunuldu. Yenidoğan döneminde şiddetli konjestif kalp yetmezliği ve tedaviye dirençli persistan sistemik/suprasistemik düzeyde pulmoner hipertansiyonun ayırıcı tanısında Galen ven malformasyonunun akılda bulundurulması amacıyla bu olgu sunuldu. Ayrıca, alternatif tedavi seçeneği olarak anormal besleyici arterlerin embolizasyonunun vurgulanması için de önem teşkil etmektedir.

Anahtar sözcükler: Konjestif kalp yetmezliği; yenidoğan; Galen ven anevrizması.

The vein of Galen aneurysmal malformation (VGAM) is a rare cerebrovascular disorder characterized by abnormal direct communication between one or several cerebral arteries and the vein of Galen.[1] It is the most common form of symptomatic cerebrovascular malformation in neonates and infants. Depending on the increase in volume accompanied by different levels of pulmonary hypertension, advanced high-flow

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Correspondence: Şebnem Çalkavur, M.D. Dr. Behçet Uz Çocuk Hastanesi Neonatoloji Kliniği, 35210 Konak, İzmir, Turkey.
Tel: +90 232 - 411 61 35 e-mail: sebnemcalkavur@yahoo.com

intracerebral shunts can cause high-flow congestive heart failure. Mortality is especially high during the neonatal period.[2] Without intervention, over 90% of these babies progress rapidly to multiple organ failure resulting in death. Surgery and arterial embolization have only been partially successful in controlling flow through Galenic fistulas. In the past 10 years, these patients have been successfully treated with embolic coils.[3] In this report, we present a newborn baby who presented with severe high-flow congestive heart failure and was diagnosed with VGAM.

CASE REPORT

A 2550-gram, full-term, male baby born from the third pregnancy of a 26-year-old mother via normal spontaneous vaginal delivery with Apgar scores of 5/8 at one and five minutes respectively was admitted to our neonatal intensive care unit (ICU) with the diagnosis of respiratory distress. On admission, the baby was edematous and was experiencing subcostal-intercostal retractions along with tachypnea, tachycardia, and impaired peripheral circulation as well as preconvulsive movements. His anthropometric measurements were between the 10th and 50th percentiles. It was noted that he had a systemic blood pressure rate of 65/35 (mean 41) mmHg, a heart rate of 160/min, a respiratory rate of 82 breaths/min, oxygen saturation of 85% (while receiving 5 lt/min oxygen with a hood), and a temperature of 36.8 ºC (axillary). No murmur or additional heart sound was audible on cardiac auscultation. Widespread fine crackles were auscultated over both lungs. His liver was 2 cm palpable on the midclavicular line. The newborn reflexes were weakened, and the baby was remarkably hypotonic. He did not have any dysmorphic features.

His blood gas analysis revealed compensated metabolic acidosis with a pH: 7.33, pCO2: 22 mmHg, pO2: 63 mmHg, HCO3: 12 mEq/l, and BE: -13. No significant pathology could be seen on his chest radiography. An echocardiographic (ECHO) examination revealed severe pulmonary hypertension with pulmonary arterial pressure (PAP) of 65 mmHg, insufficiency of the tricuspid and mitral valves, patent ductus arteriosus (PDA), and patent foramen ovale (PFO). Biochemical studies showed mildly impaired hepatic and renal function tests (blood urea nitrogen: 48 mg/dL, serum creatinine: 1.3 mg/dl, alanine transaminase: 90 IU/L), and slightly elevated creatinine kinase levels (CK-MB 98 IU/L). Inotropic and anticongestive treatment with dopamine, dobutamine, and spironolactone was administered immediately. Intravenous magnesium sulphate infusion was started for pulmonary hypertension, and a wide spectrum antibiotic combination was arranged with ampicillin and netilmicin. On his follow-up, his respiratory distress worsened, and his arterial oxygen saturation gradually decreased. The baby was intubated and started on mechanical ventilation after detecting decompansated metabolic acidosis on blood gas analysis. Despite all treatments, his tachycardia continued, and his hepatomegaly increased. Therefore, sildenafil was started and a furosemide infusion was added to his existing therapy after repeating his ECHO. In order to eliminate the reasons for high-flow congestive heart failure other than cardiac malformations, transfontanel and abdominal ultrasonographic (USG) examinations were performed. After demonstrating a suspicious vascular anomaly on his transfontanel USG and auscultating a continuous murmur over the baby's anterior fontanelle, contrast-enhanced axial computed tomography (CT) scanning of the brain demonstrated a dilated, varicose, persistent porencephalic vein (Vein of Galen) with a diameter of 1.5 cm at the posterior of the third ventricle, lateral and third ventricular dilation, periventricular white matter hypodensities, and secondary encephalomalacic changes. The baby's clinical findings related to his congestive heart failure worsened rapidly, and he was referred to the invasive radiology department of a university hospital in order to perform an endovascular embolization. Magnetic resonance (MR) imaging and MR angiography confirmed VGAM and compression to the lateral and third ventricle (Figures 1 and 2). Catheterization of the umbilical artery could not be performed because the baby was only 10 days old. Therefore, he was catheterized under general anesthesia via the right internal carotid artery. This branch was catheterized by using a superselective microcatheter, and it was embolized over the fistula level first from the right and then from the left side with N-butyl-cyanoacrylate (NBCA). The control angiography performed after embolization demonstrated that the vein of Galen was still being fed by the left vertebral artery (Figure 3). Only two holes were embolized in order not to exceed the contrast agent dose that the baby could tolerate. Postembolization ECHO revealed severe pulmonary hypertension, insufficiency of the tricuspid and mitral valves, and right atrial and ventricular dilation. His existing treatment was continued, and his postangiography thyroid function tests were completely normal. However, the patient developed a rapid multiorgan failure, including renal failure, hepatic failure, and grade 1 necrotizing enterocolitis after embolization. Despite strong supportive therapy, the baby died four days after embolization on his 14th postnatal day of life.

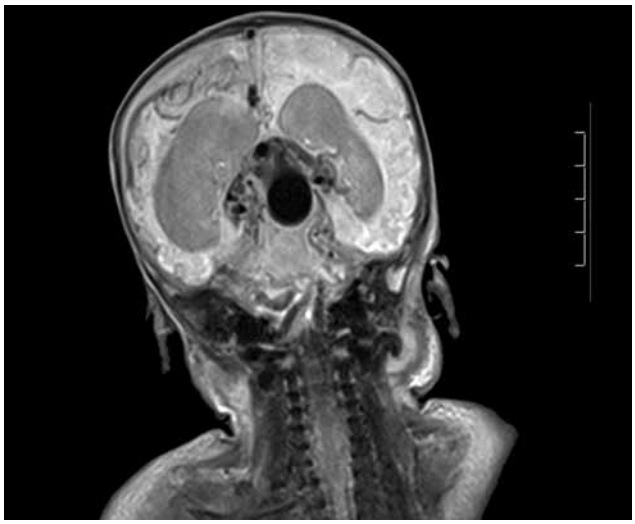

Figure 1. Magnetic resonance imaging, T_1-weighted coronal cross-section revealing dilation of both lateral ventricles and the vein of Galen aneurysmal malformation.

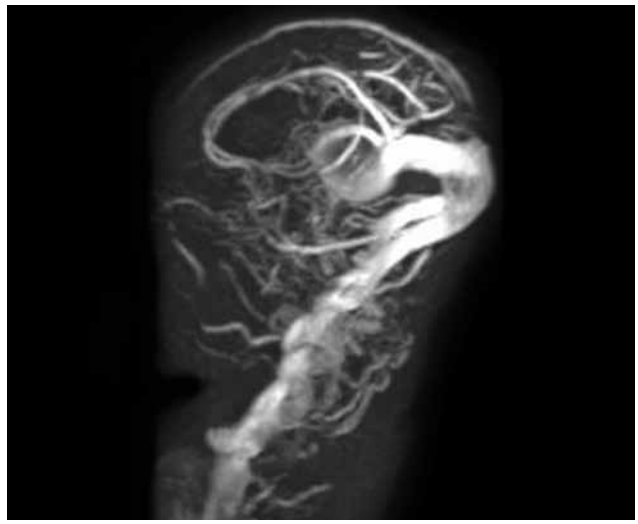

Figure 2. Magnetic resonance angiography, sagittal cross-section revealing the vein of Galen aneurysmic malformation compressing ventricles.

DISCUSSION

The VGAM is a venous ectasia secondary to an arteriovenous shunt, which drains either directly into the vein of Galen or into a tributary.[4] If it is dilated enough, prenatal diagnosis is possible by antenatal Doppler USG.

The most frequently observed clinical finding is high-flow congestive cardiac failure in neonates and infants. Other common symptoms are intracranial hemorrhage, seizures, and hydrocephaly.[5,6] Our patient had not received regular antenatal follow-up; therefore, he could not be diagnosed prenatally. He was referred to our hospital with severe respiratory distress. On admission, his physical examination revealed high-flow congestive cardiac failure, and preconvulsive movements were observed.

It is much harder to manage congestive cardiac failure due to VGAM with medical agents when compared with other causes of cardiac failure. The reason for high-flow congestive cardiac failure is the continuous volume overload to the systemic circulation. During the intrauterine period, low-resistance blood flow from the placenta prevents the filling of excessive blood into the malformation. Therefore, cardiac failure does not occur during the antenatal period. After delivery, high-flow intracranial shunts are begun, and the diastolic aortic pressure decreases rapidly. As a consequence, coronary blood flow decreases, endomyocardial ischemia arises, and the existing cardiac failure further deteriorates. Because of excessive blood accumulation at the site of the malformation, blood flow in the descending aorta is reduced. At the same time, the excessive blood returning to the right atrium causes an increase in pulmonary pressures and results in pulmonary hypertension. This hyperdynamic circulation results in high-flow congestive cardiac failure.[2,7]

The blood which was stolen from the cerebral parenchyma leads to ischemic damage. A reduction in venous drainage secondary to the poorly developed venous system may result in high cerebral venous pressure and cerebral edema. It may also cause progressive hydrocephaly due to compression of the ventricular system. If not treated efficiently and rapidly, mortality rates may rise up to 100% during early infancy, and

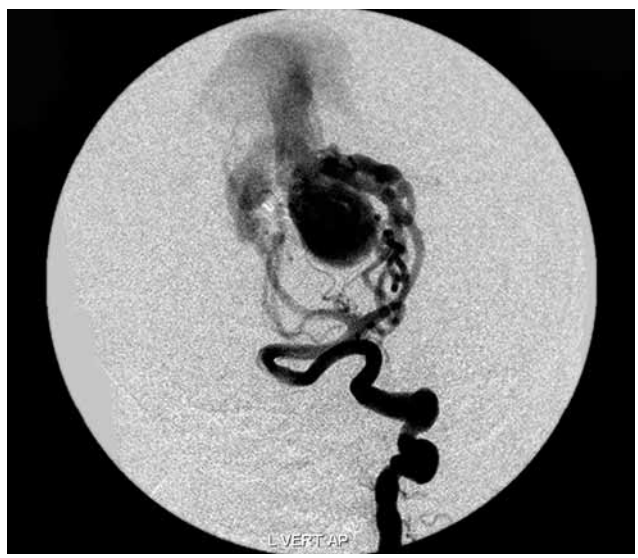

Figure 3. Postembolization magnetic resonance angiography revealing the vein of Galen still being fed by the left vertebral artery.

severe neurological sequels may develop.[2,8] Our patient had developed severe cardiac failure and respiratory distress soon after birth. However, his ECHO findings did not reveal any congenital cardiac malformation which might have led to severe suprasystemic pulmonary hypertension. The existing pathology ameliorated in his follow-up, and he remained refractory to aggressive anticongestive treatment.

Diagnosis of VGAM is made by Doppler USG in the antenatal period and by transfontanel Doppler USG, CT angiography, and MR angiography in the postnatal period. Because of his initial unresponsiveness to medical treatment, a bedside transfontanel USG was performed, and a suspicious arterial venous flow (AVF) was demonstrated. Cranial CT and MR angiography performed later confirmed the initial diagnosis of VGAM.[2,7] Our patient also had early encephalomalacic changes secondary to cerebral ischemia and remarkable ventricular dilation. Multiple organ failure and death is inevitable in a newborn baby who presents with severe congestive heart failure and who receives only anticongestive supportive medical treatment. In recent years, endovascular embolization of the feeding artery has especially been used successfully. However, the long-term neurological outcome still remains poor even after embolization. In most patients, a remarkable regression of symptoms related to cardiac failure can be observed after embolization.[9-11] In one study, the worst prognosis was observed in newborns with a mortality rate of 35.6% after endovascular intervention.[12] In another study, the general mortality and neonatal mortality rates were calculated at 10.6% and 52%, respectively.[13] Embolization was performed relatively late on the postnatal 10th day in our patient. Because the cerebral perfusion could not be improved in the early period, irreversible brain damage was demonstrated on his CT scans. On MR angiographic images, vascular malformation was demonstrated as a huge, varicose, multifeeder AVF. The amount of embolizing agent that could be administered was not sufficient enough to close all the holes, and a second angiography performed soon after embolization demonstrated that the vein of Galen was still being fed by the left vertebral artery. For this reason, congestive heart failure remained refractory to medical treatment even after embolization. Furthermore, multiorgan failure due to renal, hepatic, and mesenteric ischemia accelerated his death. Perhaps radiocontrast agents used during the embolization and angiography might have worsened the existing renal failure. Another issue is that iodine exposure during cardiac catheterization or surgery may induce transient hypothyroidism in neonates.[14] Although we lost our patient in the early neonatal period, his postangiography thyroid function tests were completely normal.

In conclusion, a diagnosis of VGAM should be considered in neonates with high-flow congestive heart failure and a structurally normal heart or in neonates with unexplained hydrocephaly and intracranial hemorrhage. Auscultation of the anterior fontanelle is a simple examination technique to be kept in mind. Once the diagnosis is made, prompt diagnostic and therapeutic interventions should be planned. Aggressive medical treatment can stabilize the symptoms of heart failure, and urgent transarterial embolization can lead to hemodynamic and neurological recovery and survival. The main goal should be recognizing this lethal malformation prenatally by providing adequate antenatal care. This is the only method that can minimize the postnatal brain injury.

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