

Intracranial Arteriovenous Malformation In An Infant: Vein Of Galen Malformation

Bir infantta intrakraniyal arteriyovenoz malformasyon galen ven malformasyonu olgu sunumu

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Summary

The vein of Galen malformation is a rare cerebrovascular disorder which is characterized by an abnormal direct communication between one or several cerebral arteries and the vein of Galen. In neonates, it usually causes congestive heart failure. Infants, older children, and adults usually present with mass effect, seizures, or intracranial hemorrhage. Here we report a 9-months-old infant diagnosed with the vein of Galen malformation. This report emphasizes that the unexplained congestive heart failure, with or without neurologic symptoms, during neonatal or early infancy period might be due to the vein of Galen malformation.

Key Words : Vein of Galen malformation, congestive heart failure, infancy

Özet

Galen ven malformasyonu bir veya birkaç arterle Galen veni arasındaki anormal bağlantı ile karakterli nadir bir serebrovasküler hastalıktır. Yenidoğanlarda genellikle konjestif kalp yetmezliğine neden olur. İnfantlar, daha büyük çocuklar ve yetişkinler sıklıkla kitle etkisi, nöbetler veya intrakraniyal kanama ile gelirler. Burada Galen ven malformasyonu tanısı almış 9 aylık bir süt çocuğu sunulmuş ve nörolojik bulgular olsun ya da olmasın, yenidoğan veya erken süt çocukluğu döneminde açıklanamayan konjestif kalp yetmezliğinin Galen ven malformasyonuna bağlı olabileceğinin vurgulanması amaçlanmıştır.

Anahtar Kelimeler: Galen ven malformasyonu konjestif kalp yetmezliği, süt çocukluğu

Introduction

The vein of Galen malformation is a rare cerebrovascular disorder which is characterized by an abnormal direct communication between one or several cerebral arteries and the vein of Galen. Respectively, posterior choroidal artery, anterior cerebral artery and transmesencephalic artery are the most common arteries that join the malformation (1). It may appear in the neonatal period or afterwards (2). Pediatric population is most sensitive to this shunt. During the neonatal or infancy period the disease is presented with congestive heart failure, intracranial hemorrhage, seizures or focal neurologic symptoms. Thereafter, progressive hydrocephaly or headache are the most frequent presentations of the vein of Galen malformation (3). Because it is a rare situation, we present here an infant who had a history of neonatal seizures and developed congestive heart failure afterwards and diagnosed with the vein of Galen malformation.

Case Report

A 9-month-old girl was brought to our hospital because of tachypnea and getting tired rapidly by breastfeeding. She was born to a 38-year old healthy mother with meconium aspiration in an outside institution. At the first day of life she had convulsions. Emergency cranial ultrasonography revealed a right frontoparietal subdural hematoma which was supposed to be related to asphyxiated delivery. She was given phenobarbital and was seizure-free thereafter. By 2 months of age she was noted to have irritability and tachypnea and be diagnosed with congestive heart failure.

In physical examination, she was 7 kg in weight (3-10th percentile), 71 cm in length (75th percentile). Her head circumference was 42cm (3-10th percentile). The vital signs were; blood pressure 85/45 mmHg, heart rate 120 beats/min, respiratory rate 44 breaths/mm and temperature 37°C. By the cardiac auscultation there had been double S2 and 1/6 systolic murmur at the left parasternal region. Liver was 2 cm palpable on the midclavicular line. A continuous cranial bruit was evident on auscultation. On the neurological examination she had

left hemiparesis The deep tendon reflexes were increased on the left side The remainder of the physical examination was unremarkable.

In spite of absence of any cardiac defect pulmonary hypertension was detected in the electrocardiography, telecardiography and echocardiography. An intracranial arteriovenous malformation was suspected. Her cranial magnetic resonance imaging revealed an appearance fitting vein of Galen malformation (Figure 1) The diagnosis was confirmed by the cerebral angiography (Figure 2) She was decided to treat via embolisation. But the parents refused the treatment and the patient was managed symptomatically.

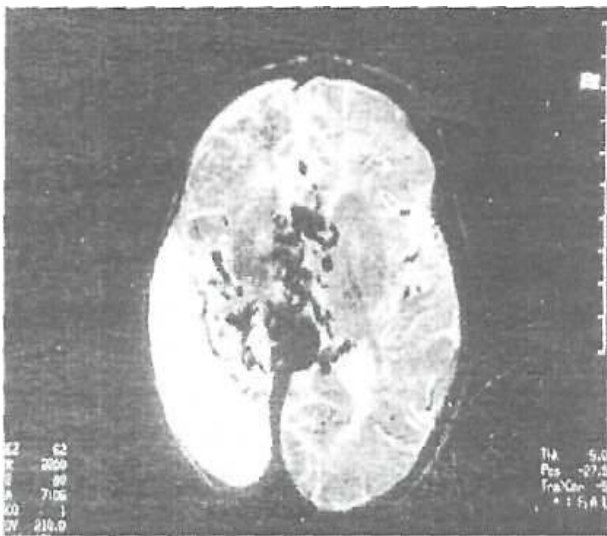


Fig 1. The cranial magnetic resonance imaging of the patient revealed an appearance fitting vein of Galen malformation and subdural effusion on the right



Fig 2 The cerebral angiography of the patient demonstrates the vein of Galen and early filling of the sinus rectus and confluens sinuum (Right internal carotid artery injection lateral view)

Discussion

Intracranial arteriovenous malformations (AVM) are complex medical emergencies in infants It is reported that 30% of AVM in childhood is vein of Galen malformation. (4) Systemic manifestations and congestive heart failure are the most common presentations encountered in the neonatal period and infancy whereas neurological signs and symptoms and hemorrhage belong mostly to the adult symptomatology (5) The flow of arterial blood into the venous side ruins cerebral perfusion and cause neurologic signs as well as congestive heart failure Breuer et al reported 5 neonates with the diagnosis of the vein of Galen malformation presented with cyanosis and symptoms of congestive heart failure without any evidence of congenital heart disease (6) Our patient presented with neonatal seizures which may be due to the vein of Galen malformation But the most possible cause of the congestive heart failure diagnosed when she was 2 months old of age is the vein of Galen malformation. The cardiac decompensation of the patient could be controlled medically at that moment. As time passed she presented with pulmonary hypertension and diagnosed with the vein of Galen malformation.

Lasjaunias et al classified vein of Galen malformation into 5 different types in their series of 36 cases 44% parenchymatous AVM, 30% choroidal AVM, 20% mural AVM, 7% vein of Galen varices and 3% dural AVM. They reported that the pediatric population was most sensitive to this shunt whatever its type (2) It was also reported that the classification of the malformation was difficult in most of cases. We considered that our case encounter a complex type of the malformation.

Because of the poor outcome of vein of Galen malformations, all authors believe that these malformations have to be treated aggressively (2,3) The aim of the therapy is to decrease blood flow through the AVM and control cardiac failure (4) In order to carry out elective embolisation and surgical occlusion convulsions and cardiac failure have to be controlled by medical therapy. (7) By the newborn cases presented with congestive heart failure the mortality rate is over 90% without embolisation, 46 4% with complete anatomic occlusion and 82% of cases healed clinically after embolisation methods (8) It has been showed that, the neurological deficits get better, psychomotor development increases and pulmonary hypertension regresses dramatically after treatment (6,9) Our patient had been decided to treat with embolisation. But the parents refused the treatment.

In conclusion, we suggest that newborns or infants who present unexplained congestive heart failure with or without neurologic symptoms, have to be considered as having vein of Galen malformation.

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