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Case Report

VEIN OF GALEN ANEURYSMAL MALFORMATION IN A NEONATE

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SUMMARY – In this article, we present a neonate with the vein of Galen aneurysmal malformation, a rare blood vessel malformation of the brain, which was clinically manifested by signs of cardiac failure. Cranial ultrasound suspected the existence of the vein of Galen aneurysmal malformation, and the diagnosis was confirmed by computed tomography and magnetic resonance imaging. Based on our case, we believe that cranial ultrasound should be included in the screening protocol for neonates with cardiac failure. In addition, we believe that endovascular embolization of the blood vessels that supply the aneurysm is the method of choice in treating the vein of Galen aneurysmal malformation. Treatment of neonates with the vein of Galen aneurysmal malformation is challenging and requires a multidisciplinary approach (neonatologists, cardiologists, neuroradiologists, and neurosurgeons).

Key words: *Neonate; Vein of Galen aneurysmal malformation; Cardiac failure*

Introduction

The vein of Galen aneurysmal malformation (VGAM) is an uncommon form of congenital vascular malformation that causes arterial blood to divert into an enlarged cerebral vein (arteriovenous (AV) fistula) that is located dorsal to the tectum. VGAM is a persistence of the embryonic prosencephalic vein of Markowski and is defined as direct AV fistulas between choroidal or quadrigeminal arteries and an overlaying single median venous sac. VGAM develops because of a teratogenic insult during 6-11 weeks of fetal life[1]. The incidence of VGAM is about one *per* three million people, and it accounts for less than 1% of cerebral AV malformations[2].

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The low systemic resistance of the fetus *in utero* may reduce flow through the fistula and minimize cardiac decompensation[3]. Ultrasound allows *in utero* diagnosis of VGAM[4]. A large percentage of VGAMs are diagnosed in the neonatal period, and the remainder during early childhood. The main clinical presentations in neonates is cardiovascular and respiratory distress. Neonatal congestive cardiac failure can occasionally be caused by VGAM[5].

Untreated VGAMs have an almost 100% morbidity and mortality[6]. Endovascular embolization is currently preferred for the treatment of VGAMs due to improved mortality rates (18%-33%) and clinical outcomes[7,8].

We present a neonate with early (in the first hours of life) signs of respiratory distress and cardiac failure due to VGAM. In our patient, interventional radiological embolization of the aneurysm feeder vessels proved to be successful.

Case Report

A male neonate weighing 3470 g was born in good condition by spontaneous vaginal delivery. Pregnancy progressed uneventfully and the mother had no noteworthy medical history. Fetal ultrasound revealed no hypoechoic lesions in the brain. However, at the age of eight hours, the neonate displayed acrocyanosis, grade 3/6 systolic murmur, and hepatomegaly. Both femoral pulse and blood pressure were normal. Other systemic examinations were normal and no dysmorphic characteristics were present. Upon retrospective clinical evaluation, distinct cranial murmur was detected.

In the first few days, the neonate became tachypneic, had a hyperdynamic precordium, and gradually developed symptoms of congestive cardiac failure. The neonate required ventilatory support, cardiac failure drug treatment (inotropics, diuretics), and other supportive therapies.

Chest x-ray showed cardiomegaly (Fig. 1). The echocardiogram performed at 10 hours of life showed pulmonary hypertension, patent foramen ovale, dilated ventricular chambers, tricuspid (+2.5/4) and mitral (+1.5/4) regurgitation, and diastolic steal phenomenon in the descending aorta. A midline cystic mass posterior to the third ventricle was detected during bedside cranial ultrasonography, which was suggestive of AV malformation of the vein of Galen.

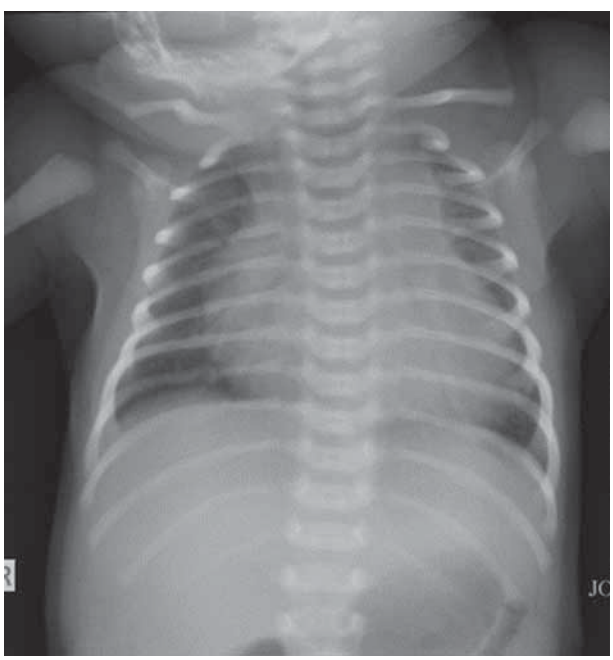

Fig. 1. Chest x-ray showing cardiomegaly.

Computed tomography (CT) of the brain revealed a hypoechoic lesion in the region posterior to third ventricle with the vessels feeding the aneurysm and mildly dilated lateral ventricles (Fig. 2). The third and fourth ventricles were of normal size. There were no signs of intracranial hemorrhage.

After stabilization, the infant was transferred to the cardiology unit where magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were performed. MRI and MRA showed choroidal type of VGAM and the vessels feeding the aneurysm. The feeding arteries originated from the left anterior cerebral arteries and left posterior cerebral arteries. On the eighth day of life, after a period of stabilization, effective transfemoral embolization of the arteries feeding the aneurysm with Onyx 18 was carried out. The postoperative course was complicated by the development of sepsis, along with deranged renal and hepatic function.

During hospital stay, hypopituitarism and secondary hypothyroidism were diagnosed, so hormone replacement therapy was started. In the further course, there was gradual clinical and laboratory improvement of systemic infection, and gradual improvement of renal, hepatic, cardiac and pulmonary function. The patient was extubated on day 13. At the time of discharge, there were no signs of congestive heart failure, neurological complications (seizures) were stopped with treatment, so it was concluded that the neuroradiological intervention produced satisfactory results.

Discussion

The VGAMs are uncommon intracranial malformations that frequently manifest dramatically in infancy. Cardiovascular and respiratory distress in neonates is the most prevalent symptom (94%) and is caused by high-flow AV shunts of VGAMs and increased heart workload, which can lead to cardiac failure. This can also lead to increased workload on the lungs, liver or kidneys, causing them to fail as well[5].

Imaging is typically used to confirm the diagnosis because VGAM is extremely rare and the symptoms are nonspecific. VGAM can be diagnosed by ultrasound before birth[4]. However, in most cases, VGAM is diagnosed postnatally by cranial ultrasonography when the neonate starts to have problems. CT of the brain, MRI and MRA are used to confirm the diagnosis[5].

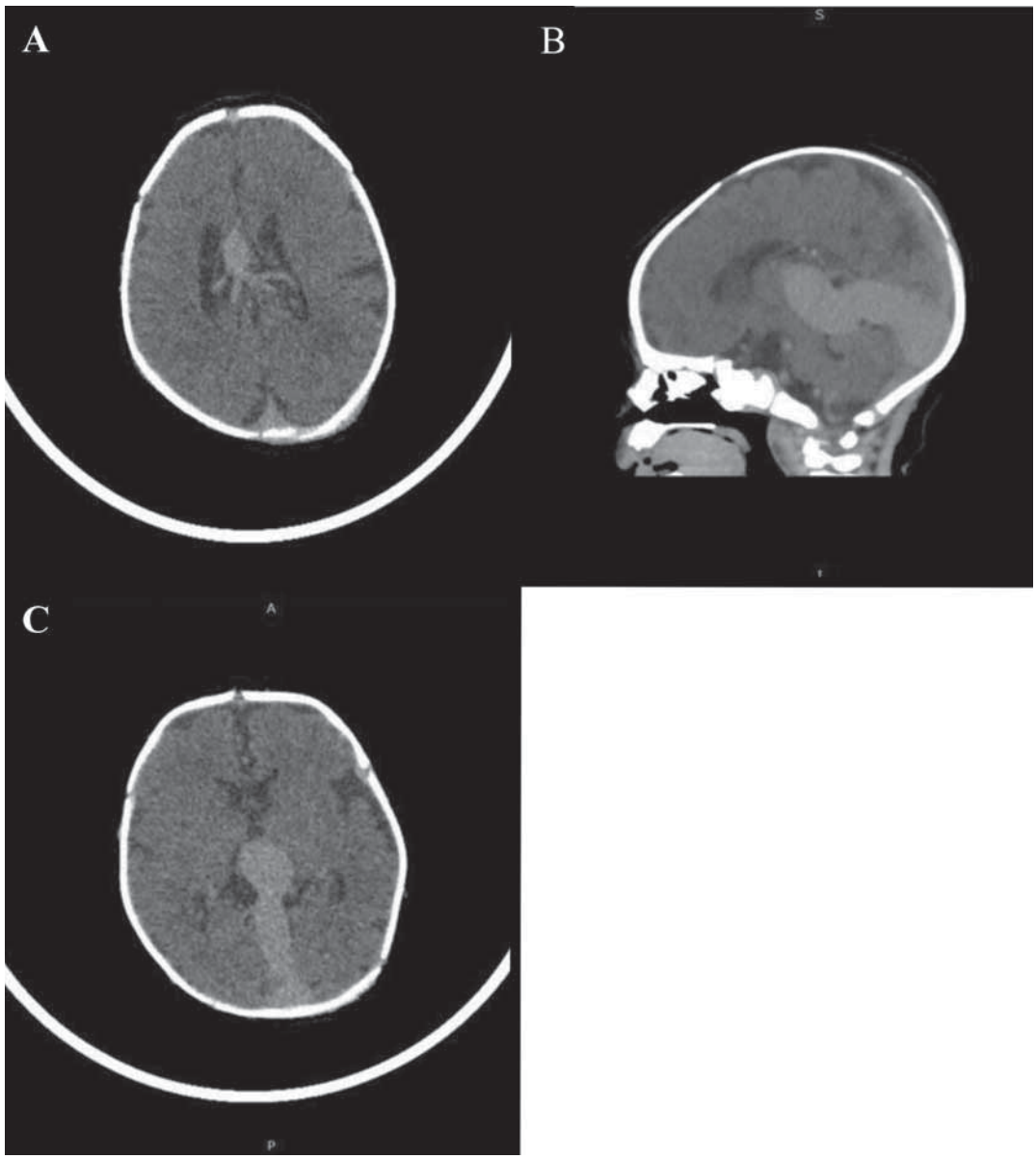

Fig. 2. Computed tomography of the brain: (A and B) mid-sagittal and coronal images showing a hypoechoic lesion in the region posterior to third ventricle; (C) the vessels feeding the aneurysm and mildly dilated lateral ventricles.

Embolization or surgical correction are two treatment options for VGAM. A review of the clinical history and research literature indicates that endovascular embolization for VGAM can produce a favorable clinical outcome, an acceptable mortality rate, and few sequels[7]. Surgery is rarely used in the current management of VGAM[5]. Untreated VGAMs have an almost 100% morbidity and mortality. According to the latest studies, the mortality rate after endovascular embolization is 18%-33%[8,9].

We report a case of the neonate with postnatally diagnosed VGAM. Shortly after birth, the newborn began to have problems (respiratory distress and cardiac failure). The x-ray showed enlargement of the cardiac silhouette, but ultrasonography revealed no abnormalities in the cardiac structure. VGAM was suspected by cranial ultrasound. CT, MRI and MRA confirmed the diagnosis, and showed aneurysm feeders arising from the anterior and posterior cerebral arteries. Embolization of the vessels feeding the aneurysm on

day 8 of life resulted in improvement of the general condition of the neonate and disappearance of cardiac symptoms. VGAMs are rare intracranial anomalies that can manifest dramatically during the neonatal period, even with features of cardiac failure. Therefore, the neonate with cardiac failure should be evaluated for a possible intracranial etiology. If VGAM is the cause of cardiac failure, early embolization of the aneurysm is necessary for survival and improvement of cardiac and respiratory symptoms. Treatment of neonates with VGAM is challenging and requires a multidisciplinary approach (neonatologists, cardiologists, neuroradiologists and neurosurgeons).

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Sažetak

ANEURIZMA VENE GALENAE U NOVOROĐENČETA

L. Banjac i *B. Banjac*

U ovom radu prikazujemo novorođenče s aneurizmom vene Galenae, rijetkom krvožilnom malformacijom mozga koja se klinički manifestirala znakovima zatajenja srca. Ultrazvučnim pregledom mozga posumnjali smo na postojanje aneurizme vene Galenae, a dijagnoza je potvrđena kompjutoriziranom tomografijom i magnetskom rezonancijom. Na temelju našeg slučaja smatramo da ultrazvuk mozga treba uključiti u protokol probira za novorođenčad sa zatajenjem srca. Također smatramo da je endovaskularna embolizacija krvnih žila koje hrane aneurizmu metoda izbora u liječenju aneurizme vene Galenae. Liječenje novorođenčadi s aneurizmom vene Galenae je složeno i zahtijeva multidisciplinarni pristup (neonatologa, kardiologa, neuroradiologa i neurokirurga).

Ključne riječi: *Novorođenče; Aneurizma vene Galenae; Zatajenje srca*