Olgu Sunumu Case Report

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Prenatally Diagnosed Vein Of Galen Aneurysm Presenting With Cardiac Failure

Kardiyak Yetmezlikle Başvuran Galen Ven Anevrizmasının Prenatal Tanısı

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ABSTRACT

Vein of Galen aneurysm is a rare congenital anomaly, originated from a defect in fusion of internal cerebral veins. It constitutes 1% of all intracranial vascular malformations. Due to low resistance, it produces high-output cardiac failure(1,2). A 39-year-old woman was diagnosed Galen vein aneurysmal malformation with the ultrasonographic finding of dilated structure in the midline of the brain and turbulent flow within the dilated vascular structure showed with color flow Doppler ultrasonography at the 26th gestational week. We report the subject diagnosed prenatally in our clinic in light of the literature.

Keywords: Vein of Galen aneurysm, prenatal diagnosis, cardiac failure

ÖZ

Galen ven anevrizması, internal serebral venlerin füzyon defektinden kaynaklanan nadir bir konjenital anomalidir. İntrakraniyal vasküler malformasyonların %1'ini oluşturur. Düşük direnç nedeniyle, yüksek debili kardiyak yetmezlik yapar(1,2). 39 yaşındaki olguya, ultrasonografide beyin orta hattında genişlemiş yapı ve renkli akım Dopler ultrasonografide bu yapı içinde türbülan akım izlenmesi ile 26. gebelik haftasında Galen ven anevrizması tanısı konuldu. Kliniğimizde prenatal tanısı konulan bu olgu literatür bilgileri ışığında sunulmaktadır.

Anahtar Kelimeler: Galen ven anevrizması, prenatal tanı, kardiyak yetmezlik

Introduction

Vein of Galen aneurysm is a rare congenital anomaly, originated from a defect in fusion of internal cerebral veins. It constitutes 1% of all intracranial vascular malformations. Vein of Galen aneurysm results from an arteriovenous connection between the primitive choroidal vessels and the median prosencephalic vein of Markowski. In the course of normal development of intracranial vasculature, Markowski's vein undergoes regression and middle cerebral veins develop. Due to low resistance, vein of Galen aneurysm produces high-output cardiac failure(1,2).

We report a case that was diagnosed prenatally in our clinic in light of the literature.

Case Report

A 39- year-old woman, gravida 4, para 1, first trimester abortions 2, was referred us for evaluation of cardiac structure due to suspicion of anomaly at 25 weeks 2 days of gestation. Ultrasonographic examination resulted with increased nuchal thickness, left deviation of cardiac axis, cardiomegaly filling the half of the thorax, significantly dilated superior vena cava and vasculature of the neck.

As we check the cranial anatomy;dilated structure in the midline of the brain was noticed and color flow Doppler ultrasonography demonstrated turbulent flow within the dilated vascular structure, diagnosed Galen vein aneurysmal malformation (Figure 1,2). In the first visit, we also detect; cardiomegaly, bilateral ventriculomegaly (ventricular atria 17mm bilaterally) and mild polyhydroamnios.

Figure 1: Cranial anatomy, dilated vascular structure in the midline

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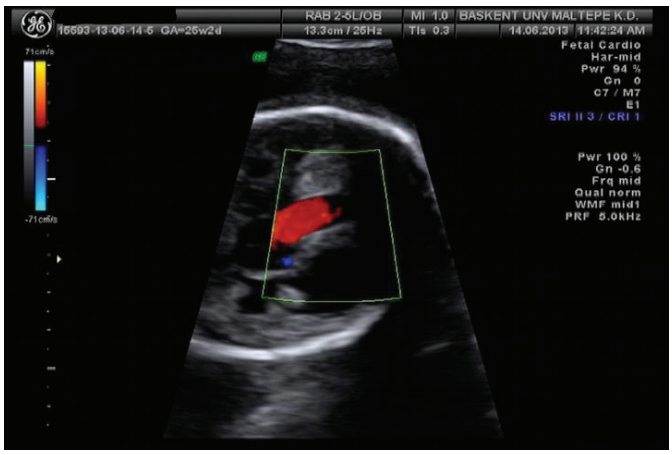
Figure 2: Color flow Doppler demonstrating dilated vascular structure



Dilated right cardiac chambers, cardiomegaly were detected without cardiac structural abnormalities with fetal echocardiography. Fetal magnetic resonance imaging(MRI), a complementary method, can not be performed but it is important to confirm the diagnosis, to detect associated brain abnormalities and to rule out differential diagnoses including arachnoid, porencephalic or choroid plexus cysts, pineal tumors, choroid papilloma and intracerebral hematoma(3,6).

Antenatal visits were continued at frequent intervals. Considering the possibility of premature birth, single cure betametasone treatment was given for the fetal lung maturation at 30th weeks of gestation. In another visit, symptoms of the cardiac insufficiency such as; edema of the scalp, pericardial effusion, cardiomegaly and severe polyhydroamnios were detected and cesarean section was performed at 32 weeks' of gestation. A female infant weighing 1750 g, with apgar scores of 6 at 1 min, 7 at 10 min, was delivered. The newborn with respiratory distress was intubated and intratracheal surfactant was applied.

Vascular and interventional radiology planned embolization but the newborn can not be stabilized there for eshe did not receive embolization.After postpartum 10 hours, following the decrease in saturation and brady cardia, cardiopulmonary arrest was occured and the newborn died.

Placental pathology report was including; heavy placenta according to gestational age, dismaturation in the chorionic villi and such findings were consistent with non-immune hydrops.

Discussion

Galen vein aneurysmal malformation is usually established in the third trimester of pregnancy, presenting with cardiomegaly. As Galen vein aneurysmal malformation is not associated with chromosomal abnormalities, fetal karyotyping was notroutinely carried out. Out come is particularly poor when associated anomalies such as signs of cardiac dysfunction (which is directly related to the size of arterovenous shunt)or cerebral defects (the extent of cerebral ischemia caused by increased venous pressure and so-called cerebral steal) were detected prenatally. Vein of Galen aneurysm can be complicated with nonimmune hydrops, as it has occured in our case. Regardless of the type of treatment, perinatal morbidity and mortality are high, often due to cardiac failure.Treatment of choice involves performing transarterial embolization in the postnatal period and its efficacy depends largely on the size of malformations and developed complications (7,10).

In the literature, some of the publications suggest birth before the fetal decompensation but, some of them say that premature birth does not alter the newborn outcomes. Severe cardiac failure, is the most important predictor of morbidity and mortality(1,11,13).

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