Prenatal Ultrasonography in the Diagnosis of Vein of Galen Aneurysmal Malformation

Galen Ven Anevrizmal Malformasyonunun Prenatal Ultrasonografik Tanısı

Prenatal Tanı Konan Galen Ven Malformasyonu / Prenatal Diagnosis of Galen Vein Aneursyma

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Bu çalışma 33. Ulusal Radyoloji Kongresi'nde poster olarak sunulmuştur

Özet

Galen veni anevrizmal malformasyonu (GVAM) Galen veni ile bir veya birkaç serebral arter arasındaki anormal doğrudan ilişki ile karakterize nadir görülen bir konjenital intrakranial arteriovenöz şanttır. Tüm intrakranial vasküler malformasyonların % 1'inden azını oluşturmaktadır. GVAM nedeni tam olarak aydınlatılamamış olsa da savunulan hipotez anevrizmanın progresif dilatasyonuna neden olan embriyonik vasküler desteğin sebat etmesidir. Prenatal ultrasonografi ile tanı konabilmektedir. Aşırı yüksek akımlı intraserebral şant, pulmoner hipertansiyonun farklı dereceleri ile beraber olan hacim artışına bağlı yüksek debili kalp yetmezliğine neden olabilir. Kalp yetmezliğine neden olabildiği için prenatal ve erken postnatal dönemde prognoz kötüdür. Güncel tedavi seçeneği endovasküler tedavi olup kalp yetmezliği gelişmeden erken dönemde yapılması hayat kurtarıcıdır. Bu nedenle multidisipliner yaklaşımla tedavi edilmesi gereken bu durumun prenatal dönemde tanı alması önemlidir. Bu çalışmamızda 28 yaşında B-mod ve renkli Doppler sonografi kullanarak prenatal dönemde GVAM tanısı konan 33 haftalık gebeliği bulunan bir olguyu sunduk.

Anahtar Kelimeler

Prenatal Tanı; Galen Veni; Anevrizma; Ultrasonografi

Abstract

The vein of Galen aneurysmal malformation (VGAM) is a rarely-seen congenital intracranial arteriovenous shunt characterized by an abnormal direct relationship between one or more cerebral arteries and the vein of Galen. While the causes of VGAM are not fully known, the hypothesis advocated is that it is due to persistence of embryonic vascular support causing progressive dilatation of the aneurysm. It can be diagnosed by prenatal ultrasonography. Together with extremely high-flow intracerebral shunt and differing degrees of pulmonary hypertension, high-output heart failure linked to volume increases may occur. Current treatment choice is endovascular treatment which if administered in the early period before heart failure develops may be life-saving. For this reason, prenatal diagnosis of this multidisciplinary treatment approach requiring situation, is important. Here, we present a 28-year-old woman at 33 weeks' gestation who was evaluated using B-mode and color Doppler sonography for the prenatal diagnosis of VGAM.

Keywords

Prenatal Diagnosis; Galen Vein; Aneurysm; Ultrasonography

DOI: 10.4328/JCAM.2244 Received: 22.12.2013 Accepted: 07.01.2014 Printed: 01.11.2013 J Clin Anal Med 2013;4(suppl 3): 255-7
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Introduction

VGAM is a rarely-seen congenital intracranial arteriovenous shunt characterized by an abnormal direct relationship between one or more cerebral arteries and the vein of Galen. It forms less than 1% of all arteriovenous malformations [1,2].

VGAM is a vascular anomaly that presents with congestive heart failure symptoms generally in the prenatal and neonatal periods. In the infantile period, hydrocephalus, seizures, encephalomalasia and subcortical calcification may be observed linked to its obstructive effects [3].

In this report a vein of Galen aneurysm case accompanied by secondary findings is presented with color Doppler ultrasonography (US) and B-mode US.

Case Report

The 28-year old pregnant patient, with no history of intermarriage, was identified to be carrying a female fetus with biometric readings appropriate for 33 weeks. On obstetric US B-mode examination of the fetal cranium showed an anechoic cystic lesion on the midline localized posterior-superior to the 3rd ventricle, extending towards the occipital region (Figure 1). Color Doppler US indicated venous flow pattern with high flow speed and many feeder arteries around the lesion (Figure 2). Vein of Galen aneurysm diagnosis was made with the findings of B-mode and color Doppler US.

In addition parenchymal disorganization and vascular malformation in the cerebral and cerebellar hemispheres, dilatation of the cerebral lateral ventricles, tortuosity-dilatation in the vena cava superior and jugular vein in the neck (Figure 3-4), and clear cardiomegaly and minimal pericardial fluid linked to hyperdynamic flow in the atrium (Figure 5) were observed.

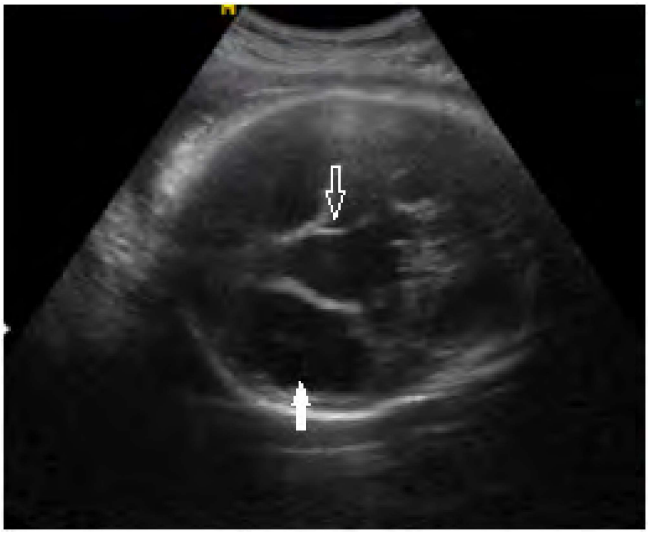


Figure 1. Gray scale axial image of the fetal head shows supratentorial-located, smooth-edged, anechoic cystic lesion (empty arrow) and hydrocephalus (filled arrow).

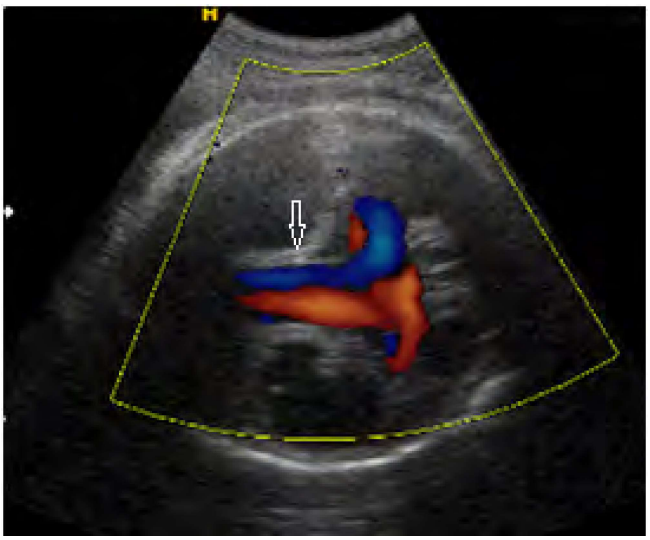
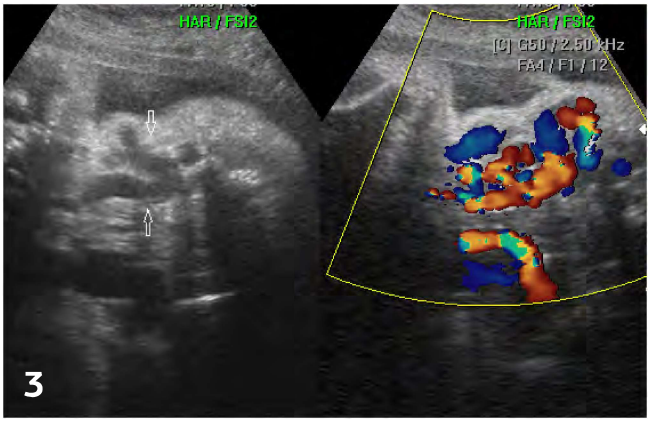
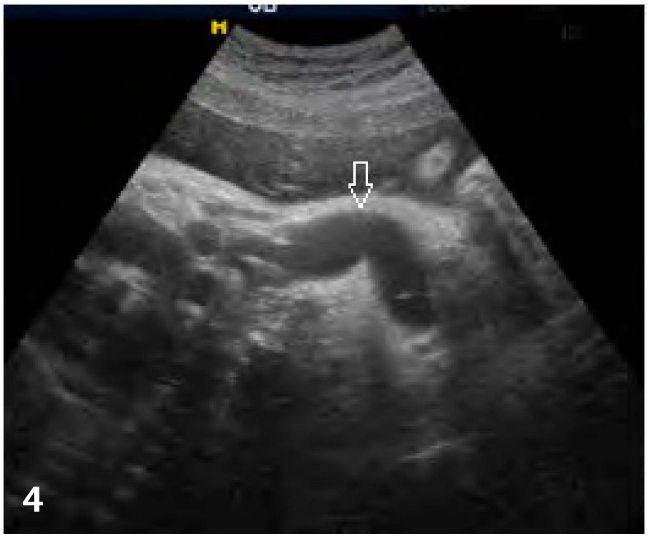


Figure 2. Doppler US axial image of cerebrum shows midline cystic lesion with color fill and AVM type turbulent flow pattern (arrow).





Figures 3-4. Coronal US images including neck and upper thoracic region show tortuosity and dilatation of vena cava superior and bilateral jugular veins (arrow).



Figure 5. Axial US image at thoracic level shows cardiomegaly and minimal pericardial fluid.

In spite of prenatal diagnosis of this case, before endovascular treatment could be administered, the patient died due to development of heart failure in the early postnatal period.

Discussion

The vein of Galen is located under the cerebral hemispheres and drains the front and mid parts of the brain into the posterior cerebral fossa sinuses. VGAM forms when the vein of Galen dilates as a result of an arteriovenous fistula between the choroid branches of the cerebral artery and/or basilar arterial branches and dilated veins of the mesencephalon [4].

The most important characteristic of VGAM is the aneurysmal dilatation of the venous structure called the vein of Galen. The most frequent feeder arteries of this dilated venous structure are, in order of frequency, posterior choroidal artery, anterior cerebral artery, middle cerebral artery, anterior choroidal artery and posterior cerebral artery [5].

While the cause of VGAM is not fully known, the currently accepted hypothesis is persistence of embryonic vascular supply which normally regresses, known as the choroidal arteries and the anterior segment of the median prosencephalic vein of Markowski. This persistence and gradual dilation between aneurysmal components forms the typical VGAM.

It is observed with equal rates in both genders [6]. In our case the fetus was female.

It most frequently presents as high-output heart failure and its secondary findings in the prenatal and newborn period [7]. As a result of the "steal" phenomenon of severe high-output heart failure there is a clear increase in cardiac preload due to the venous return from the brain. Hydocephalus may develop secondarily due to obstruction of the aquaductus Sylvius by the dilated aneurysm. The "steal" phenomenon may cause more cerebral infarctus due to self-routed blood flow from the parenchyma of the aneurysm and periventricular white matter lesions [8].

Currently obstetric ultrasonography is widely used. When evaluating the cerebral structures if an intracranial cystic structure is observed VGAM should be suspected. Diagnosis can be made by high-flow observed within the cystic structure on color Doppler investigation. Due to the compressional effect of the aneurysm in the advanced weeks of pregnancy slight or moderate hydrocephalus may develop. In the prenatal or postnatal period high-output congestive heart failure and serious hemodynamic dysfunction may cause death in the intrauterine or postnatal periods. It may cause cardiomegaly, ventriculomegaly, intrauterine growth retardation, dilatation of the jugular vein, hepatomegaly, polyhydramnios and non-immune hydrops fetalis. The main factor determining prognosis is the effect of high flow from this AV fistula on the fetus or newborn [9]. In our case on prenatal ultrasound cardiomegaly and minimal pericardial effusion linked to heart failure, and prominent jugular veins were present.

Currently the alternative treatment for VGAM is to embolize the abnormal feeder arteries before development of heart failure, which increases the importance of prenatal diagnosis.

At the moment endovascular treatment is the first step treatment for VGAM. The previous 100% mortality rate has reduced with endovascular treatment approaches. Embolization of both feeder arteries and veins reduces the speed of blood flow increasing the survival rates. Due to neuro-embolization the extreme blood flow in the vein of Galen is reduced, correcting cardiac function and preventing the formation of brain damage [10].

As endovascular treatment reduces mortality if done before heart failure develops, to increase the survival chances prenatal diagnosis gains even more importance. To administer the necessary treatment in the early postnatal period, the birth must occur where appropriate conditions can be provided. In addition while diagnosis can be easily made by prenatal US, in the postnatal period differential diagnosis of VGAM is only considered after heart failure findings have developed and diagnosis may be late.

In our case although prenatal diagnosis was made, due to development of heart failure the patient died in the early postnatal period.

In conclusion, vein of Galen aneurysm is a rare cause of prenatal heart failure and hydrops which may be diagnosed prenatally. Early diagnosis and treatment with endovascular intervention in conditions with a multidisciplinary approach may reduce the rate of mortality and morbidity.

Competing interests

The authors declare that they have no competing interests.

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How to cite this article:
Uysal F, Kurtulmuş S, Uysal A. Prenatal Ultrasonography in the Diagnosis of Vein of Galen Aneurysmal Malformation. J Clin Anal Med 2013;4(suppl 3): 255-7.