

Case Report

Prenatal diagnosis of vein of Galen aneurysm: A case report

Ayşe Kirbas^{1,*}, Ebru Biberoglu¹, Halil Korkut Daglar¹, Nida Erol¹, Suat Fitoz², Dilek Uygur¹, Nuri Danisman¹

¹ Department of Perinatology, Zekai Tahir Burak Women's Health, Education and Research Hospital, Ankara, Turkey

² Department of Radiology, Ankara University Faculty of Medicine, Ankara, Turkey

Abstract

Aneurysm of the vein of Galen (AVG), a complex malformation that has a generally poor prognosis, with a very high risk of neurologic sequelae and a mortality rate of 50%, represents less than 1% of cerebral arteriovenous malformations. The most noticeable prenatal finding is the existence of a posterior, midline, or slightly lateral supratentorial cystic structure in the fetal brain. Visualization of the blood flow in the cystic structure facilitates the diagnosis of AVG; prenatal magnetic resonance imaging is useful for evaluating the vascular connection and the degree of cerebral damage. In addition, prenatal findings indicative of heart failure are associated with a poor prognosis for the newborn. Therefore, early detection is important in order to proceed to effective therapeutic management. Premature delivery does not improve the perinatal outcome. The prenatal diagnosis of AVG has been reported in only a few occasions. In this report, we present a prenatal diagnosis of AVG with cardiomegaly in a fetus at the 31 weeks' gestation.

Key words:

Aneurysm of the vein of Galen, ultrasonography, prenatal diagnosis

Introduction

Aneurysm of the vein of Galen (AVG) is a complex arteriovenous malformation that affects multiple communications between the vein of Galen and the cerebral arteries. It is seen very rarely, and it can cause severe morbidity and mortality in neonates [1]. Here is the presentation of a prenatal diagnosis of AVG with cardiomegaly in a fetus at the 31 weeks' gestation.

Case presentation

A 33-year-old primigravida was referred to our unit with a cystic mass in the median line of the fetal brain, evidenced in a two-dimensional (2D) ultrasonography scan conducted in the

31st week of gestation. There was a midline tubular anechoic structure situated above the thalamus (Figure 1). Color Doppler imaging showed turbulent flow and blood draining into an enlarged straight sinus, suggesting an AVG (Figure 2). The ultrasonography did not indicate any further structural brain abnormalities. The heart was considerably enlarged, indicating intrauterine fetal cardiac insufficiency. The parents were counseled by a multidisciplinary team, and, considering the size of the cerebral arteriovenous shunt, they were informed about the risks of serious mental and physical handicaps, as well as imminent cardiac failure. Fetal magnetic resonance imaging (MRI) performed at 33 weeks' gestation to confirm the diagnosis showed a grossly dilated vein of Galen with prominent choroid arteries as supplying vessels and significant dilatation of the straight sinus, the confluence of sinuses, and the transverse sinuses. The brain parenchyma was normal, without hydrocephalus or structural brain damage. An elective caesarean section was performed at 37 weeks' gestation after lung maturity, and a male neonate weighing 2970 g was born. The diagnosis of AVG was confirmed by post-natal ultrasonography. Unfortunately,

Article history:

Received 09 10 2014

Accepted 15 12 2014

* **Correspondence:** Ayşe Kirbas MD.

Cankaya Mahallesi, Ziaurrahman Caddesi, 4A/2 Postal code:06680. Cankaya, Ankara, Turkey

Email: drayse1982@yahoo.com

Phone: +90 533-646-9213

the newborn died at 48 hours of age due to cardiac decompensation. The parents did not permit an autopsy.

Figure 1.



Midline cystic lesion suggests a vein of Galen aneurysmal malformation at 31 weeks of gestation

Discussion

In this report, we described a very rare case of fetal AVG complicated with cardiac insufficiency. We diagnosed the condition by using 2D and color doppler ultrasonography modalities and confirmed the diagnosis with a fetal MRI. While AVG is a rare congenital anomaly of the cerebral vasculature, it is the most common cerebral arteriovenous malformation detected prenatally [2]. Prenatal diagnoses of AVG are usually made during the third trimester, and ultrasonography is usually sufficient for making a diagnosis. The classic finding is an echolucent structure in the posterior region of the midline superior to the tentorium cerebelli and superior to the thalamus [1, 3, 4]. It is important to perform a fetal MRI to confirm AVG, in order to detect associated brain abnormalities and to rule out differential diagnoses. MRI allows a better evaluation of the lesion and helps in the decision-making process [5]. Considering the useful information obtainable with fetal MRI regarding viability of the brain and degree of cardiac compromise, it should be considered a reasonable diagnostic tool when a diagnosis of AVG is based on prenatal ultrasonography [4, 6]. AVG is not associated with chromosomal abnormalities, but it can demonstrate signs of volume overload, such as cardiomegaly and hydrops. Besides, the neonatal progno-

sis is usually poor, with high incidence rates of morbidity and mortality. The differential diagnosis includes arachnoid, porencephalic, and choroid plexus cysts; pineal tumors; intracerebral hematomas; and choroid papillomas [4]. In most cases, the lesion does not resolve spontaneously, and definitive postnatal treatment is required [7]. Management is a major therapeutic problem, despite recent advances in radiologic diagnosis, neonatal intensive care, and neurosurgery. Pregnant patients diagnosed with prenatal AVG should be referred to a tertiary neonatal intensive care unit, where a pediatric neurologist, a cardiologist, an interventional neuroradiologist, and a neurosurgeon are available for management of the infant. Careful obstetric management and early postnatal intervention might lead to a favorable outcome [7, 8].

Figure 2.



Doppler color flow image demonstrates presence of prominent convoluted vessels and a turbulent flow in the lesion.

Most infants with AVG need urgent treatment. The main goal in treating an infant with high-output cardiac failure is to decrease the flow in the vein of Galen malformation, and thus, reduce cardiac output failure, improve coronary perfusion, and increase blood flow to the renal arteries. Endovascular interventional treatment has been accepted as the first treatment option for AVG patients [9, 10]. In summary, AVG can cause severe morbidity and mortality in neonates. Optimal management of infants with AVG is achieved by only through the multidisciplinary approach offered by specialized tertiary care centers.

Conflict of Interest

Authors declare no conflict of interest

References

1. Paternoster DM, Manganelli F, Moroder W, Nicolini U. Prenatal diagnosis of vein of Galen aneurysmal malformations. *Fetal Diagn Ther.* 2003;18:408-11.
2. Sepulveda W, Vanderheyden T, Pather J, Pasquini L. Vein of Galen malformation: prenatal evaluation with three-dimensional power Doppler angiography. *J Ultrasound Med* 2003;22:1395-1398.
3. Pearl M, Gregg L, Gandhi D. Cerebral venous development in relation to developmental venous anomalies and Vein of Galen aneurysmal malformations. *Semin Ultrasound CT MR.* 2011;32:252-63.
4. Doren M, Tercanli S, Holzgreve W: Prenatal sonographic diagnosis of a vein of Galen aneurysm: Relevance of associated malformations for timing and mode of delivery. *Ultrasound Obstet Gynecol* 1995; 6:287-289.
5. Maheshwari PR, Pungavkar SA, Narkhede P, Patkar DP. Vein of Galen aneurysmal malformation: antenatal MRI. *J Postgrad Med.* 2003;49:350-1.
6. Messori A, Polonara G, Salvolini U. Prenatal diagnosis of a vein of Galen aneurysmal malformation with fetal MR imaging study. *AJNR Am J Neuroradiol.* 2003;24:1923-5; author reply 1925.
7. Lasjaunias PL, Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R. The management of vein of Galen aneurysmal malformations. *Neurosurgery.* 2006; 59(5 Suppl 3): S184-94.
8. Chisholm CA, Kuller JA, Katz VL, McCoy MC. Aneurysm of the vein of Galen: prenatal diagnosis and perinatal management. *Am J Perinatol.* 1996;13:503-6.
9. Gupta AK, Rao VR, Varma DR, Kapilamoorthy TR, Kesavadas C, Krishnamoorthy T, Thomas B, Bodhey NK, Purkayastha S. Evaluation, management, and long-term follow up of vein of Galen malformations. *J Neurosurg.* 2006;105:26-33.
10. Lasjaunias PL, Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R. The management of vein of Galen aneurysmal malformations. *Neurosurgery.* 2006;59(5 Suppl 3):S184-94.