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A Case of Vein of Galen Aneurysmal Malformation Diagnosed Prenatally in a Monochorionic-Diamniotic Twin Pregnancy

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Abstract

We present a case of a vein of Galen aneurysmal malformation (VGAM), a rare congenital arteriovenous malformation, in one fetus of a monochorionic-diamniotic twin pregnancy. The diagnosis was made with color Doppler ultrasonography at 28 weeks and the affected fetus was found to have worsening cardiomegaly on subsequent fetal echocardiograms. She was emergently delivered at 32 weeks for abnormal fetal heart rate tracing of the affected twin. Magnetic resonance imaging of the brain findings after delivery demonstrated severe neurological injury; therefore, postnatal embolization was not performed. The neonate died on day of life 9. The cotwin survived without neurological complications. This is the first case in the literature of a VGAM diagnosed prenatally in a monochorionic-diamniotic twin pregnancy and demonstrates the challenge of delivery timing with prenatal diagnosis in a twin pregnancy.

Keywords
- twin anomaly
- twin pregnancy
- vein of Galen aneurysmal malformation

Case Report

A 32-year-old woman with a monochorionic-diamniotic twin gestation was referred at 23 weeks for suspected fetal cardiac anomalies for twin A. Upon transfer, the fetal echocardiogram in twin A showed a reversal of flow in the ductus arteriosus and a dilated superior vena cava and innominate vein. On follow-up fetal echocardiogram at 28 weeks, twin A was found to have borderline cardiomegaly, reversal of flow at the aortic isthmus, and findings concerning increased vascularity in the fetal brain. She was urgently referred to the Maternal-Fetal Medicine ultrasound unit where a cystic midline structure with turbulent blood flow was identified measuring 2.4 × 2.0 cm, as well as dilation of the straight sinus (8.7 mm at thinnest point and 12 mm thickest point), consistent with vein of Galen aneurysmal malformation (VGAM) (►**Fig. 1**). A 29 weeks ultrasound showed new mild polyhydramnios for both twins (deepest vertical pocket 9.1 cm/9.3 cm) and antenatal corticosteroids were administered. She underwent twice weekly biophysical profiles with reassuring results and the polyhydramnios remained mild for both twins. However, subsequent fetal echocardiograms at 30 and 32 weeks showed worsening cardiomegaly with dilation of the right atrium and ventricle in twin A. Fetal magnetic resonance imaging (MRI) at 30 weeks confirmed VGAM and showed otherwise normal cerebral parenchyma (►**Fig. 2**). While limited due to the large VGAM, ultrasound at 32 weeks suggested possible interval disruption in the brain

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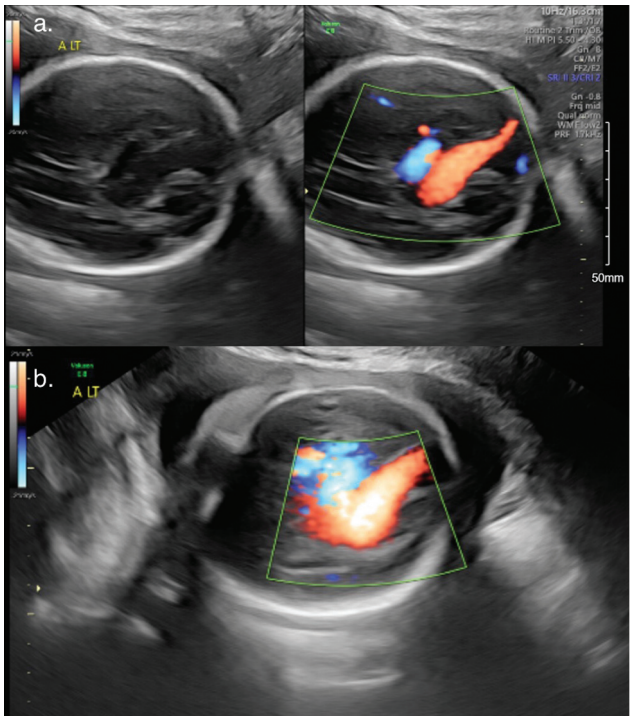

Fig. 1 Color Doppler transabdominal ultrasonography at 28 weeks showing an axial view of twin A's head with (**a**) vein of Galen aneurysmal malformation with comet tail sign and (**b**) dilated straight sinus.

parenchyma. A multidisciplinary meeting was held and given possible signs of progression of the lesion by ultrasound, to optimize outcomes for the unaffected twin, a plan was made for a 34-week delivery to balance the risk of worsening status and intrauterine demise of the affected twin with the risks of prematurity for the unaffected twin.

At $32^{6/7}$ weeks, a nonstress test for twin A was nonreactive with recurrent variable decelerations, and emergent repeat cesarean delivery was performed. Twin A was intubated shortly after birth for respiratory failure, started on dopamine for hypotension, and found to be in high output cardiac failure.

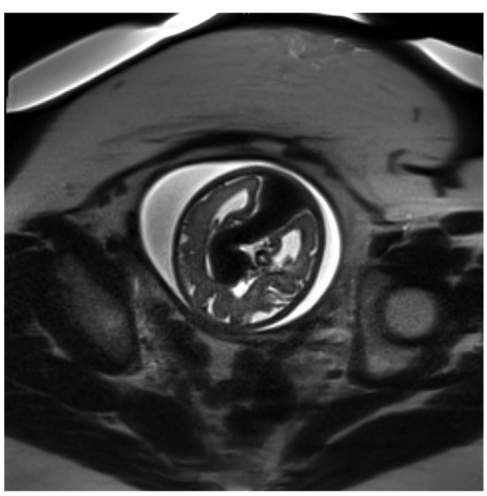

Fig. 2 Magnetic resonance imaging at 30 weeks showing vein of Galen aneurysmal malformation with normal surrounding brain parenchyma.

Postnatal brain MRI showed cerebral atrophy and chronic global ischemia suggesting poor neurological prognosis; therefore, postnatal embolization was not offered. On day of life 5, the parents decided on a do-not-resuscitate order, and twin A died on day of life 9. Twin B had a routine neonatal intensive care unit stay for prematurity and was discharged in stable condition. Placental dye studies were performed showing dilated and poorly arborized vessels on twin A's portion of the placenta (►**Fig. 3**).

Discussion

VGAM is a rare congenital cerebral arteriovenous malformation (AVM) between the choroidal vessels and the median prosencephalic vein of Markowski, which prevents its involution into the vein of Galen.[1] If diagnosed prenatally, the diagnosis is typically made in the third trimester with color Doppler ultrasonography and is commonly associated with hydrocephalus, ischemic brain injury, and cardiomegaly with high output cardiac failure as a result of shunting of blood to the AVM.[2,3] While endovascular embolization is a potential cure for VGAM, it is ideally performed at 4 to 6 months of age if the patient remains stable, but many neonates are not candidates for this intervention due to the complexity of the lesion, the extent of brain injury at birth, or the severity of heart failure. Therefore, the mortality rate is up to 32% in singletons.[4]

There are few data on the morbidity and mortality of VGAM in twin pregnancies. Of the four published cases of VGAM in twin pregnancies to date, only one affected twin survived.[5–8] This twin was part of a monochorionic-diamniotic pair delivered at 25 weeks for nonreassuring fetal status with *postnatal* diagnosis of VGAM.[7] The two cases with *prenatal* diagnosis of VGAM were dichorionic twin pregnancies (one with preterm birth at 35 weeks due to preterm labor and the other with planned 37 weeks delivery); both of the affected fetuses died in the neonatal period in the setting of heart failure and inability to successfully embolize the AVM.[5,8]

Determining the optimal timing of delivery for pregnancies diagnosed with VGAM is challenging and no current guidelines exist; therefore, management must include a multidisciplinary approach. The risk of in utero progression of the AVM (and its associated impact on the developing brain and heart) must be weighed against risks of prematurity to increase survival and optimize the neonate for endovascular intervention. Ultrasound features associated with poor prognosis include tricuspid regurgitation and AVM volume $> 20{,}000\ mm^3$, which can help guide prenatal counseling and management.[4]

The challenge of delivery timing is further complicated in twin pregnancies, especially monochorionic twins, given the demise of the affected twin imparts a risk of neurological injury and the demise of the healthy cotwin.[9] In our case, to optimize outcomes for the healthy twin per the patient's wishes, prevention of an intrauterine fetal demise of the affected twin was critical due to the shared placental circulation. This ultimately led to a preterm delivery at 32 weeks. However, the heart failure had already progressed and neurological injury was already severe (despite normal brain parenchyma on prenatal MRI).

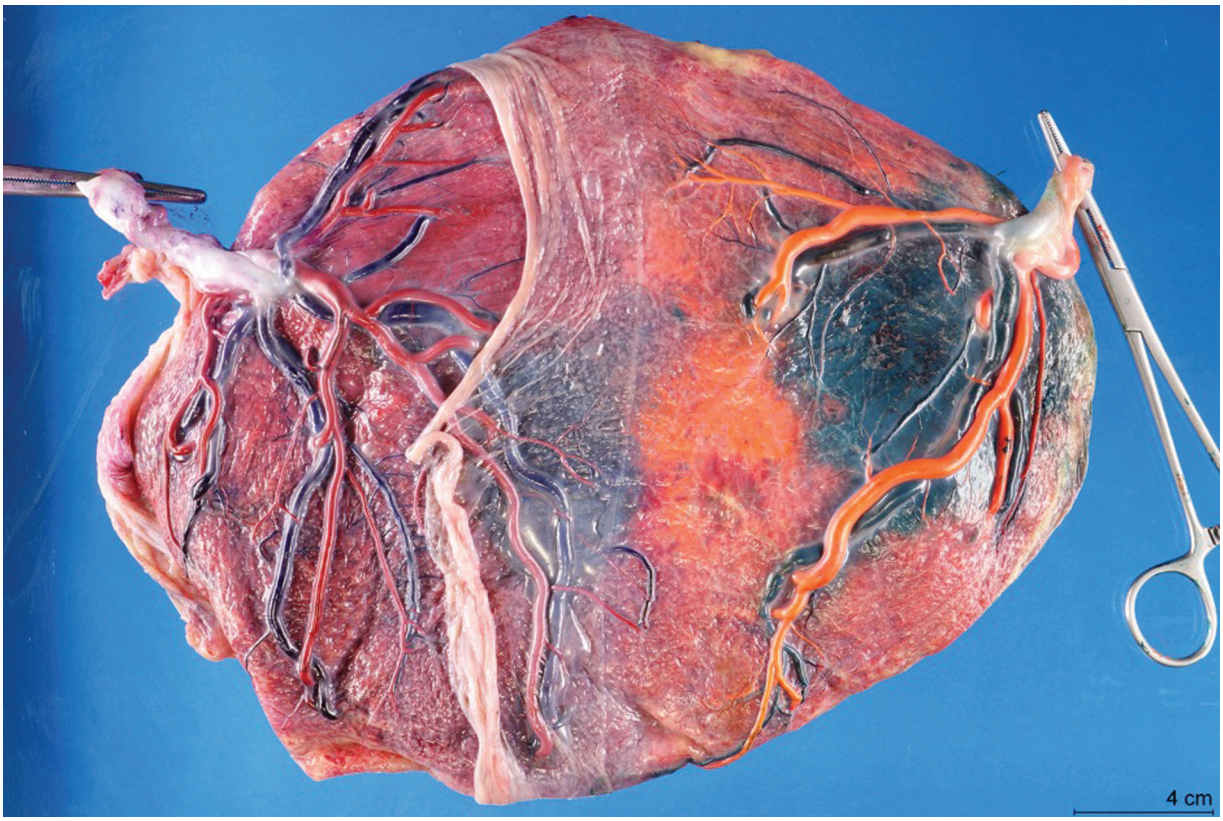

Fig. 3 Placental dye studies with twin A right (vein green, artery orange) and twin B left (vein blue, artery red).

The timing of delivery of fetuses with VGAM is often dictated by other complications, especially among twin gestations. In the other mentioned published cases of VGAM in twin pregnancies, preterm birth occurred in three out of four cases; two for fetal distress (one in the setting of twin-to-twin transfusion syndrome) and one as a result of preterm labor.[5–7] The one affected twin in the literature who survived was born at 25 weeks (for fetal distress) but only had mild heart failure therefore remained stable through two embolization procedures at 6 and 8 months of life.[7]

Given the rare clinical scenario of VGAM in twins, it remains unclear how to counsel and manage patients after prenatal diagnosis is made, and guidelines around delivery timing are limited. It remains to be determined if early delivery is associated with less severe heart failure or less extensive neurological injury, and if so, if these benefits outweigh the impact of prematurity on prognosis. This case contributes to our greater understanding of this rare diagnosis for the management of both singletons and twins.

Conflict of Interest

None declared.

References

1 Raybaud CA, Strother CM, Hald JK. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology 1989;31(02): 109–128
2 Sepulveda W, Platt CC, Fisk NM. Prenatal diagnosis of cerebral arteriovenous malformation using color Doppler ultrasonography: case report and review of the literature. Ultrasound Obstet Gynecol 1995;6(04):282–286
3 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(12): 1395–1398
4 Paladini D, Deloison B, Rossi A, et al. Vein of Galen aneurysmal malformation (VGAM) in the fetus: retrospective analysis of perinatal prognostic indicators in a two-center series of 49 cases. Ultrasound Obstet Gynecol 2017;50(02):192–199
5 Liriano B, Levy R. Prenatal sonographic diagnosis of a vein of Galen aneurysmal malformation in a twin. J Diagn Med Sonogr 2003;19 (04):242–247
6 Steggerda S, Lopriore E, Sueters M, Bartelings M, Vandenbussche F, Walther F. Twin-to-twin transfusion syndrome, vein of Galen malformation, and transposition of the great arteries in a pair of monochorionic twins: coincidence or related association? Pediatr Dev Pathol 2006;9(01):52–55
7 Komiyama M, Miyatake S, Terada A, Ishiguro T, Ichiba H, Matsumoto N. Vein of Galen aneurysmal malformation in monozygotic twin. World Neurosurg 2016;91:672.e11–672.e15
8 Hackett J, Hall L, Sadiq M, et al. Vein of Galen aneurysmal malformation affecting cotwin in dichorionic diamniotic pregnancy. Ultrasound Obstet Gynecol 2020;56(03):474–475
9 Nicolini U, Poblete A. Single intrauterine death in monochorionic twin pregnancies. Ultrasound Obstet Gynecol 1999;14(05): 297–301