Vein of Galen Malformation

Prenatal Evaluation With Three-dimensional Power Doppler Angiography

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Arteriovenous malformation of the vein of Galen, also known as vein of Galen aneurysm, is a distinct intracranial anomaly characterized by a midline, deeply located, high-flow lesion with an extremely complex vascular architecture. Although they represent fewer than 1% of all cerebral arteriovenous malformations seen in children and adults, almost all cases diagnosed in the fetus and neonate involve the vein of Galen.1,2 During the perinatal period, a timely diagnosis is of paramount importance because the large systemic shunting within the fetal brain often results in a substantial steal of blood, potentially leading to cardiac failure, hydrops, and perinatal death.1–4

Prenatal diagnosis of vein of Galen malformation has been documented by several authors.2 In this report, we present an additional case that was evaluated further with the recently developed three-dimensional (3D) power Doppler angiographic imaging modality. This new technique provided us with improved images of the malformation in utero, which were helpful for characterizing the vascular anatomic features of the lesion before planned delivery and neonatal treatment.

Case Report

A 31-year-old woman, gravida 3, para 1, was referred for sonographic evaluation at 34 weeks because of suspected vein of Galen malformation. Her history was notable for a previous stillborn fetus delivered by cesarean section at 36 weeks for abruptio placentae. Although the current pregnancy was uncomplicated, regular scans were performed to monitor fetal growth and fetoplacental perfusion. At 34 weeks, a midline, supratentorial anechoic area measuring $29 \times 21 \times 19$ mm was found in the fetal brain, which was confirmed by color Doppler sonography as vascular in nature. In addition, the lesion was noted to extend posteriorly through the dilated superior sagittal sinus (Fig. 1); the fetal neck vessels were mildly enlarged; and there was mild cardiomegaly but no other signs of cardiac insufficiency.

Three-dimensional power Doppler angiography (Voluson 730 Expert; GE Medical Systems, Livingston, Scotland) was also used to characterize the...
vascular pattern of the lesion. Multiple feeding vessels were clearly identified draining directly into the dilated vein of Galen, which were not depicted on conventional color flow imaging. Superimposed gray scale sonography (glass body mode) also helped in determining the precise location of the malformation and the feeding vessels (Fig. 2). Subsequent fetal magnetic resonance imaging (MRI) confirmed the diagnosis of vein of Galen malformation in association with prominent vessels coursing around the corpus callosum both inferiorly and anteriorly but no other cerebral structural anomalies (Fig. 3).

The parents were referred for full pediatric specialist consultation involving a neuroradiologist, a neurologist, and an intensivist to optimize perinatal management. Serial follow-up scans were performed weekly to monitor the cardiovascular status of the fetus, which remained stable despite mild cardiomegaly and high flow through the dilated vein of Galen. At 37.5 weeks, a male neonate was born by elective cesarean delivery weighing 3358 g with Apgar scores of 9 and 9 at 1 and 5 min, respectively. The diagnosis of vein of Galen malformation was confirmed by postnatal transfontanelle sonography. The neonate was transferred for neurosurgical evaluation on the same day and, after assessment, was discharged in good condition. At the age of 3 months, he was doing well and awaiting embolization of the lesion.

Discussion

In this report, we describe the prenatal sonographic findings in a case of vein of Galen malformation, including those obtained with 3D power Doppler angiography. This technique allows spatial angle-independent visualization of blood vessels, which may assist in the prenatal evaluation of several malformations involving the fetal vascular system. Vein of Galen malformation is a complex vascular anomaly. It is thought to be the result of increased blood flow through arteriovenous shunting into the vein of Galen in association with venous ectasia or obstruction of a dural sinus distal to the aneurysm. Before the advent of sonography, neonates with this condition usually had congestive heart failure of “unknown” etiology in the first week of life, and in many, the correct diagnosis was made only at the time of untreatable cardiac failure or autopsy. With the widespread use
of sonography in antenatal care, most cases of vein of Galen malformation are currently detected in utero. The most striking prenatal feature is the detection of a cerebral midline tubular anechoic structure superior to the thalamus, which is contiguous with the dilated sagittal sinus (the “comet tail” or “keyhole” sign). In the last 15 years, prenatal diagnosis of this vascular anomaly has been facilitated greatly by the use of color Doppler sonography, which is crucial for differentiating this lesion from other cystic lesions of the fetal brain because the vein of Galen malformation is the only lesion that clearly displays blood flow within it. According to a review based on 25 prenatally diagnosed cases, associated prenatal sonographic features were present in 76% of the fetuses, including cardiomegaly in 64%, enlarged neck vessels in 32%, and ventriculomegaly in 24%.

In severe cases, hydrops secondary to high-output cardiac failure already may be present in the fetus at the time of diagnosis, but fetuses with mild degrees of cardiac insufficiency are prone to have decompensation in the early neonatal period. This could be mediated by the unique features of the fetal and neonatal circulations. In the fetus, the placenta is the organ with the lowest resistance to blood flow. When the cord is clamped after delivery, there are abrupt circulatory changes characterized by redistribution of blood flow, resulting in low resistance in the fetal lungs and brain, which increases the blood flow through the vein of Galen malformation considerably. This particular hemodynamic phenomenon could explain the rapid deterioration in neonates with borderline cardiac insufficiency or could lead to neonatal death in those already having cardiac failure. Therefore, accurate determination of cardiovascular status should be made routinely before and after delivery in all cases.

Vein of Galen malformation is a developmental anomaly that usually appears late in pregnancy. Because third-trimester sonography is still not routinely performed in most units, many cases will be detected only in the neonatal period when clinical symptoms arise. The main presenting feature is cardiac failure; therefore, vein of Galen malformation should be strongly considered in the differential diagnosis of cardiac insufficiency in the first week of life. Perinatal predictors of poor outcome have been suggested, including assessment of the width and blood flow of the straight sinus, the presence of steal retrograde aortic flow, and the presence of signs of high-output cardiac failure. However, a full clinical evaluation during the first days of life is still the best way to determine the best timing for intravascular embolization. In the case described here,
we used 3D power Doppler angiography to further characterize the vascular anomaly in the fetal brain. In keeping with the findings of previous authors, we confirmed that this noninvasive technique generated high-quality images comparable with those generated by conventional neonatal angiography for recognizing the dilated vein of Galen, the straight sinus, and abnormal arterial vessels feeding the malformation. We used an automated sweep for acquiring the volume sets, whereas other authors had used a freehand technique. The ability to generate automatic slices certainly improves the quality of the images by reducing the possibility of motion artifacts. In addition, the reduction in the acquisition time to 2 to 4 sec, depending on the data volume, is another notable advantage of this technique over traditional MRI because it allows almost immediate depiction of the vascular anomaly. Superimposed gray scale sonography (glass body mode) can also assist in the diagnosis because it allows exact localization of vessels in relation to fetal tissue structures.

In conclusion, this report suggests that 3D power Doppler angiography adds useful information in the prenatal evaluation of vein of Galen malformation. Because this technique provides images similar to those produced by conventional neonatal angiography, 3D power Doppler angiography should be the first-line investigation and could potentially replace MRI in the prenatal noninvasive evaluation of arteriovenous malformations of the fetal brain.

References