The significance of 3D power Doppler in prenatal diagnosis and the evaluation of the anatomical structure of vein of Galen aneurysmatic malformation: case report

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Summary

Prenatal diagnosis of vein of Galen aneurysmatic malformation (VGAM) is made with the use of color Doppler, while in B-mode it is seen as a centrally placed supratentorial cystic structure. 3D-power Doppler (3D-PD) is a method that enables precise visualization of the vascular anatomy of this complex malformation. In our case, VGAM was detected in the 33rd week of gestation with power Doppler, and the use of 3D-PD enabled better visualization of the angioarchitecture and detection of feeding and drainage vessels of aneurysmatic widening. The diagnosis was confirmed postnatally with the use of MRI. A prenatal study of the angioarchitecture could have prognostic significance as well as being important in the therapeutic approach during the postnatal period.

Key words: Intracranial cystic structure; Vein of Galen aneurysmatic malformation; Prenatal diagnosis; 3D-power Doppler.

Introduction

One of the severe causes of mortality and morbidity in the early neonatal period and during childhood is vein of Galen aneurysmatic malformation (VGAM), a rare congenital disorder, forming up to 1% of total cerebral vascular disorders [1]. VGAM is viewed by unltrasonography as an anechoic tubular intracranial lesion in the midline, superiorly located in relation to the cerebellum, and with high turbulence flow detected by color Doppler [2]. Due to the possibility of the three-dimensional power Doppler angiography (3D-PD) to provide views comparable to those formed by traditional neonatal angiography, additional valuable information can be obtained in the evaluation of VGAM [3, 4].

We present a case of aneurysmatic malformation of the vein of Galen, prenatally diagnosed by power Doppler with particular attention to the significance of 3D-PD in terms of evaluation of anatomical structure of this vascular malformation, and accordingly the potential prognostic role of this method.

Case Report

A 26-year-old women became pregnant after a two-year history of infertility. Ultrasound (US) was performed with a Voluson 730 (GE Medical Systems, Kretz, Austria) which is equipped with an abdominal transducer of 4-8 MHz. Previous US screening examinations performed in the 9th, 12th, and 22nd gestational weeks revealed a vital singleton fetus without apparent structural abnormalities in the uterus bicornis (Figure 1). A cystic structure 22 x 13.9 mm in diameter, located dorsal to the tectum was visualized on B-mode imaging at the 33rd gestational week. Blood flow in this mass was demonstrated using power Doppler (Figure 2). To understand what sort of vascular lesion was involved and also to study its angioarchitecture, 3D-PD was used. Using the 3D-PD made possible not only a confirmation of the diagnosis, but also a more precise orientation of the area, meaning a visualization of the anatomy of the earlier noticed vascular anomalies (Figures 3 and 4). This technology clearly displayed the existence of arteriovenous communication between the arterial circle of Willis, more precisely the a. cerebri posterior, and aneurysmatic widening of the vein of Galen (Figures 4 and 5), as well as a widening of the straight sinus opening into which blood drains from the lesion (Figures 3, 4, and 5). The morphology of the fetus as a whole was normal, without any signs of hemodynamic disorders. The fetal heart was also morphologically normal with normal size.

The birth was completed at term by cesarean section and resulted in a vital female baby weighing 2,900 g. The Apgar score was 8/9 at one and five minutes after birth, respectively. The prenatal diagnosis was confirmed postnatally by echosonography and magnetic resonance imaging (MRI) of the central nervous system (Figure 6). Echocardiography revealed the existence of pronounced hypertrophy of the myocardium of both ventricles, particularly the right one. At that moment, the shunt was not large, so there were no elements to cause congestive heart insufficiency. Echo-sonographic findings in the abdomen were regular with no signs of ascites. All the laboratory findings were within the normal parameters. Bearing in mind that the newborn was hemodynamically and neurologically normal, the medical team that was following the progress of the newborn took an expectative position. At this time the child is two years old, without any negative implications and is under regular pediatric and neurological supervision. The child is developing well with regular neurological development and with no aneurysm enlargement. The cardiac state is stable with no signs of heart function decrease. The left ventricle is slightly smaller in size than it was at previous examination with discrete remodeling, first of all, of the myocardial mass, with normal ranges of the heart insufficiency biomarkers.

Revised manuscript accepted for publication March 20, 2012

Fig. 1

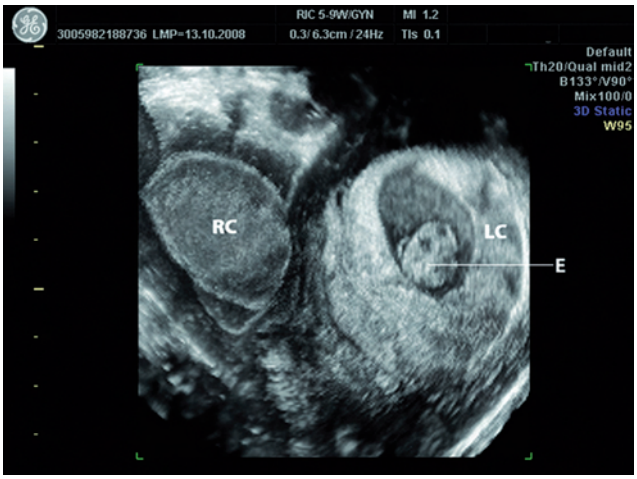

Fig. 2

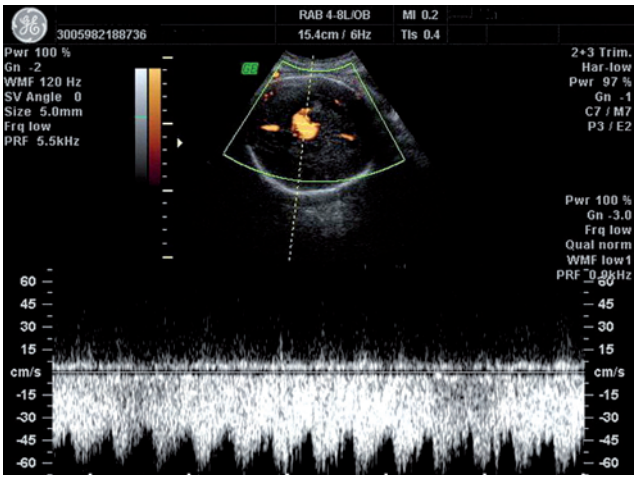

Fig. 3

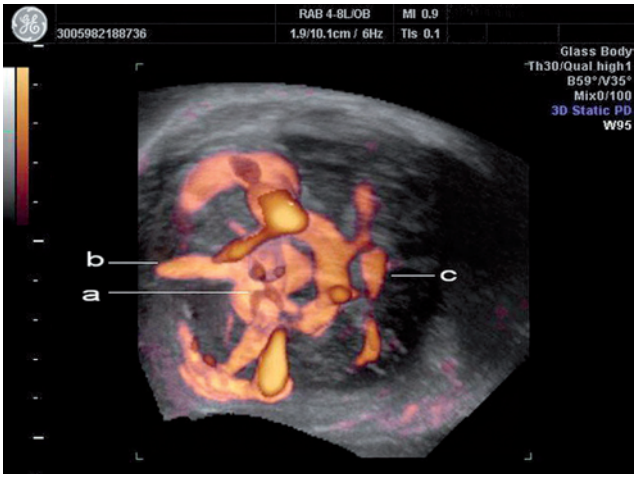

Fig. 4

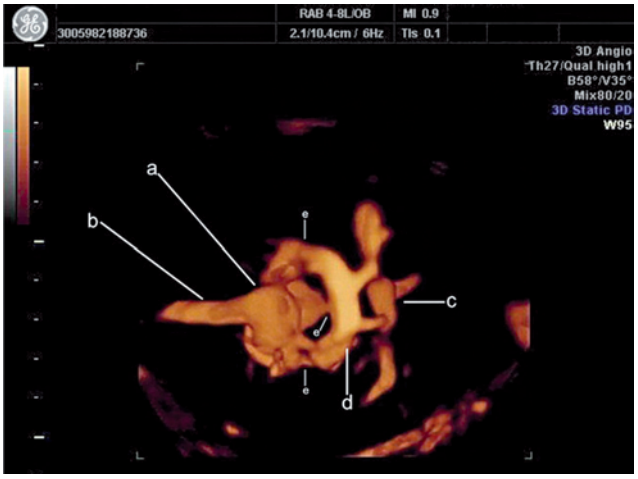

Fig. 5

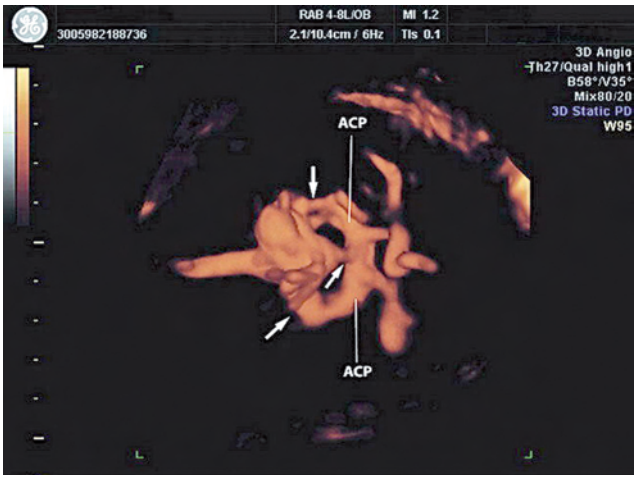

Fig. 6

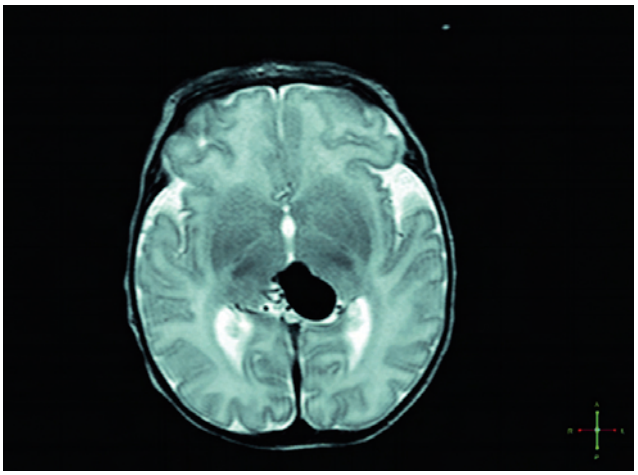

Figure 1. — Three-dimensional rendering technique. The scan shows uterus bicornis in the ninth week of gestation with an embryo (E) embedded in the left horn (LC) of the uterus. (RC) the right horn of the uterus.
Figure 2. — Doppler analysis showing pulsatile venous blood flow pattern through an aneurysm of the vein of Galen.
Figure 3. — A 3D power Doppler analysis of the angioarchitecture of a VGAM in body glass B-mode. Scan showing the aneurysm (a), dural sinuses (sinus rectus (b)) and the circle of Willis (C) as well as shunts between the circle of Willis and the aneurysm.
Figure 4. — Scan showing the aneurysm of the vein of Galen (a) which directly drains in the sinus rectus (b) by 3D-PD mode. There is also the circle of Willis (c) as well as feeding arteries (e) which mainly come from a.cerebri posterior (d).
Figure 5. — By rotating Figure 3, the relation between the aneurysm and the circle of Willis is shown better. The feeding arteries (arrows) mainly come from a.cerebri posterior (ACP).
Figure 6. — Postnatal magnetic resonance imaging showing aneurysmal malformation of the vein of Galen.

Discussion

VGAM, which makes up 1% of all intracranial malformations, is a complex arteriovenous malformation which appears in different forms, from a considerable widening of the vein itself up to multiple communications between the vein of Galen system and the cerebral arteries (carotid and/or vertebral basin) [3, 5]. The etiological origin of this disorder is controversial but it is thought that it appears between the 6th and 11th week of embryonic development. The assumption is that reduced capillary resistance, probably combined with stenosis of the dural sinus leads to progressive enlargement of the anterior segment of the medial prosencephalic vein (vein of Markowski), which is considered to be the precursor of the vein of Galen and which normally regresses, and to the formation of an aneurysmatic component typical of the arteriovenous malformation of the vein of Galen [6]. Several types of VGAM have been recorded. Arteriovenous communications create an intracranial shunt (L-R shunt) which can cause cardiomegalia with cardiac insufficiency and non-immunological hydrops fetalis as a result of cardiac decompensation. Low resistance in utero circulation can reduce the flow through the fistula and minimize cardiac decompensation, but a sudden increase in systematic vascular resistance after birth results in a considerably increased flow through arterio-venous communication, resulting in cardiac weakness. Changing the course of cerebral circulation can lead to cerebral infarction and the resulting porencefalia. This is often accompanied by hydrocephalia and it is thought that it occurs because of aneurysm pressure on the Sylvian aqueduct or because of increased intracranial venous pressure [7].

While the malformation of vein of Galen may be diagnosed by pulse and color Doppler, it is still hard to accurately localize feeding arteries and venous drainage, and yield satisfactory prognosis. Using MRI antenatally has turned out to be useful in the final diagnosis [8]. This paper showed that it is possible to perform not only diagnosis, but also the accurate localization, as well as detailed identification of elements forming VGAM by using 3D-PD. The advantage of 3D-PD is the possibility to memorize volume and additional processing, i.e. rotation and visualization of scans from various angles, which enables identification of structures unavailable to 2D technology.

Several factors may have an effect on the prognosis of this anomaly. Yuval *et al*. illustrated for the first time prenatal sonographic indices that may predict fetal outcome. In their report, anatomic brain changes, hydrops, dilated drainage tract, multiple (five or more) feeding vessels, dilated jugular vein and/or inferior vena cava, retrograde aortic flow, and cardiomegaly were associated with adverse outcome [9]. The volume of the aneurysm is also a factor that may influence outcome [3, 10]. Sepulveda *et al*. reported ventriculomegaly, cardiomegaly, and dilated neck vessels being a common associated finding [1]. On the other side, Pilu *et al*. reported changes of the cerebral structure in cases with vein of Galen aneurysm being a negative sign [11].

The true therapeutic approach has not yet been established. Overall, what is needed is a detailed anatomic picture of the changes in order to evaluate a method that could be suitable in each individual case. As many studies have shown, the neurological outcome of subjects with VGAM [6, 12, 13] is highly improved by endovascular embolization [6, 12, 13]. Transtorcular embolization of Galen's aneurysm through the confluence of sinuses after delivery is possible owing to identifying straight sinus drainage [14].

The age of the newborn infant is a critical factor when talking about the success of endovascular therapy. As long as the patient shows no signs of heart or kidney weakness and is free of neurological disorders, meaning if the patient is clinically stable, it is better to delay treatment until the fifth or sixth month [6, 13]. There is evidence of rare cases of spontaneous thrombosis or regression of arteriovenous malformation. Patients without any symptoms may be treated with adequate medical therapy, and with repeating angiography with the possibility of occurrence of spontaneous thrombosis [6, 15].

Even though angiography represents the gold standard in the precise evaluation of the angioarchitecture of this malformation, including the detailed anatomy of the supply arterial blood vessels, as well as the venous drainage which allows precise endovascular access and successful therapy, the pictures obtained in the case described are comparable with postnatal diagnostic methods. It has been demonstrated that the use of 3D-PD enables detailed identification of all elements of VGAM such as feeding arteries and venous drainage or the localization of the same. These findings suggest that 3D-PD is one more additional prenatal diagnostic method which, as well as MRI, can also be very informative and significant not only in the context of the diagnosis of VGAM, but also in the verification of the seriousness of the illness.

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