

Prenatal Ultrasound Diagnosis of Dural Sinus Malformation: Analysis of Three Cases And Review of the Literature

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Abstract

Three cases of prenatal ultrasound diagnosis of sinus venous malformation of dura mater have been described, which in two cases was complicated by thrombosis of confluence of sinuses during the second trimester of pregnancy. In one case, the baby died 10 days after birth, and in two cases, pregnancies were aborted for medical reasons. The issues of prenatal diagnostics and perinatal outcomes in cases of sinuses' thrombosis in the dura mater in the fetus have been discussed based on the previously published observations.

Keywords: fetus, dural sinuses, dural sinus malformation, thrombosis, prenatal diagnosis, perinatal outcome.

INTRODUCTION

Sinus venous malformation (SVM) is a rare congenital abnormality characterized by enlarged sinuses in the dura mater. Abnormal development most often affects the confluence of sinuses (torcular Herophili), i.e. the place of their confluence, localizing in the posterior cranial fossa, and is often accompanied by thrombosis.

Etiology of the dura mater sinuses' thrombosis in the fetus remains not fully studied. The literature discusses the following possible causes: trauma, infection, hypercoagulopathy, preeclampsia; however, in 40% of cases, the reason remains unknown. In some cases, thrombosis may develop post primarily as a result of SVM, which is probably a consequence of abnormal embryonic development (uncontrolled development of dorsal sinuses).

In SVM cases, ultrasound examination of the posterior cranial fossa of the fetus reveals hypoechogenic or anechogenic formations. With that, the pathologic process most commonly affects confluence of sinuses (torcular Herophili); therefore, the formation has triangular shape, mimicking the shape of the dilatated sinus. In the Doppler Color Mapping (DCM) mode, pulsating blood flow is seen in the dilatated sinus. In cases of thrombosis overlap, an echogenic component with clear contours is visualized in the dilatated sinus, and in case of development of absolute thrombosis, blood flow is not observed in the DCM mode.

Currently, the published literature describes individual cases of prenatal diagnostics of brain SVM in fetuses [1-20]. Due to the rarity of this pathology, the authors present 3 own clinical observations.

MATERIAL AND METHODS

Observation 1. Patient B., 33 years old, contacted a private medical center for consultative ultrasound examination in the 22nd week of pregnancy. The ultrasound examination in the 17th week of pregnancy did not reveal any pathological changes and congenital malformations in the fetus. In the 20th week of pregnancy, an anechogenic formation with a diameter of 28 mm was found in the fetus, containing fine particulate slurry and parietal echogenic inclusion. Based on these data, the presence of arachnoidal/porencephalic cyst was assumed. Magnetic Resonance Imaging (MRI) was used to confirm the prenatal diagnosis, which stated the presence of "an oncoma of the posterior cranial fossa with compression of the cerebellum and posterior (more to the left) divisions of the cerebral hemispheres (might correspond to a hematoma), the presence in its thickness of an expanded vessel and the area of hemorrhagic suffusion (probably, an arteriodural fistula)".

Observation 2. A primip reprenant patient I., 35 years old, went to a private center for Doppler velocimetry of blood

vessels in the fetus due to previously diagnosed hydramnion. 10 days before the visit, she was subjected to an ultrasonic study at the nursing home with the appropriate conclusion "30-31 weeks of pregnancy. Hydramnion". No pathological changes were found in the fetus.

Observation 3. Patient A., 26 years old, was sent for consultative ultrasound examination in the 21-22 weeks of pregnancy due to the detected brain oncoma in the fetus during the screening ultrasound examination in the second trimester of pregnancy. No other congenital malformations and markers of chromosomal anomalies were found.

Ultrasonic studies were performed on apparatus Voluson E8 (GE) with the use of the transabdominal volumetric scanning transducer RAB6-D, and Voluson S6 (GE) using the transabdominal volumetric scanning transducer RAB 4-8.

RESULTS

Observation 1. Consultative ultrasound examination in the 22nd week of pregnancy diagnosed the accordance of the phetometric characteristics to the period of pregnancy. Fetus position was longitudinal with cranial presentation. Transabdominal access confirmed the presence of a 45x34x47 mm abdominal oncoma with finely divided content.

For further prenatal diagnosis, transvaginal scanning was additionally performed, which clearly identified the triangular shape of the oncoma in the projection of confluence of sinuses, and homogenous echogenic parietal component of rounded shape with the diameter of 10 mm. In the Doppler directional power mapping mode, with the use of low values of the velocity scale (4 cm/s), no blood flow was detected in the expanded confluence of sinuses. Based in these data, a conclusion was made about sinus malformation with thrombosis.

During prenatal consultation, the patient made a decision in favor of abortion.

Observation 2. During the ultrasound examination of the fetus, severe cardiomegaly without any evidence of congenital heart disorders, and significant expansion of jugular veins on both sides were noted. The detailed ultrasound examination of the structures of the brain of the fetus, including tomographic mode of volumetric echography, revealed no changes. In the DCM mode, a pronounced aneurysm of the circle of Willis and sinuses was discovered, especially the upper ones, with a turbulent blood flow. No data about aneurysm of the vein of Galen and arteriovenous malformations were found during the ultrasonic volumetric angiography. Based in these data, a suggestion was made about the presence of lesions of the sinuses in the dura mater, and magnetic resonance imaging (MRI) was recommended for confirming the prenatal diagnosis.

By the results of MRI performed almost 4 weeks later, thrombosis of the upper, sagittal and transverse sinuses was

diagnosed without malformation in the vein of Galen and without damage of brain matter. Thus, the sinus venous malformation was complicated by thrombosis. The patient delivered, and the baby was immediately transferred to the children's hospital, where it died in the 10th day.

Observation 3. Consultative ultrasound examination in the 21-22nd weeks of pregnancy diagnosed accordance of the phetometric characteristics to the period of pregnancy. Ultrasound examination of the structures of the brain of the fetus confirmed the presence of a 46 x 35 x 30 mm hypoechogenic abdominal oncoma of triangular shape located in the posterior cranial fossa, and containing a 14.8 x 11.7 mm homogeneous echogenic component with clear contours.

In the directional power Doppler mapping mode, with the use of the values of velocity scale of 11 cm/s, no blood flow was detected in the expanded confluence of sinuses. Based in these data, a conclusion was made about sine malformation with thrombosis. During the prenatal consultation, the patient made a decision in favor of abortion.

DISCUSSION

The time of SVM diagnostics in course of the three observations ranged between the 21st and 32nd weeks of pregnancy, the average being 25 weeks. SVM that was complicated by thrombosis of confluence of sinuses was diagnosed in 2 fetuses in the 22nd week of pregnancy. In these cases, ultrasound study of brain structure revealed an abdominal hypoechogenic oncoma of triangular shape located in the posterior cranial fossa containing a homogeneous echogenic component with clear contours. In one case, SVM diagnosed in the 32nd week of pregnancy, a pronounced aneurysm of the circle of Willis and sinuses was discovered, especially the upper ones, with a turbulent blood flow, which was later complicated with thrombosis. All cases had unfavorable outcome: in one case, death of the baby in the 10-th day after birth, and abortion in the other two cases.

The authors managed to find complete clinical description of 29 cases of SVM diagnostics during prenatal ultrasound examination, including the described cases. The published cases of SVM were diagnosed in the period between the 19th and 35th weeks of pregnancy, the average being 25.2 weeks [1–20]. According to the published data, prenatal ultrasound diagnostics of sinuses' malformations in the dura mater of the fetus is possible starting from the 19th week of pregnancy [9], which corresponds to the time of screening ultrasound examination in the second trimester of pregnancy. Diagnostic criteria for sinuses' malformations in the dura mater during the prenatal period are the following: 1) a triangular formation in the posterior cranial fossa (most common localization of SVM is confluence of sinuses), behind the cerebellum with its forward displacement; 2) in the DCM mode, pulsating, often turbulent blood flow is detected in the dilatated sinus; 3) in cases of thrombosis, a homogeneous echogenic component with clear contours is visualized in the dilatated sinus, and in case of development of absolute thrombosis, blood flow is not detected in the DCM mode.

In case of SVM, according to published data [1-20], prediction may range from completely favorable to death after birth. According to analysis of 29 published observations, including the reported cases, favorable outcome was observed in 14 (48.3%) cases, abortion was made in 7 (24.1%) cases, and death after birth occurred in 8 (27.6%) cases. Unfavorable predictory prenatal symptoms are associated with the following changes: ventriculomegalia, expressed compression and displacement of brain structures, cardiomegaly, tricuspid regurgitation, pericardial effusion, neck varication and ascite. According to some specialists, the size of the thrombus and the

degree of sinuses' dilatation do not have significant effect in the perinatal outcomes.

CONCLUSION

Thus, SVM is a rare congenital abnormality of sinuses in the dura mater of the fetus, which may be diagnosed starting from the second trimester of pregnancy. Diagnostic criteria for sinuses' malformations in the dura mater during the prenatal period are triangular formations in the posterior cranial fossa, pulsating, often turbulent blood flow in the dilatated sinus in the DCM mode; in cases of thrombosis, a homogeneous echogenic component with clear contours is visualized in the dilatated sinus. The unfavorable predictory prenatal symptom is the presence of associated changes.

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