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Prenatal findings and postnatal follow-up of a midline dural sinus malformation

Hana Shabana[1,2], Johannes Leidinger[3], Johan Wikström[4] and Ove Axelsson[1,5]

Abstract

Dural sinus malformation is a rare condition. We describe a prenatally detected case followed by repeated ultrasound scans and a prenatal magnetic resonance imaging examination. A substantial spontaneous regression was observed, which is associated with a favorable outcome. We believe that our observations, including a long postnatal follow-up, will add to the present knowledge of prenatally detected cases, and thus improve management of the pregnancies as well as our possibilities to counsel the parents-to-be.

Keywords

Dural sinus malformation, prenatal diagnosis, ultrasound, magnetic resonance imaging, outcome

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Introduction

Dural sinus malformation (DSM) is an uncommon congenital anomaly seldom met by feto-maternal physicians.[1] It is better known by pediatric neurologists and neuroradiologists. The etiology of DSM is far from established. The condition often includes partial thrombosis of the affected sinus, possibly secondary to slow flow in the dilated sinus and is most often associated with arteriovenous shunts. The outcome for the fetus in prenatally diagnosed cases is varying, and our ability to give information concerning the prognosis to the parents-to-be is limited.

The case presented here adds to our previous knowledge by presenting frequent examinations throughout pregnancy with imaging and Doppler ultrasound and prenatal magnetic resonance imaging (MRI). Moreover, we have been able to follow the child postnatally up to an age of three and have a control person in the shape of a healthy twin brother.

Case report

A 35-year-old primipara with a dichorionic twin pregnancy came for a scheduled scan at 27 weeks. A minor hyperechoic structure located superior and anterior of the cerebellum was observed in twin 1 (Fig. 1). At a previous scan at 18 weeks no deviant findings were noticed. TORCH-serology was negative. At 31 weeks the biparietal diameter (BPD) was substantially enlarged, and so was the occipito-frontal diameter. A homogenous ovoid cyst-like structure measuring 65×53 mm and containing concentric rings was seen in the posterior part of the skull. The ventricular system was clearly widened. Doppler examinations, including the fetal middle cerebral artery, were normal and remained so throughout pregnancy. No turbulent flow was observed at any occasion within the cystic structure or elsewhere (Fig. 2(a) to (c)). Tests for idiopathic thrombocytopenia and

[1]Centre for Clinical Research Sörmland, Uppsala University, Eskilstuna, Sweden
[2]Department of Obstetrics and Gynecology, Nyköping Hospital, Nyköping, Sweden
[3]Department of Obstetrics and Gynecology, Mälar Hospital, Eskilstuna, Sweden
[4]Department of Surgical Sciences, Radiology, Uppsala University, Uppsala, Sweden
[5]Department of Women's and Children's Health, Uppsala University, Uppsala, Sweden

Corresponding author:
H Shabana, Department of Obstetrics and Gynecology, Nyköping Hospital, olrogs vägen 1, Nyköping 611 85, Sweden.
Email: hana.shabana@regionsormland.se

thrombocyte antibodies were negative. At 32 weeks BPD, although still enlarged, had decreased as well as the cyst-like structure (Fig. 3). The next scan, at 33 weeks, showed a continuous regress (Fig. 4). Days later, still in week 33, an MR examination showed a large expansive lesion in the back of the skull compressing cerebellum. At the bottom of the lesion a rounded structure was seen, interpreted as a thrombus or hemorrhage. The fourth ventricle was compressed leading to obstructive hydrocephalus. The parenchyma was assessed as normal (Fig. 5(a) and (b)). The last scan was done at 35 weeks and showed a substantial regress of the cystic structure. BPD was still enlarged and the hyperechoic structure still visible (Fig. 6(a) to (c)). Delivery was now undertaken through a caesarian section. Twin 1, a boy, weighed 2232 g and had an Apgar of 8 at 1 min and 10 at 5 min.

Twin 1 was examined by ultrasound, MRI and computed tomography a few hours after birth and a partly thrombosed DSM was diagnosed. Later, MR examinations have exhibited a normalization including recanalization of vessels. The child received no specific treatment.

Twin 1, now more than three years old, has up to now displayed normal growth and neurological development, well in line with his twin brother.

Discussion

This case had its final diagnosis after birth. With the wisdom of hindsight the hyperechoic mass observed at

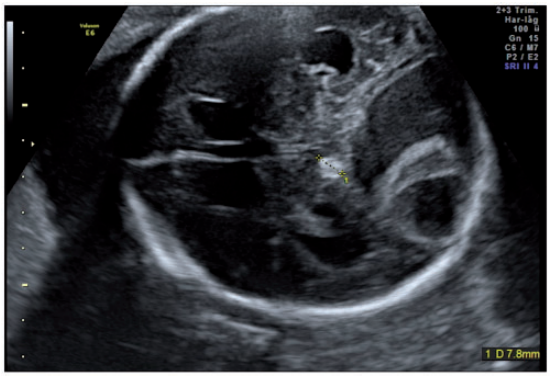

Fig. 3. Ultrasound scan at 32 weeks displays regression of the cystic structure.

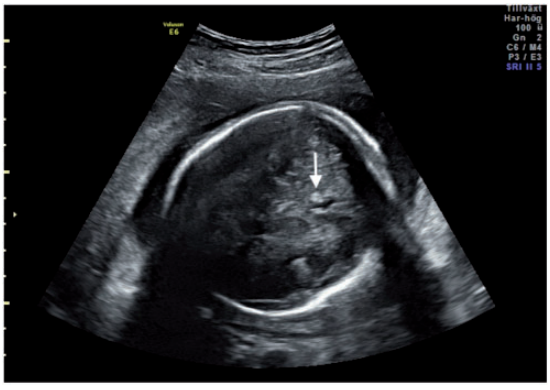

Fig. 1. Ultrasound at 27 weeks of the fetal head with a minor hyperechoic structure (arrow).

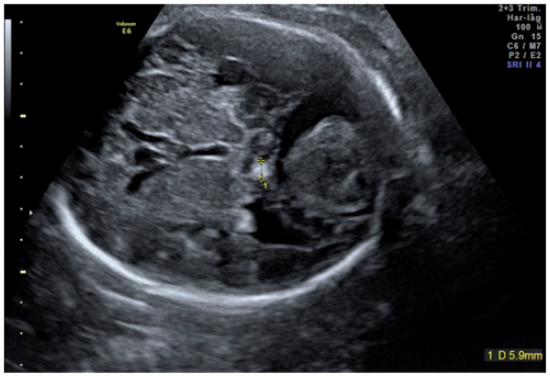

Fig. 4. Ultrasound scan at 33 weeks reveals continuous regression of the cystic structure and normalization of ventricle width.

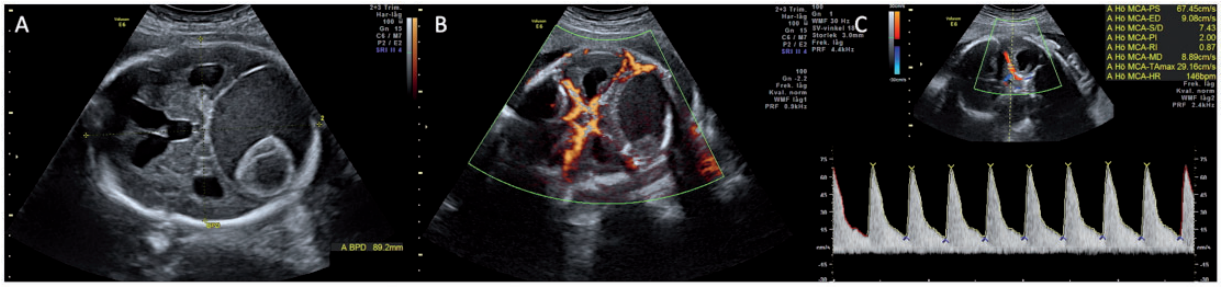

Fig. 2. Ultrasound scan at 31 weeks (a) shows a large cyst-like structure containing concentric rings in the posterior part of the skull. The ventricles are widened. Power Doppler shows normal flow pattern in the Circle of Willis (b). A normal waveform is observed in the middle cerebral artery (c).

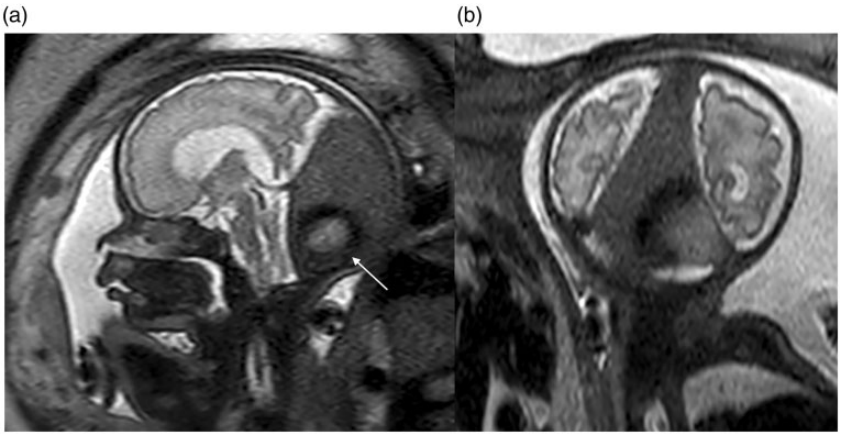

Fig. 5. Sagittal (a) and coronal (b) T2-weighted MRI at 33 weeks show an expansive lesion in the back of the skull, compressing the cerebellum. A rounded structure (arrow in a) is observed, interpreted as a thrombus or hemorrhage.

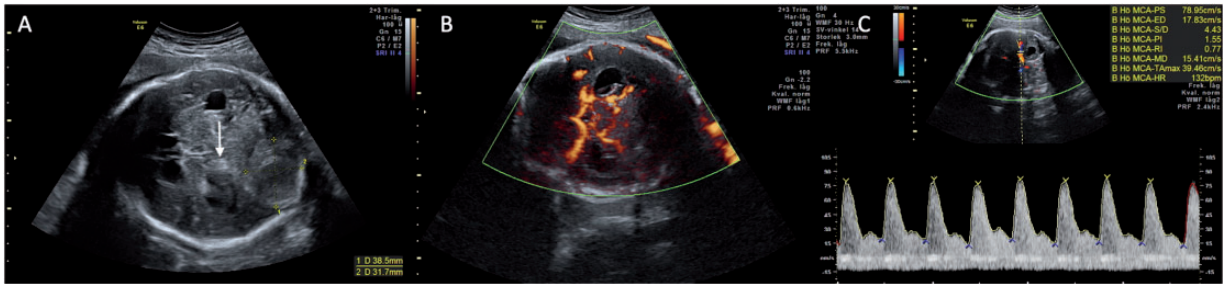

Fig. 6. Ultrasound scan at 35 weeks (a) shows a substantial regression of the cystic lesion. The hyperechoic structure (arrow) is still visible. Power Doppler shows normal flow pattern in the Circle of Willis (b). A normal waveform is observed in the middle cerebral artery (c).

the first scan (Fig. 1) is most probably a thrombosed arteriovenous shunt.[1] Nothing was then said about an anechoic mass but when we re-examine images such a mass can be suspected in the posterior part of the head. The anechoic mass observed at week 31 (Fig. 2) is the greatly widened dural sinus and the concentric rings a thrombus. The prenatal MRI at 33 weeks gave more detailed information including the finding of normal brain parenchyma emphasizing the important role of prenatal MR examinations.[2]

DSM is a congenital vascular malformation including an extensive dilatation of a dural sinus, which may be due to persistence of a physiological prenatal ballooning of dural sinuses.[3] Most often DSM is associated with arteriovenous shunts[1] between the dural sinus and choroidal arteries. They can result in an increase of blood returning to the fetal heart and lead to heart failure, which can be observed on ultrasound as fetal hydrops.[4] Our case had no such signs.

A mortality figure of 22% has been reported,[5] the figure being higher for postnatally diagnosed cases. Absence of the following prenatal signs is connected to a good outcome: absence of lesions in the brain parenchyma, of ventriculomegaly, of arteriovenous shunts and of fetal heart failure. The presence of the following prenatal signs is connected to a good outcome: presence of regression of the DSM, of a thrombus and of an increase of the size of the thrombus.[5,6] The present case displayed several findings speaking in favor of of a good outcome, namely absence of heart failure, absence of parenchymal lesions, and prenatal regression of the cystic structure. One ominous sign, ventriculomegaly, was however diagnosed, which demonstrates that a substantial widening of the ventricles can be consistent with a favorable outcome. When fetal anomalies are detected, termination of pregnancy is an option in Sweden and most European countries. The decision to terminate or continue the pregnancy lies in the hands of the pregnant woman. The information given on the prognosis for the child is of utmost importance for the woman when to decide.

It is important to remember that a spontaneous regression of even a prominent DSM with advancing gestational age is common[6] and that holds true even after a transient initial growth. Such a development was observed in our case.

Provided that the woman decides to continue her pregnancy, regular examinations including imaging, and Doppler ultrasound is recommended as well as a fetal MRI. Fetal MRI is a powerful tool to examine the fetal CNS.[7]

The most common postnatal intervention for newborns with DSM is embolization or surgery,[5] which was not needed in our case. Moreover, infants with a prenatally diagnosed DSM ought to be delivered at a hospital with access to pediatric neuroradiology and neurosurgery.[4]

Among differential diagnoses to consider are arachnoid or porencephalic cysts and Dandy–Walker malformations. A fetal MRI can rule out or verify these anomalies. The hyperechoic structure observed at the first US here can be interpreted as a sign of a fetal infection. Thus, TORCH serology is to be recommended. Fetal thrombocytopenia should be tested for since it can cause cerebral hemorrhage even prenatally.

In conclusion, to establish a correct diagnosis of a fetal anomaly is a pre-requisite to be able to give correct information on the prognosis for the child to the parents-to-be. Hopefully, our case report will add to the awareness of DSM among feto-maternal specialists as well as insight into the management and prognosis.

Declaration of Conflicting Interests

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Patient consent

Obtained.

ORCID iDs

H Shabana https://orcid.org/0000-0003-1787-820X
J Leidinger https://orcid.org/0000-0002-4957-9143
J Wikström https://orcid.org/0000-0002-9481-6857

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