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Prenatal Diagnosis of Torcular Dural Sinus Malformation: A Case Study

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Abstract

Torcular dural sinus malformations (tDSMs) are rare congenital defects representing a complex of vascular anomalies that have been grouped in one single unit. Although the current literature suggests a generally favourable prognosis for prenatally diagnosed tDSMs, there are still only limited data and published papers on the subject. Factors resulting in an adverse outcome of the fetuses and children have to be taken into consideration to determine precisely the nature of the consultation and management.

A 33-year-old primipara at 21 weeks, 5 days of gestation was referred to our clinic with the suspicion of a central nervous system (CNS) malformation of the fetus, and the diagnosis of tDSM with thrombus was made. No factors contributing to an adverse outcome such as arterialization of the lesion, ventriculomegaly or neuroparenchymal damage were present. The pregnant woman was scheduled for regular sonographic and magnetic resonance imaging (MRI) controls. In the third trimester the lesion decreased in size, which is a key imaging marker for a favourable prognosis. The child was born in term, and the latest neurological examination at the age of six weeks is without pathological findings.

This case study demonstrates a prenatally diagnosed tDSM with a favourable outcome with a regression in the size of the lesion during the prenatal period.

CASE REPORT

INTRODUCTION

Torcular dural sinus malformations (tDSM) are rare cerebrovascular malformations, which were first described in 1996. However, under the diagnosis of tDSM we can find different conditions with complex angioarchitectural features. This is why among tDSMs we have some cases with a very good prognosis and some cases with a serious impact on the fetus (child) with a poor outcome. According to authors, it is important to distinguish tDSMs with arteriovenous shunts and thrombosed tDSMs. The first group is usually associated with high morbidity and mortality. On the other hand, in the second group the lesions usually resolve spontaneously over the time and have a good prognosis (Liby *et al.* 2020). The most characteristic anatomical feature of tDSM is a torcular lake replacing the confluence of sinuses. Most of the lesions develop thrombus at some point. On the ultrasound tDSMs appear as triangular an- or isoechogenic malformations in the location of torcular herophili with hyperechogenic thrombus, if present. An important sign is the arterialization of tDSM, which is seen either directly as a feeder (a feeding vessel) or by demonstration

of a flow inside the lesion during sonographic examination. Other factors determining the prognosis are: ventriculomegaly and parenchymal haemorrhage in the surrounding tissue (Mankad *et al.* 2020).

Sometimes even determining the diagnosis itself may pose difficulties, and other malformations, such as arachnoid cyst, tumour or vein of Galen malformation, should be taken into consideration in the process of a differential diagnosis.

Prenatal management consists of regular sonographic and MRI controls. Postnatal management is a follow-up in asymptomatic cases and endovascular treatment in cases with confirmed arterialization, and in the most severe cases with hydrocephalus and neuroparenchymal damage often only conservative treatment is offered. (Liby *et al.* 2020; Yang *et al.*)

CASE DESCRIPTION

A 33-year-old primipara at 21 weeks, 5 days of gestation was referred to our clinic due to the suspicion of a fetal malformation of CNS during a routine second-trimester sonography. The patient's medical and family history were unremarkable, and the previous sonographic findings were considered normal. The initial sonographic examination showed pathology of 25x13mm located above the posterior fossa at the level of torcular herophili (in the differential diagnosis we considered this to be an arachnoid cyst or tumour), without signs of vascularization or blood flow inside the lesion (Figure 1). No associated structural malformations of the CNS or other organ systems were identified. The pregnant woman underwent the fetal MRI at 22 weeks, 2 days, which showed tDSM (size: 35x20x31mm) with a thrombus (Figure 2). Also a slight displacement of the left occipital lobe and cerebellum were diagnosed, without any evidence of vetriculomegaly or destructive lesions of the brain. The pregnant woman underwent amniocentesis with the result of a normal karyotype of the fetus. The possibility of an infectious origin of the process was excluded. Also the thrombophilia status of the mother was negative. The pregnant woman was scheduled for repeated sonographic and MRI controls. During the regular ultrasound scans the fetus had an estimated fetal weight proportional for the week of gestation (without asymmetry between the standard biometric parameters) and normal amniotic fluid volume. Perfusion and pulsatility both in the umbilical and middle cerebral arteries were physiologic during the prenatal scans. In the third trimester a regression in the size of the lesion was observed. At 35 weeks, 3 days of gestation a residual triangular formation with thrombus (tDSM) of 20x10x20mm in size in the occipital region appeared on the fetal MRI (Figure 3). There were no signs of venous congestion, ventriculomegaly or ischemic lesions of the brain.

An elective caesarean delivery was performed at 40 weeks of gestation (the Apgar scores were: 10,10 and 10 after 1, 5 and 10 minutes, respectively, birth weight: 3800g, length: 51 cm, sex: male). Postnatal adaptation and neurological presentation were normal immediately after birth.

The last neurological examination at 6 weeks of age of the child was physiologic, without signs of pathology or developmental delay. The hematologic consultation of the child showed normal levels of Antithrombin

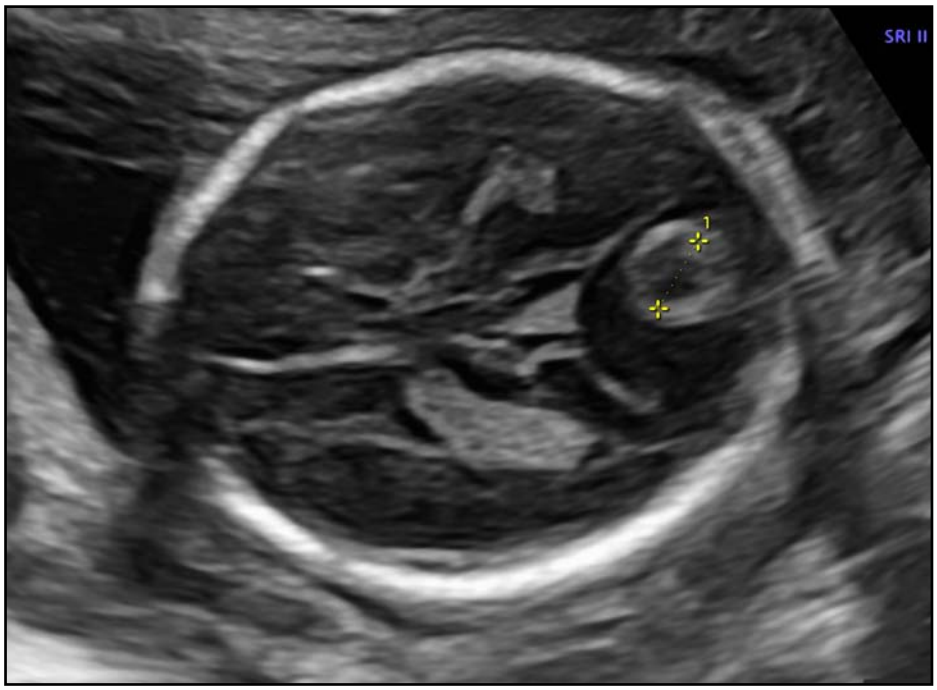

Fig. 1. The initial ultrasound at 21 weeks and 5 days. There is a torcular lake above the posterior fossa with visible thrombus of 9mm.

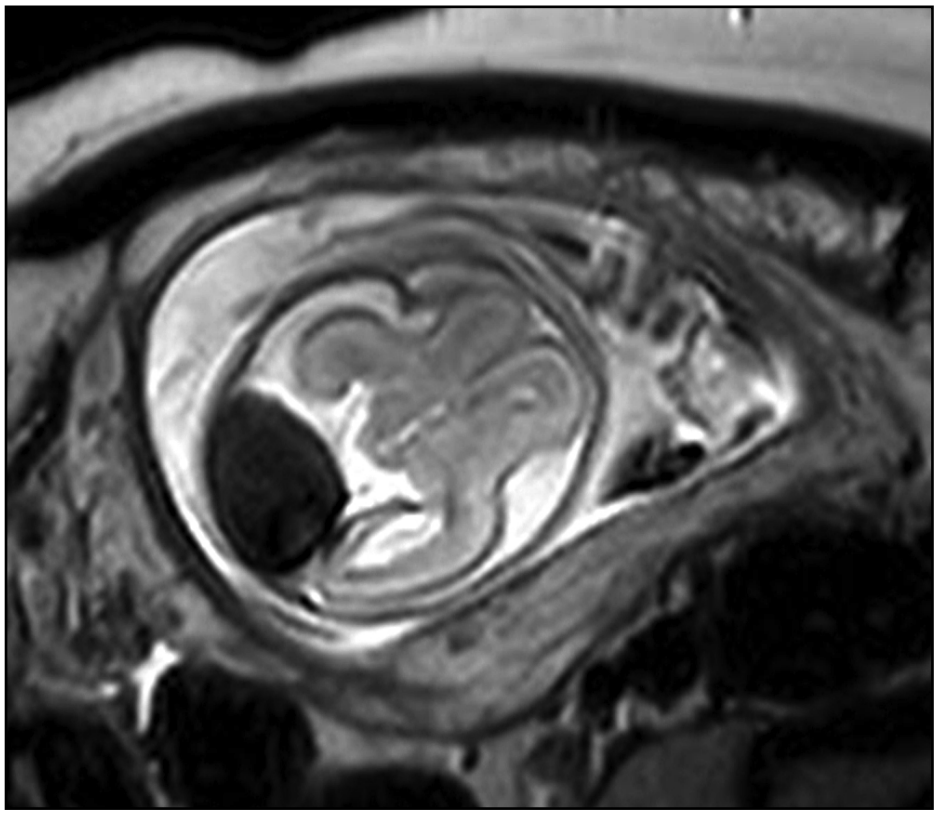

Fig. 2. Fetal axial T2W MRI at 23 weeks and 4 days shows a torcular expansion with thrombus

III and protein S. The levels of protein C were slightly lower, which was interpreted as a result of the immaturity of the hemostasis.

DISCUSSION

In the past the outcome of the fetuses and children with tDSM was described as poor, which has been a source of consternation when counselling the parents of patients either during pregnancy or postnatally. On the other hand, recently published papers showed a favourable prognosis, especially for prenatally diagnosed tDSMs, when 75–90% of fetuses survived without neurological sequelae (Yang *et al.*).

Liby *et al.* (2020) proposed a classification system of tDSMs to predict their prognosis. According to findings of the authors, two factors play a fundamental role at the time of diagnosis: the arterialization of the lesion and brain damage. Patients without a feeder were grouped as grade I and have the best prognosis (the reported mortality in this group was 7.55% according to study). Worse outcomes and higher mortality rates were reported for the grades II and III, which were defined by the identification of scarce or multiple feeders. The poorest outcome was associated with grade IV, where brain damage was the most important criterion and the mortality rate was 75%. Most of the grade I tDSM resolve spontaneously during the prenatal period or during the first months of life, and only close monitoring is recommended in these cases. In tDSMs of grades II and III close monitoring and endovascular treatment of feeders postnatally is mandatory in order to avoid further decompensation. Patients with grade IV tDSM have a poor prognosis, and conservative treatment is recommended in most cases. Our case could be classified as grade I, with favourable prognosis.

Sometimes not only estimating the prognosis, but also stating the diagnosis of tDSM can be difficult since only limited data have been published on the subject so far (Robertson, 2018) Diogo *et al.* (2019) pointed out that the vast majority of fetal DSMs were referred to MRI examination with an incorrect diagnosis. Also in our case study at the time of the first sonographic evaluation a definite diagnosis wasn't made. This finding demonstrates the important role of interdisciplinary cooperation in the diagnosis and management of rare congenital malformations of the CNS (comprising tDSM).

CONCLUSION

tDSM represents a rare condition with a more favourable prognosis when diagnosed prenatally, according to the current level of knowledge. Our described case was one with a good prognosis and a regression in the size of the malformation during the pregnancy. The child has normal neurological development according to its age and no associated abnormalities on the

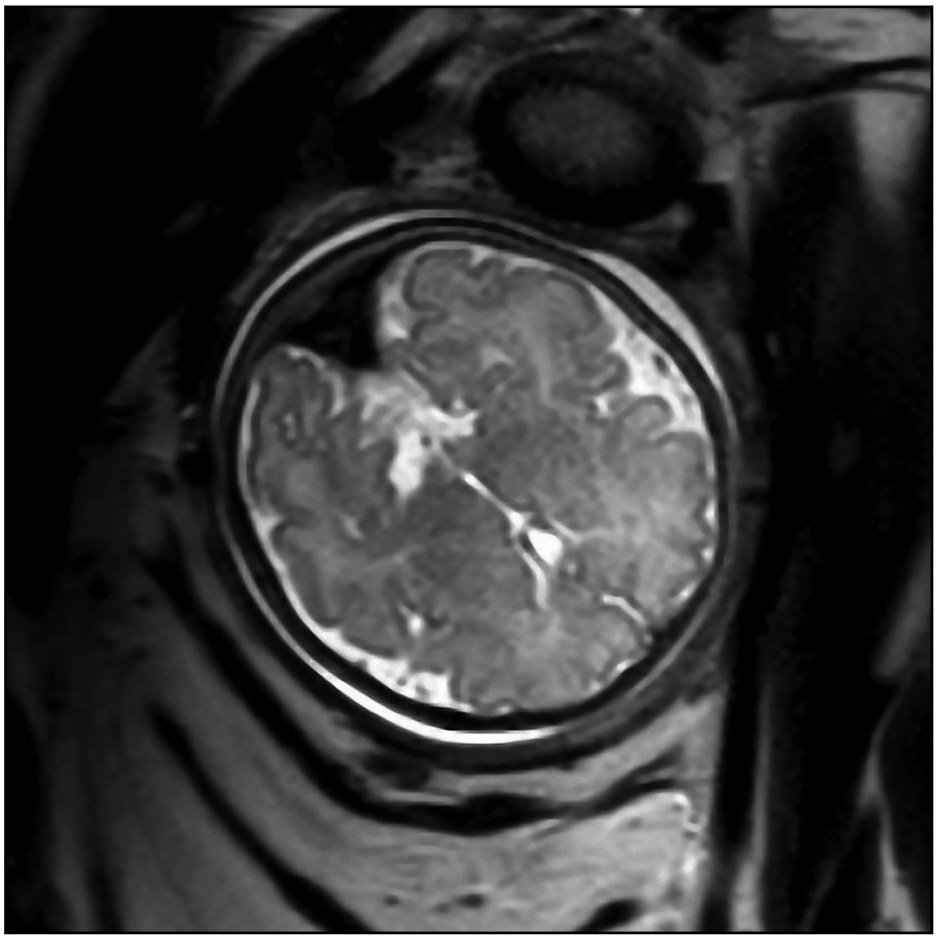

Fig. 3. The regression of tDSM documented on fetal axial T2W MRI at 35 weeks 3 days of gestation

ultrasonography of brain. Rare congenital malformations diagnosed prenatally such as the tDSM pose a difficulty when consulting future parents about the prognosis of the fetus, since only a limited number of cases and their outcomes have been published so far. With our case we wanted to emphasise a good prognosis of the tDSM without arterialization (feeding vessel or demonstration of pulsatile flow), and with no other associated abnormalities of the CNS (ventriculomegaly or parenchymal haemorrhage).

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