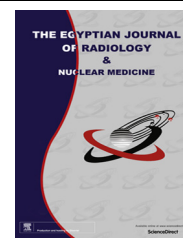




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## CASE REPORT

# Prenatal diagnosis of torcular herophili thrombosis with unfavored outcome and review of the literature



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### KEYWORDS

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**Abstract** Prenatally diagnosed thrombosis of the torcular herophili is a rare described entity. There are about 50 cases described in the literature till now. We here discuss the radiological findings of dural sinus thrombosis at the torcular herophili with extension into the superior sagittal sinus demonstrated on fetal magnetic resonance imaging (MRI) at the second trimester. After birth, the infancy was followed up to sixth months with serial MRI. During this period there was moderate reduction in the size of the thrombus. There was not any underlying cause for the thrombus both in the mother and the infancy. Unexpectedly, the cranial MRI revealed the signs of cerebral atrophy at the age of sixth month. Regarding the cerebral atrophy in a prenatally diagnosed torcular herophili thrombus, there was not a sample of this combination in the literature before. In this presentation we discuss the radiologic and clinical findings of prenatally diagnosed torcular herophili within the review of the literature.

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## 1. Introduction

The fetal dural venous sinus is a fairly uncommon entity which has been usually undiagnosed due to unfamiliarity. This condition causes serious neurological problems including ischemia

and hemorrhage which could lead to major causes of death. Even though there are several case series described in the literature, the exact cause and the pathophysiological mechanism are still a mystery. The evolution and the potential favorable outcome needs serial follow-up of magnetic resonance imaging (MRI) (1). On the behalf, the prognosis could variate in a wide range of spectrum including spontaneous resolution to severe development delay and death (2).

In this presentation, we discuss the radiological and clinical findings of torcular herophili in a prenatally diagnosed case with which is an extremely rare described in the literature.

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## 2. Case

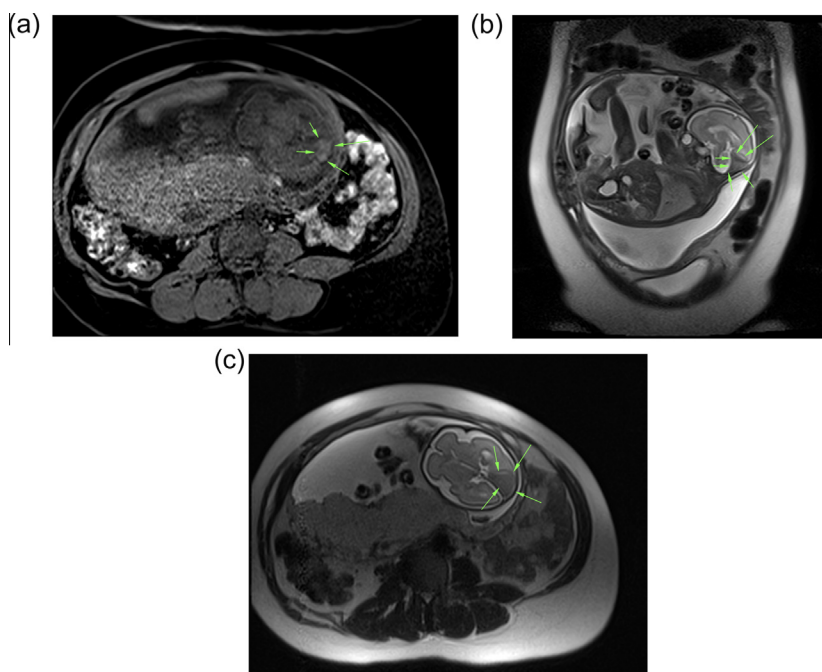
A 30 year-old Primida2Gravida2 was referred to radiology department at a gestational age of 27 weeks and 3 days for further detailed examination. The patient had no risk factor for the systemic diseases and underlying cause for the thrombus such as anemia, dehydration or coagulation disability. A midline retrocerebellar cystic structure was detected at the posterior fossa in the routine sonography by the perinatology physician.

The fetal MRI which was revealed at the 27th gestational age, demonstrated a large mass at the torcular herophili extending to superior sagittal sinus. The thrombus was isohyperintense to gray matter on T1-weighted images and isohypointense on the HASTE (T2-W) pulse sequences. The thrombus caused a slight mass effect on the cerebellar vermis within the displacement laterally and a bit superiorly. Except the posterior fossa findings, the fetal brain was normal for the supratentorial region including the ventricular system (Fig. 1(a)–(c)). To identify the vascularization of the cystic structure, Doppler sonography examination was obtained. Neither power Doppler, nor spectral Doppler imaging sequence showed any blood flow into the midline cystic structure. The patient refused to come for follow-up routine controls. An elective cesarean delivery was performed at 38 weeks gestational age in an outer centre. Birth weight was 3650 g. MR venous angiography and brain MRI were obtained on the third day of the newborn. Venous MR angiography showed filling defect in torcular herophili with small extension into the superior sagittal sinus (Empty Delta sign). The thrombus was hyperintense to gray matter on T1-W and hypointense on T2-W (Fig. 2(a)–(d)). During the follow-up

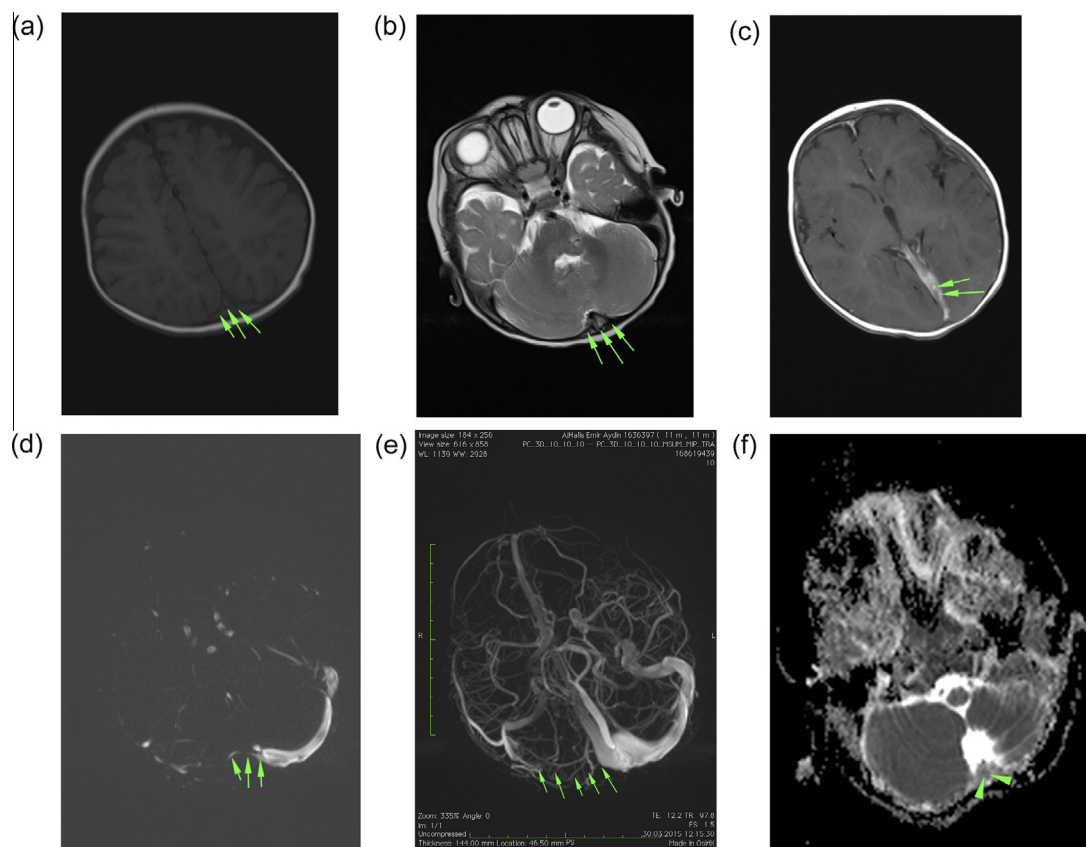
MRI up to sixth months there becomes a slight resolution of the thrombus in which a reduction occurred in the size. The infant was checked for any evidence of supporting underlying cause for thrombophilia, but the protein-C and protein S-dependent anticoagulation system and lupus anticoagulants were within normal limits. Additionally, the child revealed no signs of the fever, dehydration and anemia. Beginning from the neonatal period until six months age the child gets clexane treatment. Unexpectedly, at the sixth month, the conventional MR sequences demonstrated mild dilatation of the third and lateral ventricles and the enlargement of the hemispheric sulcal structures which is a suggestive finding of cerebral atrophy (Fig. 3(a)–(d)).

## 3. Discussion

Thrombosis of the fetal dural sinus is an extremely rarely described entity which was not to be shown more than about fifty cases in the literature (3). Within the usage of fetal MR the diagnosis of the fetal dural venous sinus thrombosis has increased by the meantime. The etiology of the dural sinus thrombosis is not clear and a subject to discuss. The fetomaternal risk factors such as infection, hypercoagulability, trauma and preeclampsia are the most considerable reasons to declare (4). The embryological development of cerebral vascularization is very complex. Arterial vessel organization occurs in the fifth–twelfth day after conception after than up to 20 weeks the central upper and lower cerebral venous system develops. The major venous system development ends but the connection of the other vessels system and inner calibration of the transverse sinus still goes on (5,6). Therefore a weakness in the transverse sinus occurs which could not be able to



**Fig. 1** Thirty years old female patient in second trimester pregnancy demonstrated an echogenic structure in torcular herophili referred to the fetal magnetic resonance imaging of detailed examination (a) axial T1 weighted images Fat saturated images shows hyperintensity in torcular herophili (green arrow). (b) On T2 weighted sagittal HASTE images thrombus is hypointense to gray matter (green arrow). (c) T2 weighted images shows the hypointensity in torcular herophili extending superiorly (green arrow).



**Fig. 2** The one day-old male newborn of cranial magnetic resonance imaging examination shows (a) T1 weighted axial images demonstrates the slightly hyperintense thrombus confluence (green arrow). (b) Axial T2 images show the hypointense–heterogenous appearance of the thrombus at the same anatomic region (arrow). (c) T1 weighted contrast enhanced series shows the filling defect of thrombus in the supracerebellar region (arrow). (d) The phase contrast MR venography shows filling defect in the torcular herophili there is a slightly smooth intensity obtained at the torcular herophili which could represent the regression of the thrombus (arrow). (e) The phase contrast MR venography shows diffuse filling defect in the right sinus transversus (arrow). (f) Additionally there is a mild diffusion restriction in the center of the thrombus at the confluence sinus in ADC images (green arrow).

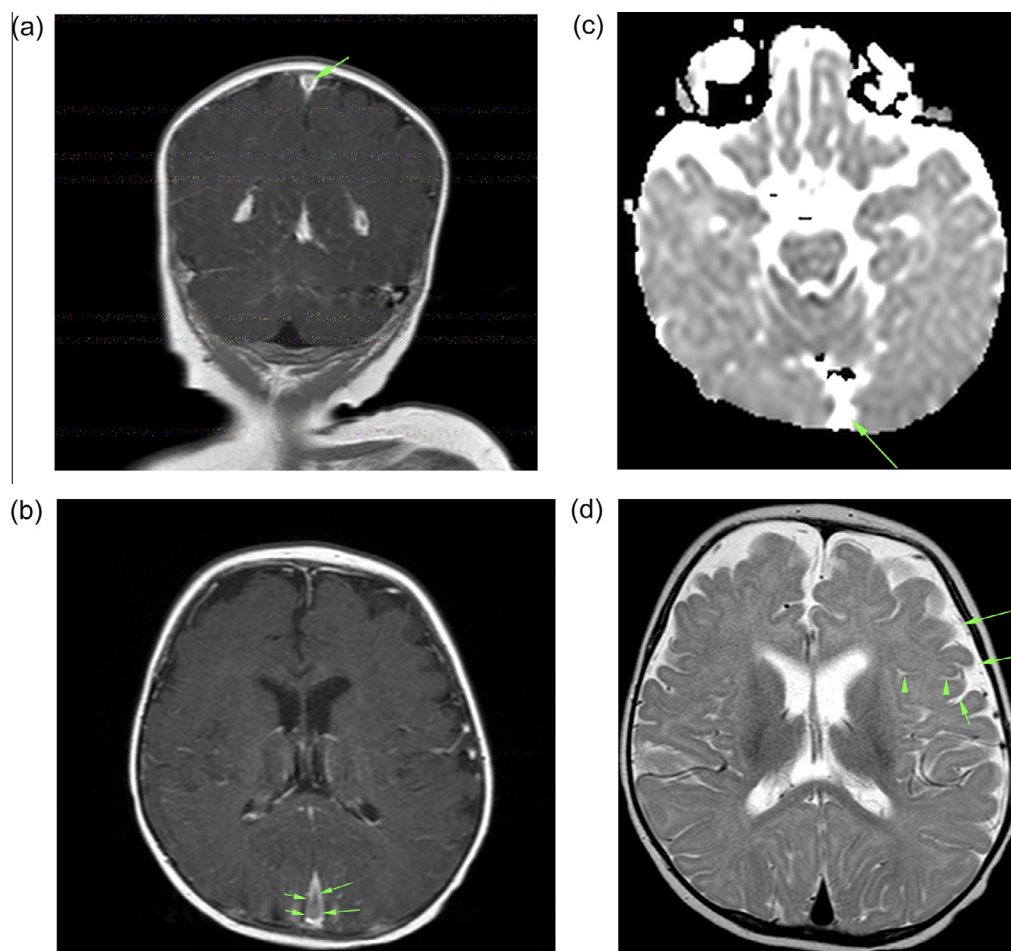
demonstrate a sufficient struggle against the higher pressure. Regarding the dural sinus thrombosis different hypotheses were described by Delmas et al. (6). The main consideration was based upon the acceptance of the dural sinus malformations which is a form of arterio-venous shunts. These shunts composed of very low-flow hemodynamics which causes a ballooning of the transverse sinus within a high pressure occurring in a long-term duration. In the basic knowledge regarding within the embryologic development of the venous structures mainly based on the transverse sinus highest pressure occurs after the 20th week (7). Additionally in the literature including our case, nearly almost all of the cases were detected after 20th gestational age (6,7).

MRI appearance of dural sinus thromboses has a heterogeneous appearance. Main advantage of MRI over Doppler sonography is to diagnose the etiology of the thrombus. It gives a detailed resolution of the posterior fossa and cerebellum, and extension of the dural venous structures (8). The presence of any tumoral structures and its structural components including blood products and the age of the hemorrhage, mass effect and solid components can be easily detected. In our case the dural sinus thrombus is iso-hyperintense on T1 in the antenatal life during the follow-up procedure the signal

intensity was decreased. The T1 hyperintensity shows fresh blood clot which consists of high proteinaceous content and low signal intensity is the hemosiderin in the chronic period. The accompanying intracranial findings of dural sinus venous thrombosis including cerebral and intraventricular hemorrhage, infarction, mass effect on the cerebral and cerebellar structures, prematurity, ventriculomegaly, schizencephaly and leptomeningeal dural vascular malformation have been considered as the bad prognosis. Spontaneous regression and normal biparietal diameter are the favorable outcomes (1,6,8).

In 2014, Rayssiguier discussed the diagnosis of the dural sinus thrombosis and outcomes of eight cases extensively. In all of eight cases MRI imaging were used to evaluate (9). This case series is the largest study regarding fetal dural sinus thrombus in the literature, they mentioned that fetal MRI is essential to diagnose and to confirm. The role of MRI in diagnosis is already discussed by the other authors in the literature. Beginning from 2006 till now almost all of the cases in MRI are a part of confirming the diagnosis (1–3,6–13).

The first step of the diagnosis is made by color Doppler sonography. MRI is usually used to confirm the findings of sonography and to understand the other accompanying abnormalities (1,2,9). However a new diagnostic tool of diffusion



**Fig. 3** The sixth month aged male boy with diagnosed thrombosis at torcular herophili in antenatal life and showed regression in the serial follow-up MRI. The brain MRI shows: (a) T1 coronal post-contrast series shows slightly hypointense filling defect at torcular herophili. (b) T1 axial postcontrast images shows small filling defect at the confluence sinus and regression compared to antenatal life. (c) Axial T2 weighted images of the brain shows the enlargement of the extraaxial CSF spaces at the frontal region. (d) There is also prominence of the sulcal structures in this level which indicates cerebral atrophy.

MRI in the role of detection is limited. On the behalf nearly all the cases in Rayssiguier's study the thrombus is hyperintense on T1 weighted images which reflects the age of thrombosis like our case (9).

There is male predominance in the literature by Delmas et al. (6), however this is not true for Rayssiguier's study (9). In our study the fetus is male.

Throughout the detailed search in literature, there only two cases with hydrocephalus. One of them belongs to Lato et al. (13), which were operated for hydrocephalus and the other is Visentin et al.'s second case who had borderline ventriculomegaly. Both of these cases do not show any signs of cerebral atrophy (14). In our case, there is an unfavorable prognosis between the fourth and sixth month follow-up. The brain volume was decreased, ventriculomegaly in lateral ventricles and slight enlargement in extradural space were occurred which is a predictive signs of cerebral atrophy in the cranial MRI. Exception of the dural venous sinus thrombus the patient was healthy and there was not any predisposition factor of cerebral atrophy. The patient was under treatment of clexane. In literature there is only one publication that explains

an indirect possible relationship of clexane and the cerebral atrophy (15). But this article focuses on the subdural hemorrhage as a side effect of the clexane. To the best of our knowledge, our case is the one in the literature with an unexpected favorable outcome of cerebral atrophy in the follow-up.

#### 4. Conclusion

In conclusion, the sonographic appearance of echogenic mass resembling a huge venous lake in the occipital region suggests an ectasia of torcular herophili thrombosis which needs further delineation of fetal MRI. The long-term follow-up should be performed in the evaluation of the dural sinus venous thrombosis although the thrombosis resolves in the meantime for further unexpected complications.

#### Conflict of interest

The author attributes that there is no conflict of interest.

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