

Torcular dural sinus malformations: prenatal diagnosis by MRI and review of prognostic factors. About a case

Malformaciones de la confluencia de los senos venosos: diagnóstico prenatal por resonancia magnética y revisión de los factores pronósticos. A propósito de un caso

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Summary

Dural venous malformation is a rare fetal cerebrovascular involvement, but represents the most frequently diagnosed cerebral thrombotic event during fetal life. We present the evolution of a case of dural venous malformation with thrombosis through magnetic resonance imaging (MRI) controls from prenatal to postnatal period, which proves its complete spontaneous resolution. It is essential to recognize this entity and its prognostic factors, since it describes the possibility of spontaneous resolution with conservative management, as shown in the case documented here.

Resumen

La malformación venosa dural es una afectación cerebrovascular fetal poco frecuente, pero representa el evento trombótico cerebral más frecuentemente diagnosticado durante la vida fetal. Se presenta la evolución de un caso de malformación venosa dural con trombosis mediante controles con resonancia magnética (RM) desde el periodo prenatal hasta posnatal, que comprueba la completa resolución espontánea de la misma. Es fundamental el reconocimiento de esta entidad y de sus factores pronósticos, ya que está descrita la posibilidad de resolución espontánea con un manejo conservador, como en el caso aquí documentado.

Introduction

Dural sinus malformations (DSM) are rare congenital anomalies, accounting for less than 2% of all intracranial vascular malformations (1).

In the neurosurgical literature, DSMs are associated with low-flow arteriovenous fistulas (AVFs), and historically have been reported to have a poor outcome (2). However, recent reviews yield more encouraging data, and show fetal magnetic resonance imaging (MRI) as a valuable tool for diagnostic confirmation, prognosis, and planning of therapeutic strategies (3).

Clinical Case

A 34-year-old woman at 24 weeks of gestation with a fetal ultrasound diagnosis of thrombosed DSM with very poor prognosis, performed in another hospital. She came to our center seeking a second opinion and to perform complementary studies with fetal MRI (Figure 1).

Due to the findings, follow-up with fetal MRI at 27 weeks + 2 days of gestation is recommended (Figure 2).

Given the decrease in the size of the DSM and increase in the size of the thrombus, successive controls at 32 weeks and 2 days, and 36 weeks and 2 days of gestation were advised. Likewise, postnatal controls were performed at 6 months, at 1 year and 6 months and at 3 years and 3 months of life, when the patient was asymptomatic (Figure 3).

Discussion

DSM is a rare congenital malformation that belongs to the group of dural arteriovenous fistulas and represents less than 2% of congenital intracranial vascular anomalies (1).

There are several hypotheses about the occurrence of dural sinus malformation. One suggests that dural malformation is due to persistent ballooning of the sinuses between the fourth and sixth month of gestation,

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which causes secondary venous hypertension and arteriovenous fistulas (2). Another theory considers that dural sinus malformation is the result of excessive and disorganized development of the dural sinuses, even in the period when the sinuses should decrease in size (2). As for thrombosis, the following theories are put forward to explain it: the result of sinus immaturity, altered blood flow and modification of the endothelial lining of the sinus (2).

Prenatal diagnosis is possible and ultrasound is the initial imaging modality of choice. The typical ultrasound finding of DSM includes a huge anechoic cystic structure in the posterior fossa attached to the dura. On color Doppler, vascularization has been described at the lateral margins of the lesion with no detectable flow in the center. It is postulated that the marginal flow represents multiple mural arteriovenous shunts generally associated with the lesion (3).

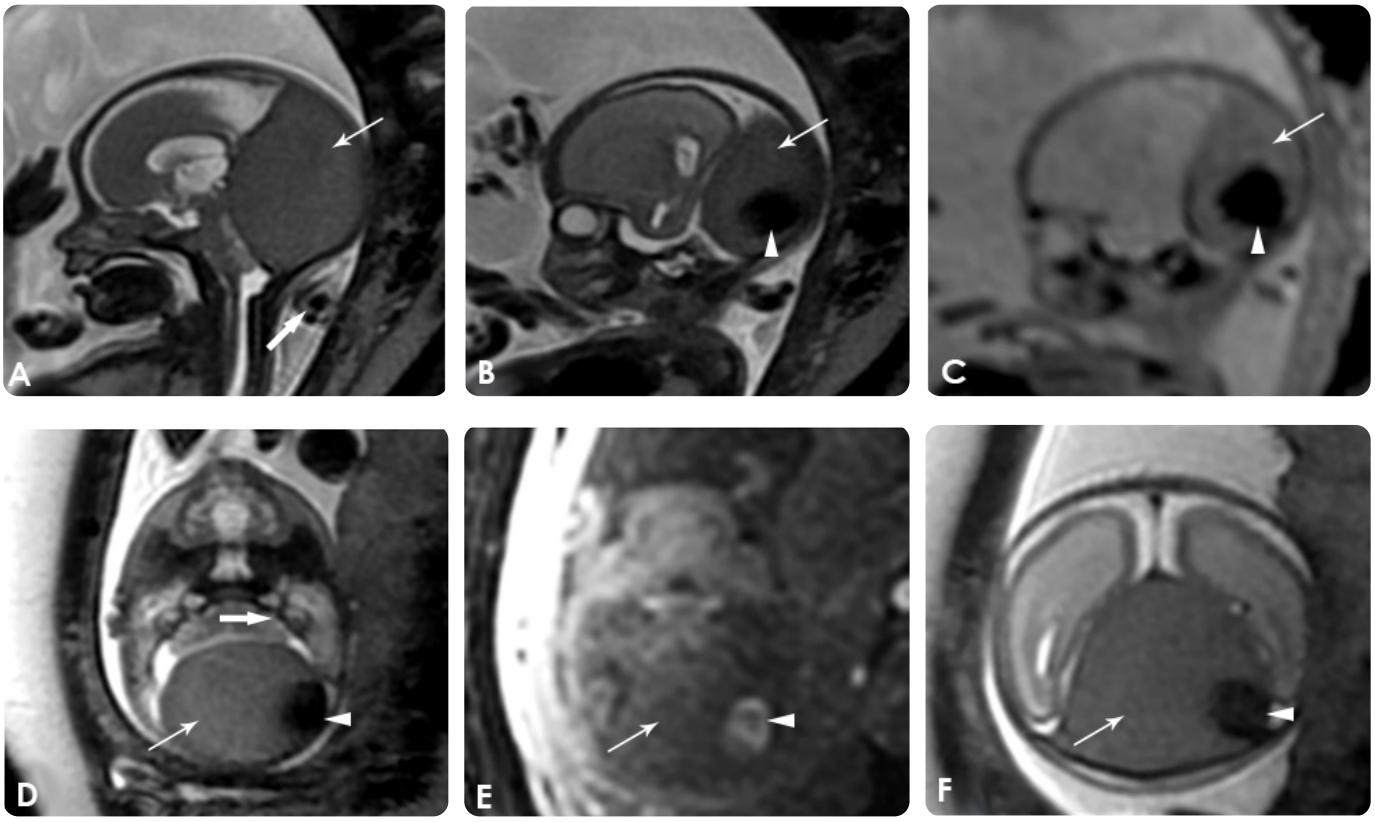


Figure 1. MRI. Dural venous malformation with subacute thrombus. Gestational age: 24 weeks. a and b) Sagittal SSFSE T2; c) Sagittal gradient echo T2*; d) Axial SSFSE T2; e) Axial 3D gradient T1 (LAVA), and f) Coronal SSFSE T2. Dural venous malformation with large cystic dilatation of the confluence of the venous sinuses, as well as the posterior third of the superior longitudinal sinus (thin white arrow) that exerts significant mass effect on the occipital and parietal lobes, occipital horns of the lateral ventricles, ventricular atria, posterior margin of the third ventricle, splenium of the corpus callosum, quadrigeminal tubercles and on the cerebellar hemispheres and cerebellar vermis (thick white arrow). Subacute thrombus in its left lateral margin and in its lower third (arrowhead), of high signal in sequences with T1 information and low signal in sequences with T2 information.

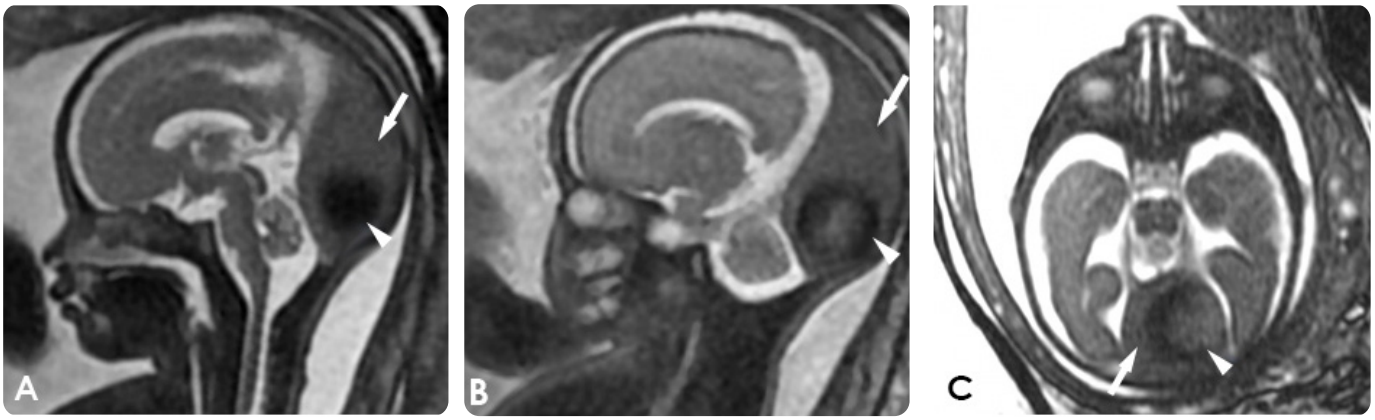


Figure 2. Control MRI at gestational age of 27 weeks and 2 days. a and b) Sagittal SSFSE T2, and c) Axial SSFSE T2. Decrease in DSM (white arrow) and increase in thrombus size (arrowhead), with improvement in the mass effect on adjacent structures.

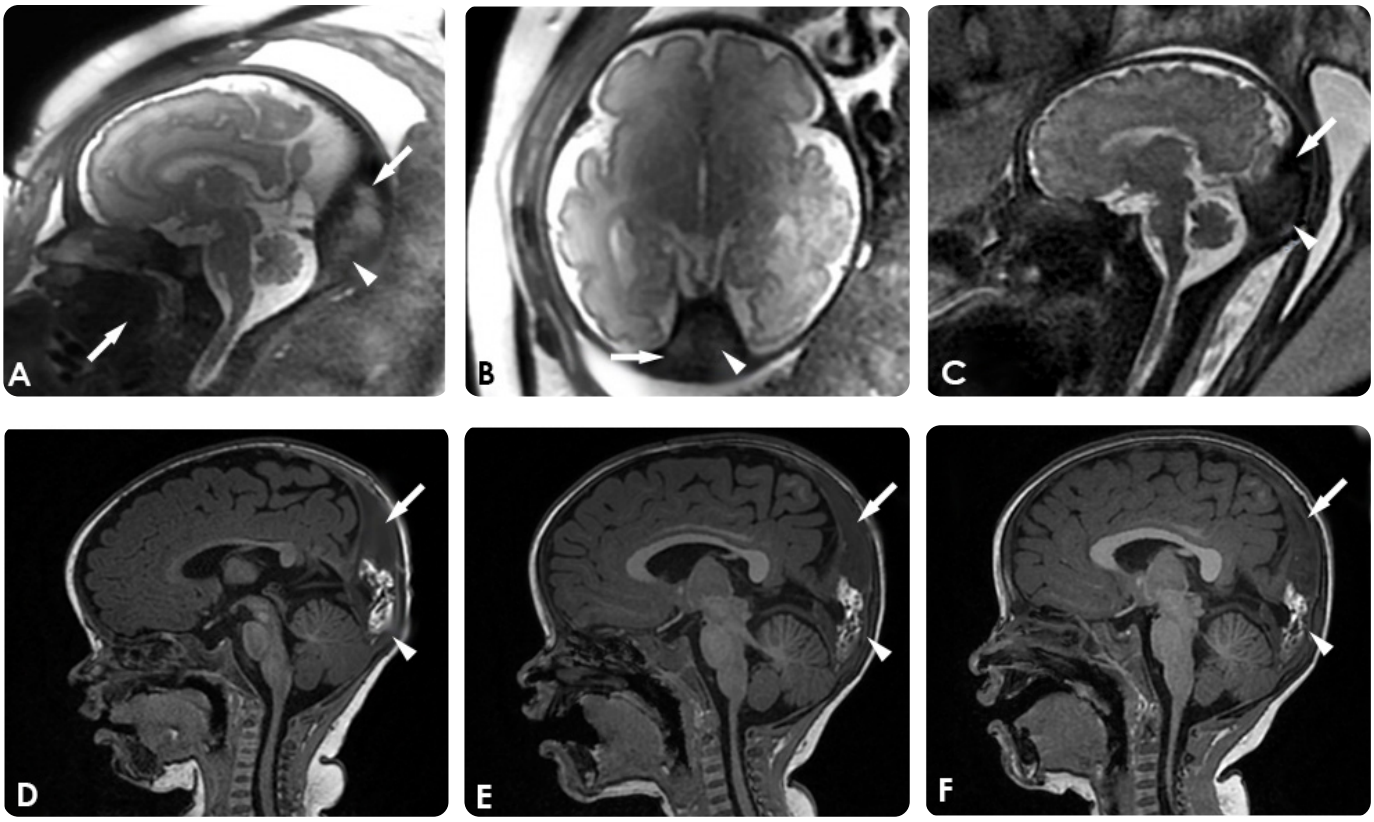


Figure 3. Control MRI. a and b) Gestational age of 32 weeks. Sagittal and axial SSFSE T2. c) Gestational age of 36 weeks. Sagittal SS FSE T2. d) At 6 months after birth. Sagittal 3D Gradient Echo T1. e) At 1 year and 6 months of age. Sagittal 3D Echo of gradient T1. f) At 3 years and 3 months. Sagittal 3D Echo of gradient T1. In successive intrauterine controls of the DSM located at the confluence of the venous sinuses with thrombosis, it was observed how the thrombus increased in size (arrowhead) and the DSM decreased in size (white arrows) until the thrombus occupied the entire sinus, indicating a good prognosis. After birth, the DSM and thrombus did not change in size.

Fetal MRI is essential to characterize and confirm the diagnosis of DSM and also to identify any intracranial complications. DSM shows a low signal on T2-weighted sequences. However, if thrombosis is present, the signal may be intermediate-high on T1-weighted sequences and intermediate-low on T2-weighted sequences, depending on the time course of the thrombus. The resulting mass effect on the cerebellum, fourth ventricle and associated hydrocephalus has also been described in some cases (4).

Diffusion sequences help in prognosis by demonstrating intralesional thrombosis and excluding potentially devastating intracerebral ischemic-hemorrhagic lesions (5).

Differential diagnosis should be made with the most common lesions of the posterior cranial fossa of the fetus, such as Dandy-Walker malformation, retrocerebellar arachnoid cysts, vascular malformations (aneurysmal malformation of the vein of Galen) and congenital tumors, especially teratomas, easily differentiated by MRI (6).

Most DSM seem to occlude spontaneously with the consequent remodeling of venous flow and favorable results (7).

Clinically progressive lesions will require endovascular intervention to stabilize them. Decrease in size during fetal life is the most important prognostic factor, while the presence of ventriculomegaly, parenchymal hemorrhage, and identifiable arterial feeders are indicators of poor prognosis (8). Initial DSM growth is not necessarily a harbinger of poor outcome, as there are cases in which transient growth was observed before involution (8).

In a recent DSM study of venous sinus confluence, 97% of prenatal cases had spontaneous shrinkage with 83% survival (62% with favorable clinical course). Postnatal cases had a 59% survival (29% with favorable clinical course) (8).

A DSM found in fetal life requires, therefore, a short-term follow-up to assess the decrease in size of the lesion or to detect early findings that imply a poor prognosis in the neurological development of the fetus.

Conclusion

DSM is a rare congenital anomaly, the initial diagnosis of which can be made by prenatal ultrasound. Fetal MRI should be performed to establish the definitive diagnosis and to exclude other posterior fossa pathologies. Fetal MRI provides useful information for perinatal management and prognosis, since it assesses better than ultrasound the involvement of the venous sinuses and the mass effect on adjacent structures. Additionally, it rules out the presence of hydrocephalus or ischemic lesions, and allows calculating the percentage of thrombosed dural venous malformation, which are prognostic factors.

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