SWISS SOCIETY OF NEONATOLOGY

Dural sinus malformation

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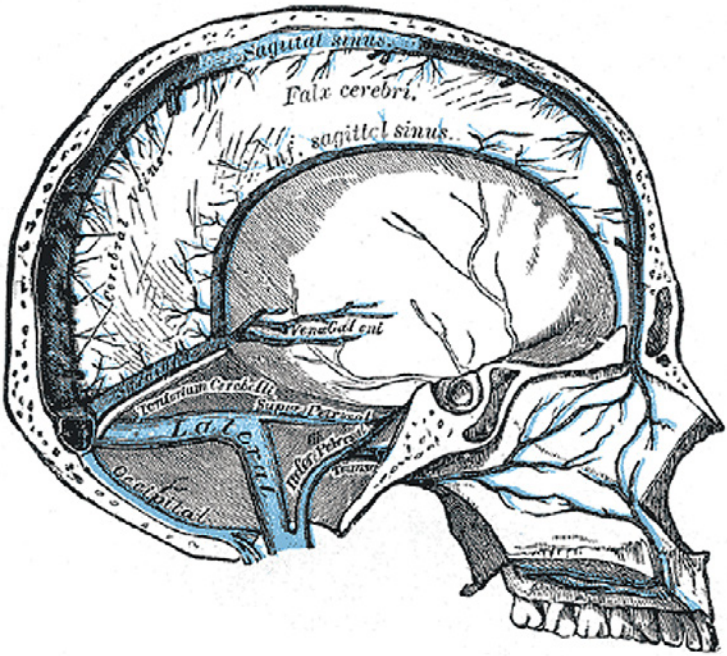

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Title figure:
Sagittal section of the skull, showing the sinuses of the dura (source: Wikipedia).

INTRODUCTION

Dural sinus malformation (DSM) is a rare congenital condition characterized by congenital massive enlargement of one of the dural venous sinuses. Historically, the disease has been mainly reported to have a poor prognosis, with a few recent reports describing a more favorable outcome.

We report the case of a preterm newborn with a dural sinus malformation with thrombosis, who presented with persistent thrombocytopenia as the only symptom.

This male preterm neonate, the first child of a non-consanguineous couple, was born at 34 0/7 weeks of gestation by spontaneous vaginal delivery. He had a birth weight of 2075 g (P 20) and a head circumference of 29 cm (P 4). The Apgar scores were 6, 6, and 6 at 1, 5, and 10 minutes of age, respectively.

Pregnancy had been uneventful and there were no relevant maternal and family histories, except for the mother being hepatitis B surface antigen (HBsAg) positive.

At birth, the infant had signs of respiratory distress for which CPAP was initiated and continued up to the 9th hour of life. The patient also had an episode of mild hypoglycemia with a blood glucose concentration of 2.2 mmol/l, which was successfully treated with an intravenous glucose bolus. In view of the maternal HBsAg positivity, the patient was actively and passively vaccinated against hepatitis B within the first 12 hours of life in accordance with local guidelines.

On the fourth day of life, the patient developed hyperbilirubinemia, which required a 24-hour-course of phototherapy. In this context, a full blood count was performed and showed moderate thrombocytopenia with a platelet count of 86 G/l.

To further investigate the cause of thrombocytopenia, an abdominal ultrasound was done, and it excluded

thrombosis of renal vessels. A cerebral ultrasound showed an anechogenic mass in the posterior fossa with no signs of hydrocephalus. Magnetic resonance imaging (MRI) confirmed the presence of an extra-axial mass in the posterior cranial fossa consistent with a thrombosed dural sinus malformation (DSM) (Fig. 1). The thrombus was localized in the confluence of sinuses (torcular Herophili, the connecting point of the superior sagittal sinus, straight sinus, and occipital sinus) as well as in multiple adjacent sinovenous structures. Deep cerebral veins such as the basal veins of Rosenthal, the internal cerebral veins and a left mesencephalic vein were enlarged, and focal mesencephalic signal abnormality due to congestion/edema was demonstrated (Fig. 2A). Mass effect with anterior dislocation of the cerebellum and the brainstem was present.

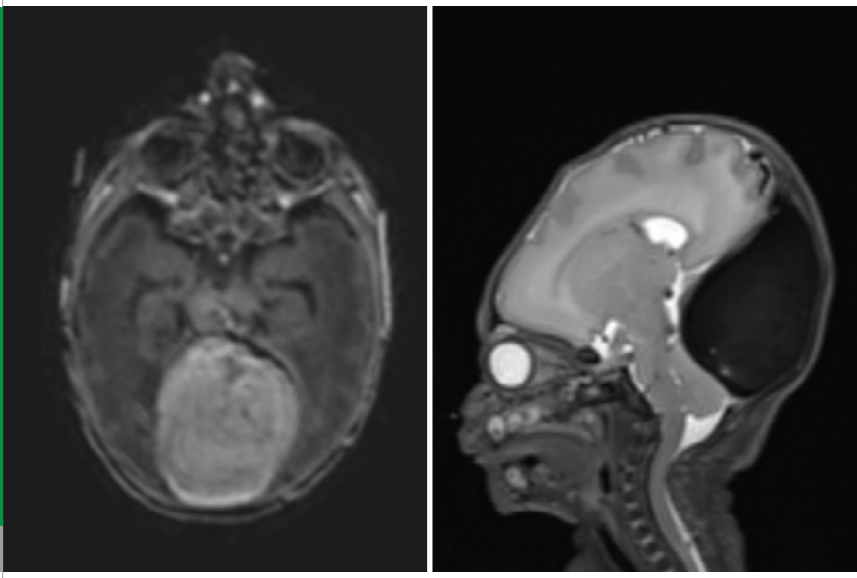

Fig. 1

Initial MRI demonstrating a large extraaxial mass in the posterior cranial fossa.

In view of the radiological findings and the rarity of the disease, the patient was transferred to the University Hospital of Bern for further assessment and treatment. MRI with MR angiography confirmed the above-described findings and showed no evidence of an associated arteriovenous shunt. The time at which the thrombus had formed could not be defined.

Full heparinization was started on the 8th day of life. Regression of the focal mesencephalic congestion/edema could already be demonstrated on a follow-up MRI 10 days after initiation of anticoagulation therapy (Fig. 2B). Additional MRIs (Fig. 3) showed progressive and almost complete recanalization of the sinovenous structures, as well as steady regression of the venous dilatation. Anticoagulation therapy with enoxaparin is still ongoing. Clinically, the patient remains fully asymptomatic at the age of almost two years.

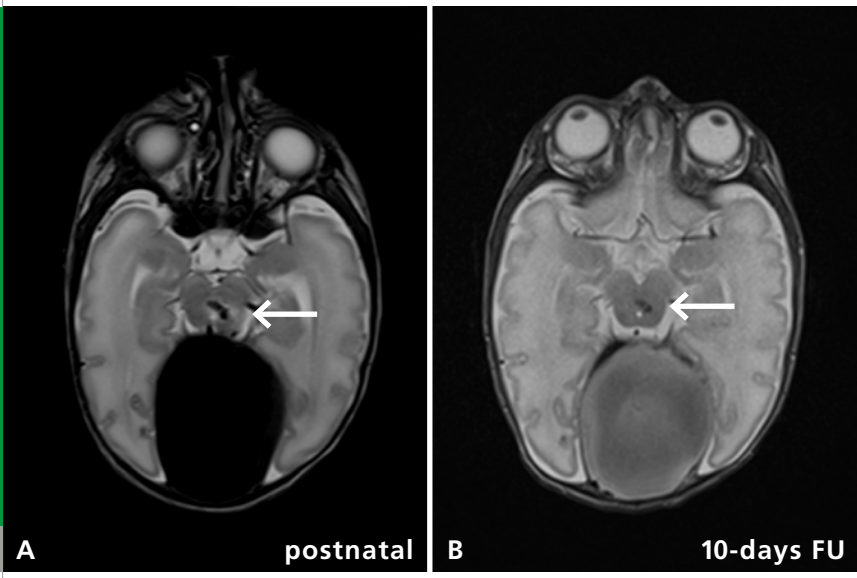

Fig. 2

A) initial mesencephalic venous congestion edema (arrow) adjacent to a dilated mesencephalic vein; B) edema regression in the short term follow-up imaging study (under heparinization).

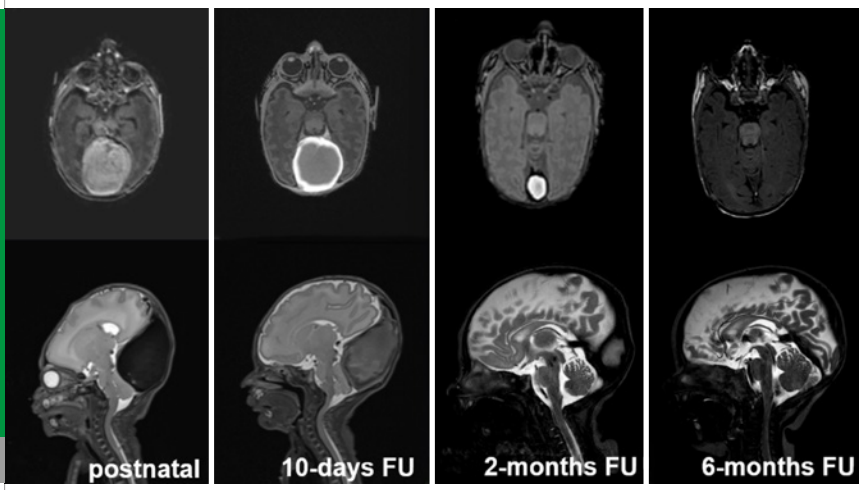

Fig. 3

Steady regression of venous dilatation and progressive thrombus organization between the initial MRIs (A) and the follow-up MRIs (B – C).

DISCUSSION

DSM is a congenital condition characterized by massive enlargement of one of the dural venous sinuses. The condition is rare and represents less than 2% of miscellaneous pediatric vascular malformations (1). In some cases, but not invariably, thrombosis of the involved sinus may be present (2).

The pathogenesis of DSMs is unclear, and different mechanisms have been considered. First, they could result from the prolonged persistence of the "sinus ballooning", which occurs physiologically in the second and third trimesters of pregnancy. Another hypothesis suggests the possibility of abnormal growth of the sinus due to a local disturbing trigger (1, 3).

DSMs can be classified into two groups according to Lasjaunias et al. (4): 1) DSMs with giant pouches or lakes and mural AV shunting involving the adjacent posterior sinuses and 2) DSMs of the jugular bulb associated with a petromastoid-sigmoid sinus high-flow arteriovenous fistula (AVF), the latter being characterized by a good prognosis (1, 3).

The clinical manifestations of this condition are diverse. Among these are macrocrania, seizures, psychomotor delay, mild cardiac failure, coagulation disorders due to consumption syndromes (such as seen in our patient), and moderately increased intracranial pressure (3).

Thrombosis of the involved sinus is a common evolution of the lesion. In contrast with sinus thrombosis not associated with DSM, which normally develops following a trigger such as ventricular hemorrhage, meningitis, shock, or dehydration, thrombosis in DSMs occurs spontaneously (1).

In patients with DSMs, several complications have been described, such as global parenchymal brain volume loss and intracranial hemorrhage (4) as well as hydrocephalus and brain infarction in patients presenting with bilateral venous drainage even though the lesions have favorable evolution (1, 3).

The prognosis of patients with DSMs involving the posterior sinuses has classically been described to be poor. However, some recent case reports and reviews have highlighted more positive outcomes (1, 4). The presence of feeders and brain damage (5) affect prognosis.

The main goal of therapy consists of securing adequate blood drainage. The treatment largely depends on the individual cases; however, it mostly comprises of heparin to treat the thrombosis or embolization in case of visible shunts in order to improve the hydrovenous equilibrium of the maturing brain (3).

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