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Fetal subdural hematoma, sickle cell disease and storage pool disease: A case report

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ABSTRACT

A fetal subdural hematoma (SDH) was diagnosed in a patient with sickle cell disease (SCD) during a routine ultrasound exam in the 30th week of pregnancy. A scan performed a few days earlier had revealed no abnormalities. After interdisciplinary consultation with neurosurgeons and neonatologists, a cesarean section was performed since acute subdural bleeding was hypothesized and the mother's condition was critical. After surgery, the diagnostic procedures revealed that the child and the mother had also suffered from thrombocytopathy, which probably jointly contributed to causing the bleeding; in general, anemia and hypoxia may also play an important role. The newborn had a good neurological outcome.

Ultrasound features do not reflect the underlying cause and therefore predicting the prognosis is challenging. In most cases, prenatal counseling is difficult because of the unknown underlying cause and because there are no ultrasound or magnetic resonance imaging criteria to define which cases can benefit from delivery or expectant management. Where there is acute bleeding, the child could benefit from delivery and surgical evacuation of the hematoma. Further investigation to identify the cause of the bleeding can improve management and prognosis.

1. Background

The estimated incidence of fetal intracranial hemorrhage (ICH) is 1:10000 [1]. Intracranial haemorrhages can be intracerebral or extracerebral. Subdural hematomas (SDH) are extracerebral bleedings located between the dura and arachnoid mater [2].

Prenatal diagnosis of a subdural hematoma is uncommon, especially in the absence of previous traumas. A trauma in a pregnant woman provides the occasion for a detailed search for fetal and maternal bleeding.

Fetal coagulation disorders, including alloimmune thrombocytopenia, and maternal use of anticoagulants that cross the placenta are common reasons for fetal intracranial hemorrhage [2,3]. Hypoxic damage (as well as fetal anemia) has also been described as a cause [1]. However, in 47% of the cases, the exact cause cannot be identified [1,2].

1.1. Subdural Hematoma

The 30th week is the average gestational age at the time of diagnosis [4]. At this time the fetal head gets closer to the wall of the uterus and to the maternal abdomen and therefore can be easily affected by minor direct trauma. In cases of additional risk factors for fetal bleeding, the probability of SDH is higher.

SDHs are probably the most difficult entity to recognize in utero, and differentiation from other intracranial masses, such as tumors, may be difficult. A large SDH will typically appear as a complex mass compressing and distorting the hemispheres [5].

A recent systematic review classified the ultrasonography features of fetal SDH [2]. The most frequent features were intracranial echogenicity (42,2%), lateral ventriculomegaly (37,8%), presence of an intracranial mass (31,1%), macrocephaly (24%), midline deviation of the cerebral falx (20%), intracranial fluid collection (11,1%) and reverse diastolic flow in the middle cerebral artery (11,1%). 32% of the affected fetuses died either due to termination of pregnancy or stillbirth/infant death [2]. Among survivors, the neurological outcome was reported to be abnormal in 42% of cases. The prognosis is better when the cause of the bleeding is identified [2].

Magnetic resonance imaging (MRI) should be performed where antenatal ultrasonography is inconclusive [1]. Fetal MRI can aid both in the diagnosis and in the counseling, because of its accuracy in diagnosing cranial anomaly in the third trimester, when the fetus is often in a cephalic presentation and cranial ossification is almost complete [4].

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1.2. Sickle Cell Disease

Sickle cell disease (SCD) is one of the most common forms of inherited hemoglobinopathy. It is caused by a homozygous mutation [6] and manifests as chronic anemia accompanied by painful vaso-occlusive crisis [7]. Individuals with SCD are more susceptible to stroke and serious bacterial infections [8]. Both the SCD mother and baby are at increased risk of adverse events during pregnancy [8].

1.3. Platelet Dense Granule Storage Pool Deficiency

Platelet dense granule storage pool deficiency (δ-SPD) is a platelet aggregation disorder associated with a vulnerability to mild to moderate bleeding. The defect may be inherited in isolation or as part of other congenital syndromes such as oculocutaneous albinism and Hermansky–Pudlak syndrome. The pattern of inheritance is variable, but is predominantly autosomal dominant [9].

2. Case Description

2.1. Clinical Presentation and Patient History

A 24-year-old woman, gravida 3 para 1, known to have sickle cell disease (SCD), presented in painful crisis to the emergency department at 23 weeks of gestation. Her first child had been born 2 years previously. A cesarean section had been performed at that time because of acute painful crisis in the 38th week of pregnancy. The father of the child was also a carrier of the sickle cell trait. No further personal or family history of bleeding disorders was known. She had a splenectomy 10 years previously. At the time of admission, her hemoglobin concentration was 4 mg/dl. Urinary tract infection was diagnosed. Even after administration of erythrocyte concentrates and antibiotic therapy, the clinical situation was fluctuating, with relapses of the painful crisis. Because of this unstable situation, antenatal corticosteroid therapy for fetal lung maturation was administrated in the 25th week.

2.2. Imaging

Routine fetal ultrasound scans were performed at least once a week, and at the 29th week of gestation a right-sided SDH was suspected with a midline shift and ventriculo enlargement of the lateral ventricles (ventricular width at the atrium of 12 mm).

The right hemispheric SDH was 15 mm × 70 mm and predominantly hypoechoic, but with a hyperechogenic region 14 mm × 33 mm (Fig. 1). The peak systolic velocity of the left middle cerebral artery was increased, at 80 cm/s. The amniotic fluid volume and the umbilical artery Doppler were normal. The estimated fetal weight was in the normal range.

2.3. Management

The situation was discussed with neurosurgeons and neonatologists. The significantly increased risks of stillbirth, neonatal death and severe neurological disability, balanced against the consequences of prematurity, were discussed with the patient. It was unanimously decided to perform an urgent cesarean section. The operation was performed without complications.

A boy was born (1600 g, 70th percentile; Apgar scores at one, five and ten minutes were 7, 7, 8, respectively). The fetal hemoglobin concentration was 11,8 g/dl (normal range 17,7-26,5), and the platelet count was 273/nl (normal range 160–320).

Postnatal transcranial ultrasound confirmed the results of the intrauterine ultrasound; neurosurgical evacuation of the hematoma was therefore performed, with the patient under general anesthesia.

2.4. Outcome and Follow-up

Regular ultrasound scans and MRI were carried out after surgery. The imaging showed no residual findings at the time of discharge two months later. The patient had no neurological deficits.

The mother had a complicated course with recurrent bleeding, multiple surgical revisions and finally hysterectomy due to uncontrolled abdominal bleeding. She was discharged 60 days after surgery.

Further diagnostic work-up revealed SCD in the child, delta storage pool disease in the mother and child as well as a maternal factor X deficiency.

3. Discussion

We present a case of a prenatally diagnosed SDH and its management. The diagnosis of this entity is reported to be rare and difficult [5]. The diagnosis was made during a routine ultrasound examination of a hospitalized patient because of pregnancy complications related to SCD. In our case, the majority of the ultrasonography features could be demonstrated: ventriculomegaly, intracranial mass, intracranial echogenicity, midline shift, intracranial fluid collection. We did not employ MRI at this time since the ultrasound findings were clear.

An acute bleeding is often echogenic on ultrasonography, whereas chronic SDH appears more translucent [2]. When we diagnosed the fetal SDH, the mixed echogenicity revealed that it was in the transitional stage due to the gradual breakdown of hemoglobin and liquefaction of the SDH, or an acute bleed was combining with the chronic SDH.

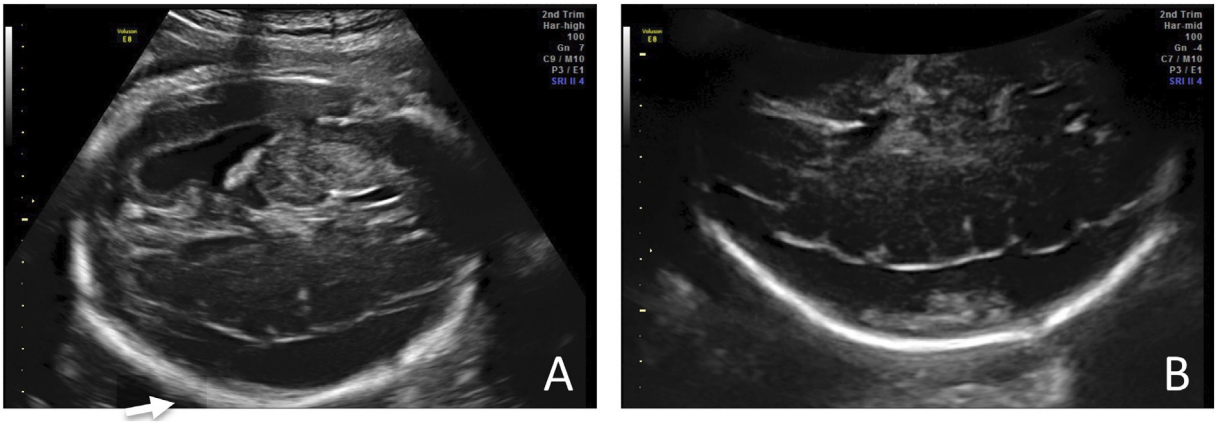

Fig. 1. (A) Transverse scan of the fetal head with midline deviation of the cerebral falx and ventriculomegaly; arrow points to intracranial fluid collection. (B) Magnification of the predominantly hypoechoic hematoma with a hyperechogenic region.

Since corticosteroids for lung maturation were already being administered, we decided to perform urgent delivery. So far there are no data regarding the optimal timing and mode of delivery. In our case, a cesarean section was performed to possibly reduce the risk of further fetal hemorrhage and to allow the neurosurgeon to rapidly evacuate the hematoma. Furthermore, the maternal condition had been critical for several weeks.

The fetus had a high chance of SCD since the mother had it, the father was a sickle cell trait carrier and because of the high peak systolic velocity of the middle cerebral artery. Fetal anemia is reported as another possible cause of antenatal hemorrhage [1]. It has been speculated that hyperdynamic circulation may cause disruption of intracranial vessels. Further coagulation disorders and vasculopathies are well known in patients with SCD, where a high rate of ischemic as well as hemorrhagic strokes is usually observed after birth [10].

Pregnancy in patients with SCDis associated with increased pain, infections, pulmonary complications and thromboembolic events [11]. A meta-analysis in 2015 (including 21 studies worldwide and 26,349 pregnant women with SCD) found an increased maternal mortality (relative risk RR 5·98, 95% CI 1·94–18·44) and an increased risk of stillbirth (RR 3·94, 2·60–5·96) [12].

The examination of all maternal deaths in France between 1996 and 2009 revealed that SCD is responsible for an increase in maternal mortality, particularly postpartum, mainly due to direct complications of the disease [13].

The diagnostic workup in the present case also demonstrated delta storage pool disease (δ-SPD) in the mother and the child. In a pregnant patient with δ-SPD, hemorrhage during surgery or labor should be anticipated. The prophylactic administration of single donor platelets and desmopressin before surgery or labor is potentially lifesaving [14]. In a case report from 2013, a woman affected by storage pool disease and with a history of severe allergic reactions to the conventional treatments was successfully treated with rFVIIa (Novoseven) [9].

Factor X deficiency (FXD) is a rare autosomal recessive bleeding disorder with variable phenotypic severity. A recent systematic review [15] of the gynecological and obstetric complications of FXD showed that these women are at increased risk of heavy menstrual bleeding and ovulation bleeding as well as adverse pregnancy outcome and postpartum hemorrhage (22% of deliveries).

4. Conclusion

SCD, δ-SPD and FXD jointly and decisively contributed to the bleeding complications of the mother and child in the case reported here. To our knowledge, this is the first case report of this kind of thrombocytopathy and SCD involved in the etiology of fetal SDH.

Contributors

Antonella Iannaccone was involved in patient care, co-authored the manuscript, and contributed to critical revision of the manuscript.

Marvin Darkwah Oppong was involved in patient care, co-authored the manuscript, and contributed to critical revision of the manuscript.

Philipp Dammann was involved in patient care and critical revision of the manuscript.

Rainer Kimmig was involved in patient care and critical revision of the manuscript.

Angela Köninger was involved in patient care, co-authored the manuscript, and contributed to critical revision of the manuscript.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Provenance and Peer Review

This case report was peer reviewed.

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