Preterm Foetal Compromise in Dichorionic Twins Due to Concordant Massive Perivillous Fibrin Deposition

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Background

Massive perivillous fibrin deposition (MPFD) is a rare placental lesion affecting <1% of pregnancies. It is associated with adverse perinatal outcomes including recurrent miscarriage, severe intrauterine growth restriction (IUGR) and foetal demise[1]. It also increases risk of neonatal morbidity including for intraventricular haemorrhage, sepsis and necrotising enterocolitis[2]. Its aetiology is unclear although it is associated with maternal coagulopathy[3], autoimmune conditions[4] and imbalance of angiogenic/antiangiogenic factors (specifically, placental growth factor (PlGF), soluble endoglin (sEng), and soluble vascular endothelial growth factor receptor (sVEGFR)[5].

Aims

To increase awareness of MPFD as a rare cause of placental insufficiency leading to adverse pregnancy outcomes.

Case

A 28-year-old G2P1 at 33+0 weeks' gestation with dichorionic diamniotic twins was referred by radiology with poor interval growth of both twins and absent umbilical artery end diastolic flow in twin 2; growth and Doppler flow studies had been normal at 31 weeks' gestation. Cardiotocography for twin 2 was abnormal on admission, with reduced variability, no accelerations and two shallow decelerations over a one-hour period. After discussion with Maternal Foetal Medicine, the woman underwent a caesarean section within the hour. The operation was uncomplicated. The placentas appeared markedly abnormal containing diffuse macroscopic thrombi. Twin 1 had a birth weight of 1710 g, APGARs of 8 and 9 at 1 and 5 minutes, respectively, and normal arterial cord blood analysis. Twin 2 had a birth weight of 1690 g and APGARs of 7 and 9. Arterial cord blood analysis had evidence of hypoxia and metabolic acidosis: pH 7.036, lactate 10.6, base excess -14. Twin 2's neonatal course was complicated by hyperinsulinaemic hypoglycaemia, anaemia and bilious vomiting.

Results

Placental histopathology showed massive deposition of fibrinoid entrapping avascular but otherwise viable chorionic villi, with the depositions affecting both placental discs and making up approximately 50% of the placental volume – consistent with MPFD *(Figures 1 and 2)*. Microscopically, there were small clusters of histiocytes within the fibrinoid, but without villitis or intervillositis. Of interest, a degree of reactive erythroblastosis was present in the vessels corresponding to twin 2, consistent with the abnormal Doppler studies. The decidua was unremarkable with maternal vessels showing appropriate adaptation. Maternal thrombophilia screen was negative.

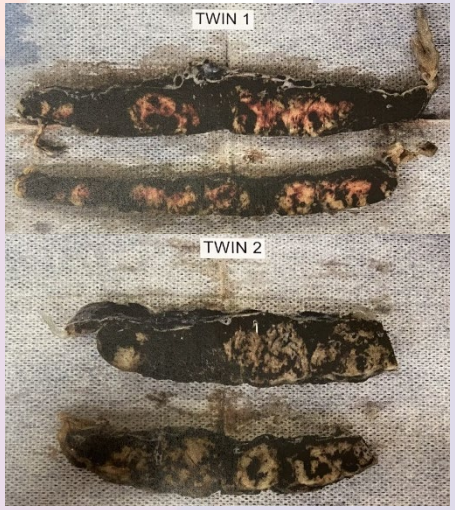

Figure 1. Macroscopic images of fibrin deposition (white) in placentas of each twin.

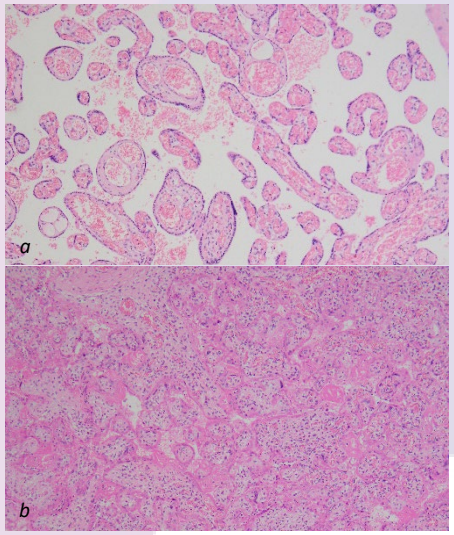

Figure 2. Microscopic images of a) normal villi and b) massive perivillous fibrin deposition.

Discussion

The diagnosis of MPFD is of clinical significance as it carries a stillbirth rate of 13-50% and there is a high recurrence rate in subsequent pregnancies of up to 78%[6]. It is also associated with foetal neurologic injury, and infants should have close paediatric follow-up. MPFD should be suspected in a history of recurrent pregnancy losses and severe, usually early-onset, IUGR. In this case, IUGR was detected on routine surveillance for twins. There is no known association between multiple pregnancy and MPFD. Interestingly, there exist a handful of case reports on MPFD discordance in twin placentas[7-9] but no case of concordance, as occurred here, has yet been described.

There is no established prevention or treatment for MPFD. However, therapies that have been successful are thrombolytics (aspirin, heparin)[10], immunotherapy (IVIg)[11], and, a statin (pravastatin) to correct angiogenic/antiangiogenic imbalance[12]. It is worth studying these further to prevent the potentially devastating effects of MPFD.

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