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CASE REPORT

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Massive perivillous fibrin deposition and single-artery umbilical cord: a case report and review of the literature

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ABSTRACT

Massive perivillous fibrin deposition (MPFD) is a rare but serious complication of pregnancy that can lead to adverse outcomes for both the mother and the fetus. The incidence of MPFD is not well established, but it is generally considered a rare complication. MPFD can be difficult to diagnose due to the similarity of its clinical signs and symptoms to those of other obstetrical complications. Diagnosis is often confirmed by histological examination of the placenta after delivery. Treatment options depend on the severity of the condition, and may include blood transfusion, coagulation products, corticosteroids, and emergency delivery. Obstetricians should be aware of the clinical signs and symptoms of MPFD and consider it in the differential diagnosis of pregnant women with similar presentations. Further studies are needed to obtain more accurate estimates of its incidence and to develop better management strategies for this rare condition. We present a case report of a patient admitted to our service for decreased fetal movements in the context of growth restriction, in whom a diagnosis of MPFD and single-artery umbilical cord was established.

ARTICLE HISTORY

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1. INTRODUCTION

Massive perivillous fibrin deposition (MPFD) is a rare but severe condition of pregnancy that can have significant consequences for both the mother and the fetus [1]. MPFD is characterized by massive fibrin deposits in the intervillous spaces of the placenta, which can lead to obstruction of placental vessels and rapid deterioration of the mother's and fetus's conditions [2]. The exact cause of MPFD is unknown, but it is thought to be related to abnormalities in the coagulation system or immune system. The condition can lead to poor fetal growth, preterm labor, stillbirth, and maternal hemorrhage. Due to the rarity of MPFD, there is limited research on the condition, and further studies are needed to better understand its pathophysiology, risk factors, and optimal management strategies. In the meantime, early diagnosis and prompt delivery may be the best course of action to minimize the risk of adverse outcomes. Monitoring for MPFD may be especially important in high-risk pregnancies, such as those involving women with a history of thrombophilia, autoimmune disorders, or recurrent pregnancy loss [1]. Here, we present the case of a patient with decreased fetal movements and growth restriction, and upon admission to our service, the diagnosis of MPFD was established.

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2. CASE PRESENTATION

A 24-year-old primiparous woman was admitted to our facility at 34 weeks of gestation due to decreased fetal movements and uterine bleeding that occurred 6 hours before admission. The patient had no notable medical history, including no hypertension, diabetes, autoimmune disease, or thrombophilia, and there was no genetic pathology in her family. During prenatal follow-up, the patient had normal blood pressure and laboratory results, including no anemia and negative serologies for hepatitis B and C, HIV, VDRL, and TPHA. An ultrasound performed at 30 weeks of gestation revealed fetal weight below normal for gestational age, but the results of umbilical and cerebral dopplers were normal. Upon admission, the patient had a blood pressure of 123/67 mmHg, no perceivable fetal heart sounds, and blackish bleeding stigmata were observed during the speculum examination. The patient had 4 contractions every 10 minutes with good interphase relaxation, and the cervix was dilated to 3 cm. Ultrasound revealed no cardiac activity and overlapping of the cranial bones. Laboratory tests (CBC, PT, AST, ALT, LDH, urea, and creatinine) were taken, and the patient gave birth to a stillborn, macerated male fetus weighing 900 g (between the 5th and 10th percentile) two hours later. The weight of the placenta was 220 g. A post-mortem examination showed organ autolysis, but no congenital abnormalities were detected. During the macroscopic examination of the placenta, small areas of white-gray, firm, irregular thickening were observed on the maternal side (Figure *1*). Examination of the umbilical cord revealed the presence of a single umbilical artery.

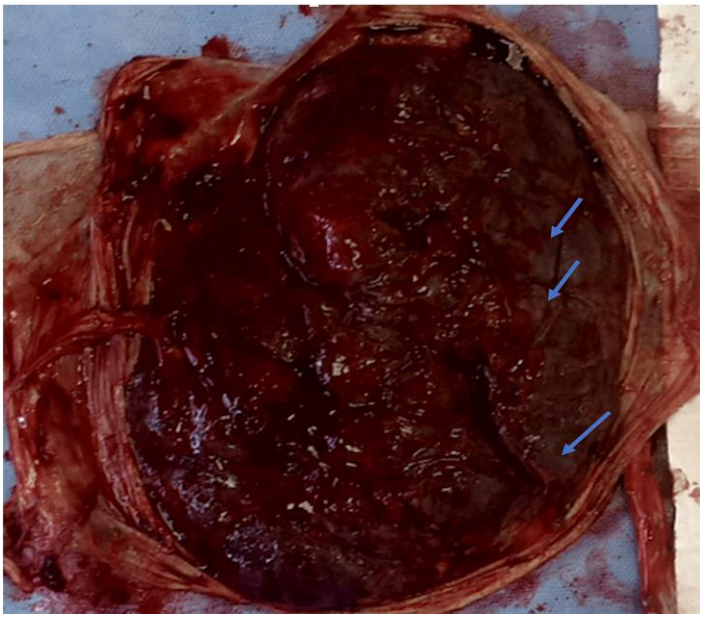

Figure 1. Small irregular placental thickenings on the maternal side - Macroscopic examination

Microscopic examination showed the presence of fibrin deposits enclosing necrotic mummified villi (Figure *2: a*) and filling the intervillous space, leaving only rare areas of healthy villi without fibrin (Figure *2: b*).

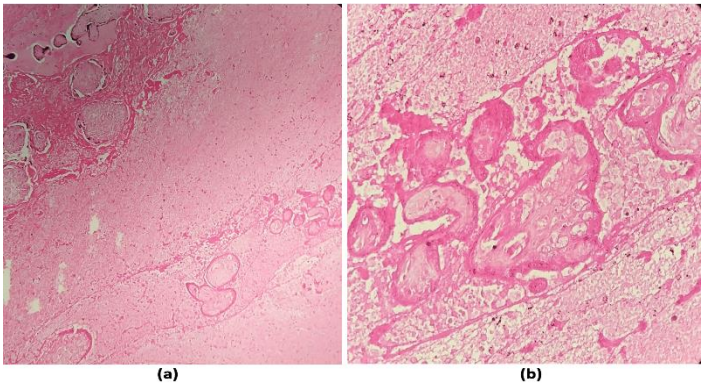

Figure 2. (a): Illustration of Ischemic Chorionic Villi Surrounded by Fibrin Deposits (HEx400) - Microscopic Image. (b): Extent of Ischemic Villous Necrosis with Fibrin Deposits (HEx100) - Microscopic Image.

3. DISCUSSION

MPFD is typically regarded as a rare pregnancy condition, while its incidence is not fully known. The reported incidence of MPFD among all pregnancies was 0.029% and 0.004%, respectively, according to two investigations [1, 2]. These numbers, however, might not fully reflect the true incidence of MPFD due to the condition's rarity.

MPFD can be difficult to diagnose as the symptoms can be similar to those of other obstetrical complications such as pre-eclampsia or placental abruption. Clinical signs include severe abdominal pain, vaginal bleeding, hypovolemic shock, and rapid deterioration of the mother's and fetus's condition. Laboratory tests can reveal disseminated intravascular coagulation (DIC) and microangiopathic hemolytic anemia (MAHA). Ultrasound may show decreased amniotic fluid, fetal growth retardation, or placental abnormalities [3].

The diagnosis of MPFD is often confirmed by histological examination of the placenta after delivery. Histological examination reveals massive fibrin deposits in the intervillous spaces of the placenta, as well as necrosis of chorionic villi and vascular thrombosis [4]. These fibrin deposits can lead to obstruction of placental vessels, compromising fetal blood circulation and causing fetal hypoxia [5]. Histological examination may also reveals lymphocytic infiltration and immune cell activation, suggesting a role of inflammation in the pathogenesis of MPFD [6].

Depending on the severity and scope of the problem, treatment for MPFD should be swift and active to reduce risks for both the

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mother and the fetus. In moderate cases, close monitoring of the mother and fetus may be sufficient, along with routine tests to track fetal growth, the volume of amniotic fluid present, and the condition of the mother. However, in severe cases, a blood transfusion or coagulation product administration may be necessary to replace lost blood components and maintain tissue perfusion. Intensive care may also be necessary for the mother and fetus, including oxygen therapy, mechanical ventilation, renal dialysis, and additional blood transfusions [3].

In cases where preterm birth is imminent, corticosteroids may be administered to stimulate fetal lung maturation. In severe cases, emergency cesarean delivery may be necessary to prevent the rapid deterioration of the mother's and fetus's condition, while in less severe cases, induced vaginal delivery may be considered under close monitoring [4].

It is crucial to emphasize that MPFD is an unusual but potentially fatal illness for both the mother and the fetus to lower risks and improve obstetric outcomes [6]. Thus, urgent and intensive treatment is necessary, treatment choices should be tailored to each circumstance based on the nature of the illness, the health of the mother, and the state of the fetus. Close and regular follow-up is also required to monitor the condition's progression and adjust management as appropriate [3].

4. CONCLUSION

MPFD is a rare but dangerous pregnancy disorder for which currently exists a paucity of epidemiological information. The frequency of it is not fully known, according to recent studies, however, it is typically regarded as a rare obstetric trouble. However, further studies are needed to obtain more accurate estimates of its incidence and better understand the pathophysiology, risk factors, and optimal management strategies. Pregnant women with similar symptoms, such as pre-eclampsia, should have MPFD considered in the differential diagnosis by obstetricians. The best course of action to reduce the likelihood of unfavorable outcomes for both the mother and the fetus is likely early diagnosis and rapid delivery of the infant.

The patient's point of view

The patient was consenting to the therapeutic management and diagnostic approach so she was satisfied with the initial results of the etiological assessment as well as the therapeutic results.

Informed consent

The patient has given her free and informed consent for the writing and publication of this manuscript.

Conflicts of interest

The authors declare no conflicts of interest.

Authors contributions

All authors contributed to the writing and editing of the article. All authors have read and approved the finalized version of the manuscript.

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