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## Case Report

# Placental mesenchymal dysplasia: a diagnostic dilemma

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## ABSTRACT

Placental mesenchymal dysplasia is a rare placental anomaly characterized by placental enlargement, dilatation and congestion of chorionic plate vessels, and villous edema without signs of trophoblastic proliferation. It is often confused with partial hydatidiform mole. It was first diagnosed in 1991 and the reported incidence is 0.02%. We hereby report a case of a 20 years PGR at a period of gestation 26 weeks 5 days with placental mesenchymal dysplasia. PMD is a rare entity that needs to be distinguished from molar pregnancy to prevent unnecessary termination of pregnancy. Once suspected, these patients should be treated as high-risk pregnancies as they are associated with intrauterine growth restriction (IUGR), intrauterine devices (IUD) and congenital anomalies.

**Keywords:** Placenta, Placentomegaly, Placental mesenchymal dysplasia, Partial mole

## INTRODUCTION

Placental mesenchymal dysplasia (PMD) is a rare placental vascular anomaly characterized by placentomegaly and grapelike vesicles. It is estimated to occur in 0.02% of pregnancies, but may be under-represented.<sup>1</sup> Characteristic features of PMD are placental enlargement, dilated and tortuous chorionic vessels which shows thrombosis. It shows a focal distribution of cystically enlarged villi, in a background of grossly normal-appearing villous tissue; hence, the similarity to partial hydatidiform mole (PHM).<sup>2</sup> In contrast to PHM, the histology of PMD features clusters of enlarged, stem villi and absence of trophoblastic proliferation or hyperplasia.

## CASE REPORT

A-27-year, unbooked G2P1001 was referred from a peripheral civil hospital and reported to Dr. Rajendra Prasad Government Medical College (RPGMC), obstetrics and gynecology out patient department at period of gestation (POG) 35 weeks with severe intrauterine growth

restriction (IUGR). An ultrasonography (USG) done showed a live intrauterine fetus with normal skull, spine, and heart with breech presentation. The placenta was fundic and enlarged, measuring 8.9 cm in thickness with multiple cystic spaces within it. No gross congenital anomalies were seen. Her antenatal period till date was uneventful.

She had one previous cesarean delivery two years prior because of fetal distress and a term male child was delivered. The baby is healthy and immunized for age. Past, family and personal history was insignificant.

On admission baseline investigations and doppler ultrasound was done. Doppler showed absent end diastolic flow in umbilical artery. A provisional diagnosis of placental mesenchymal dysplasia with severe IUGR with breech was made. An emergency lower segment caesarean section (LSCS) was done at Dr. RPGMC. An alive preterm female child was delivered with birthweight of 1229 gm. Baby cried immediately on birth. She was admitted in neonatal intensive care unit (NICU) for 2 weeks and

discharged at 1.6 kg. Placenta was grossly enlarged and weighed 550 gm and showed dilated, tortuous veins with a few vesicles ranging from 2 to 6 mm. They were interspersed among normal appearing placenta involving the entire placenta. Intraoperative findings were normal. The neonate was screened for anomalies. The neonate and her mother were discharged in a good condition from the hospital after 2 weeks. Histopathology examination showed stem villous hyperplasia with hydropic changes and thick-walled vessels at the periphery, and these were surrounded by tertiary villi with a normal appearance.



**Figure 1: Gross appearance of the placenta showing thick placenta.**



**Figure 2: Cut section of placenta showing dilated tortuous vessels interspersed in normal looking placenta.**

## DISCUSSION

PMD is a rare, benign condition characterized by placentomegaly and abnormal chorionic villi with vesicle formation, fibroblastic hyperplasia, and vascular abnormalities. PMD is found in approximately 0.02% of pregnancies, with 129 cases of PMD described to date. The diagnosis of PMD requires analysis of ultrasound, and gross and histopathologic findings. PMD is often misdiagnosed as partial hydatidiform mole because of their similarity in ultrasonographic, gross and histologic

presentations. The majority of PMD cases are associated with IUGR in 50%, intrauterine fetal death (IUFD) in 43%, and Beckwith-Wiedemann Syndrome (BWS), which includes macrosomia, exomphalos, macroglossia, omphalocele, internal visceromegaly, and placentomegaly in 25-33%.<sup>5</sup>

PMD is characterized by multiple hypoechoic vesicles which are similar to molar changes in the placenta.<sup>6</sup> By ultrasound, the placenta in PMD is described as large and thickened with multicystic, hypoechoic areas. The differential diagnosis for this appearance includes: partial hydatidiform mole, complete hydatidiform mole (CHM) with co-existent fetus, chorioangioma, and intervillous hematoma, infarct or nonspecific hydropic changes.<sup>3</sup>

Color Doppler has recently become a tool to help distinguish PMD from a molar gestation. PMD is reported to show a “stained-glass” appearance suggesting abundant blood flow in PMD while CHM shows little to no blood flow.<sup>4</sup> On color Doppler, high velocity and low resistance flow is seen in the molar mass. A large feeding vessel or increased vascularity is seen in the mass of chorioangioma. No blood flow is seen within the mass in subchorionic hematoma, and spontaneous abortion with hydropic changes.<sup>7</sup> An unusual sonographic finding of dilatation of the umbilical vein with elevated maternal serum alpha-fetoprotein on prenatal testing may be diagnosed as PMD on pathological evaluation of the placenta. Elevated alpha-fetoprotein for gestational age is elevated in PMD. Karyotype can also be of aid in diagnosis and differentiating from partial mole.

The diagnosis of PMD is only affirmed after evaluation of placental pathology. Grossly, it is characterized by placentomegaly, dilated or aneurysmal chorionic vessels and enlarged hydropic or cystic villi. Microscopic findings include mesenchymal hyperplasia and edema of stem-cell villi, which contain thick-walled vessels. A characteristic feature is the absence of trophoblastic hyperplasia. Pregnancy outcomes range from healthy, uncomplicated pregnancies, to adverse maternal and/or neonatal complications. The maternal complications are preeclampsia/gestational hypertension, and premature delivery. Fetal complications included severe fetal growth restriction in half of the cases. Fetal anomalies which have been reported with PMD included Beckwith-Wiedemann syndrome, CHARGE syndrome, fetal pleuro-pulmonary blastoma and fetal skeletal dysplasia.<sup>9</sup>

## CONCLUSION

PMD is associated with adverse pregnancy outcome regardless of the presence or absence sonographic detectable fetal abnormality. Women with PMD are at markedly increased risk of intrauterine fetal death and premature delivery. Erroneous diagnosis of PHM may lead to iatrogenic termination. The diagnosis of PMD should be considered with specific sonographic findings. Patients should be counseled regarding potential complications

such as fetal growth restriction, fetal death, premature delivery and maternal pre-eclampsia. Serial growth scans starting in the second trimester and continue until delivered should be strongly considered. Early admission to the hospital and intensive monitoring of fetal wellbeing status should be considered.<sup>10</sup>

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