Placental mesenchymal dysplasia (PMD) is a rare, benign placental, vascular anomaly. PMD is characterized by placentomegaly and grape-like vesicles resembling molar pregnancy on ultrasonography (USG).

The differential diagnosis of PMD includes partial molar pregnancy, complete mole with coexisting normal fetus, chorioangioma, subchorionic hematoma, and spontaneous abortion with hydropic placental changes. However, lack of high-velocity signals inside the lesion and a normal karyotype favor a diagnosis of PMD. PMD must be differentiated from gestational trophoblastic disease because management and outcomes differ. We report the case of an 18-year-old female at 15 weeks of gestation with sonographic findings suggestive of placental mesenchymal dysplasia. The diagnosis was confirmed on histopathology.

Key words: Partial molar pregnancy; placental mesenchymal dysplasia; sonography

Introduction

An 18-year-old female, gravida 1, para 1 was referred for routine ultrasound at 15-week gestation. No complaint of bleeding per vagina or pain abdomen was reported by the patient. Her routine hematological and biochemical parameters were within the normal limits. USG findings showed single live fetus, with mildly thickened, low lying posteriorly placed placenta. Lower part of the placenta showed multiple anechoic cysts which were extending inferiorly and covering the internal os. On color Doppler, low velocity blood flow was seen within the multicystic placental lesion [Figure 1]. A single live fetus of 15-week gestational age without any gross structural anomaly was seen [Figure 2]. Based on USG findings, differential diagnoses of a partial mole or a complete mole with coexisting twin and PMD were considered. Estimation of serum ß-HCG and maternal serum AFP levels were advised. Maternal serum levels of AFP (181.37 ng/ml) and ß-HCG (223846 mIU/ml) were raised.

In view of the presence of normal fetus on ultrasound and low velocity color flow within the cystic placental mass on color Doppler, the possibility of PMD was considered.

Patient refused for an invasive test (amniocentesis). She was kept on conservative management and informed about the increased risk of complications such as fetal growth restriction or intrauterine fetal demise and preterm delivery and association of preeclampsia.

After 1 week, the patient started complaining of heavy bleeding per vagina. Subsequently, dilatation and evacuation was done and placental tissue was sent for histopathological examination. On follow-up β-HCG showed lower levels. Pathological examination suggested hydropic stem cell villi with central cistern containing myxoid stroma with interspersed normal-sized chorionic villi. Few congested blood vessels were seen in the stroma of hydropic villi in the periphery. There was characteristic absence of trophoblastic proliferation around the periphery of the abnormal villi, absence of stromal cell inclusions, and scalloping of villous surface (diagnostic hallmark of molar pregnancy) [Figure 3]. Based on USG and histopathological findings, the final diagnosis of PMD was given.

**Discussion**

PMD is a rare disease entity. Previous studies have described the characteristic placental pathology and the possible genetic basis for the placental abnormality in PMD. PMD cases with Beckwith–Wiedemann syndrome (BWS) are associated with paternal isodisomy of the 11p15.5 region. In addition to association with BWS, there are multiple case reports in the literature describing the coexistence of PMD with fetal hepatic mesenchymal tumors, suggesting a common pathogenetic origin for the two anomalies.\[^{1,2}\]

It is important to distinguish PMD from a partial mole with an abnormal triploid fetus because this diagnosis may result in termination of pregnancy. It is difficult to distinguish PMD from a complete mole with coexisting normal fetus, which carries significant morbidity to the mother (persistent GTD). \[^{3}\] PMD is associated with intrauterine growth restriction and intrauterine fetal demise/neonatal death/without fetal development. \[^{3}\] The cause of fetal death in many cases is fetal vascular obstructive pathology causing longstanding, severe fetal hypoxia characterized by chorionic vessel thrombosis. \[^{3}\]

PMD should be included in the differential diagnosis of cystic lesions of the placenta on USG. Placenta may appear thickened. The placenta of a complete mole with coexisting normal fetus and partial molar pregnancy appears heterogeneous, with partially solid and cystic areas. On USG, a chorioangioma is a focal lesion and is hypoechogenic compared to the rest of the placenta. It is typically located on the fetal surface of the placenta. \[^{4,6}\]

PMD in the first trimester shows no blood flow in the cystic spaces of the placenta on color Doppler. However, in the third trimester, large vascular areas with turbulent blood flow are observed (either arterial or venous), which are located mainly under and at the level of the chorionic plate. These changes are due to the progressive dilatation of chorionic arteries and veins, which become aneurysmal. \[^{7}\] Low or absent venous signals may be associated with PMD during the first two trimesters, as seen in our case also. \[^{11}\]

On color Doppler, high velocity and low resistance flow is seen in the molar mass and large feeding vessel or increased vascularity is seen in the mass of chorioangioma, although no blood flow is seen within the mass in hematoma. \[^{8}\] Differentiation of PMD from a twin pregnancy with a complete mole and coexistent fetus is difficult. On the first trimester sonography, documentation of two gestation sacs, a lesion that constitute the entire thickness of the placenta,
and the lack of blood flow signals suggest the diagnosis of complete mole with coexistent fetus.[11]

The diagnosis of PMD is only confirmed after evaluation of placental pathology. Grossly, it is characterized by placentomegaly, dilated or aneurysmal chorionic vessels, and fibromuscular hyperplasia or cystic villi.[9] Microscopic findings include mesenchymal hyperplasia and edema of stem-cell villi, which contain thick-walled vessels.[2] These findings were seen in the present case along with characteristic absence of trophoblastic hyperplasia, which is a diagnostic hallmark of GTD.[5] The villi do not show proliferation of trophoblasts or stromal trophoblastic inclusions in PMD. There are no abnormal fetal vessels in the stem villi seen in cases of twin pregnancies with one complete mole, as in PMD.[3]

Some immunohistochemical and invasive tests are helpful for the diagnosis of PMD. Immunohistochemical tests using antibodies against p57KIP2 protein (an imprinting gene only expressed in the maternal genome) might prove helpful in distinguishing PMD from molar pregnancies.[10] In a complete mole, the villous cytotrophoblastic cells lack maternal genome and are negative for this test.[11]

Invasive testing (chorionic villus sampling or amniocentesis) should be performed to confirm a normal karyotype and exclude partial molar pregnancies. Partial molar pregnancies demonstrate triploidy, which is rare in PMD. Triploidy associated with PMD is presumed to occur either as a result of maternal-derived triploidy or due to placental mosaicism.[5,6] Low incidence of aneuploidy may occur in association with PMD. Cohen et al. reviewed a total of 66 cases of PMD. Normal karyotype was seen in 78% of the cases, chromosomal abnormalities in 3 cases (Trisomy 13, Klinefelter syndrome and triploidy), and BWS in 15 cases (23%).[3,4,6] Although ß-HCG levels are always elevated in molar pregnancies, PMD may also present with increased ß-HCG levels. PMD is more likely to be associated with elevated maternal serum AFP levels, as seen in our case.[10]

USG appearance suggestive of molar pregnancy along with increased maternal serum AFP levels and normal or slightly elevated ß-HCG levels can suggest the diagnosis of PMD. The possibility for PMD should be evaluated when an abnormal fetus, or an abnormal karyotype, or both are found.[5] A detailed anatomical evaluation should be performed to rule out associated fetal anomalies, mainly findings consistent with BWS. Because PMD is associated with adverse pregnancy outcome, surveillance with serial growth scans, genetic evaluation, and third-trimester assessment of wellbeing should be considered.

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Conflicts of interest
There are no conflicts of interest.

References


