Case Report

Placental Mesenchymal Dysplasia: An Enigma

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Abstract

Placental mesenchymal dysplasia (PMD) is a rare placental abnormality characterized by placentomegaly and grape-like vesicles resembling partial mole by ultrasonography, but in contrast to partial mole can co-exist with a viable fetus. Although the karyotype is normal, the fetus is at increased risk for intrauterine growth restriction, intrauterine fetal demise or perinatal death and Beckwith-Wiedemann syndrome. Prenatal diagnosis is difficult and the final diagnosis is usually achieved by postpartum histological examination of the placenta. PMD is disproportionally affects females. The purpose of reporting this case is to share the successful fetal outcome which occurs rarely, in this even rarer condition, PMD.

Key Word: Placental Mesenchymal Dysplasia.

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INTRODUCTION

First described in 1991 by Moscoso and colleagues, placental mesenchymal dysplasia (PMD) is a rare placental vascular anomaly characterized by placentomegaly and grapelike vesicles resembling partial molar pregnancy. By virtue of the enlarged placenta with multicystic, anechoic regions on sonograpghy and the widely distributed large edematous villi on gross examination, placental mesenchymal disease is very understandably confused with partial hydadiform mole. However PMD remains a distinct clinicopathological entity, molar pregnancies often have a malformed or an absent fetus. PMD is usually characterized by normal fetus, with a fortunate possibility of the pregnancy progressing to the third trimester. Even so, a co-existing phenotypically normal fetus in PMD carries a high risk for fetal growth restriction, intrauterine death and neonatal death. Also, almost a quarter of fetuses are associated with Beckwith-Wiedmann syndrome (BWS).

CASE REPORT

A 32 years, gravida 2 with one previous missed abortion, registered with us at 8 weeks of gestation. The pt had normal NT scan and a low-risk first trimester serum markers. However on anomaly scan at 19 weeks, the ultrasound revealed the placenta to have a large area (occupying more than 40% of the placenta) of multiple tiny cysts. a suspicion was raised of either Placental Mesenchymal Dysplasia or of Partial Mole. The fetus appeared phenotypically normal and there was no evidence of congenital anomalies in the fetus. The patient was counselled and the pregnancy was closely monitored. Serum levels of beta HCG were 67% higher than for the observed gestational age. The pregnancy advanced uneventfully with adequate fetal growth till 30 weeks except for a week long episode of spotting per vaginum which was managed conservatively. At 30 weeks, the follow-up scan revealed persistence of the cystic areas in the placenta along with dolicocephaly in the fetus (Cephalic Index 66). The interval growth of the fetus was still adequate. At 37 weeks, patient developed mild oligohydramnios with biparietal diameter now corresponding to 31-32 weeks. Keeping in mind the high possibility of IUD in cases of PMD, decision to terminate the pregnancy at 38 weeks was taken. The patient had an uneventful vaginal delivery of a healthy female child, weighing 2.7 kg. the gross examination of the placenta revealed pale, friable area of 15x8x2 cm with grape-like cystic structures on the maternal surface. Microscopic examination showed evidence of Placental Mesenchymal

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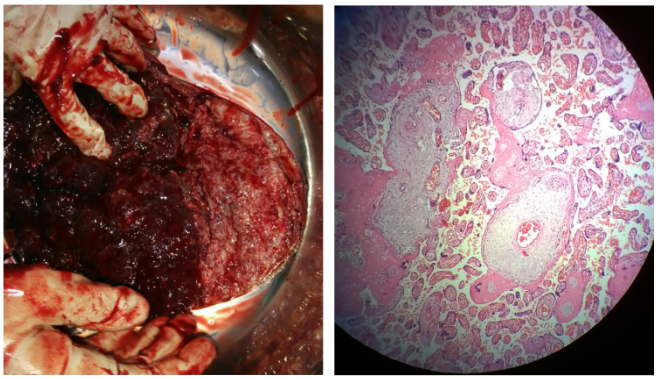
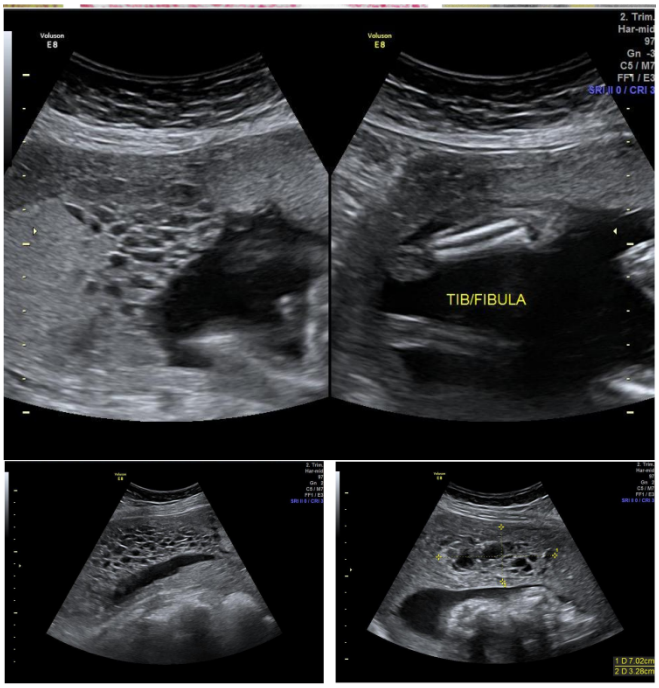
Dysplasia with absence of trophoblastic proliferation, thus ruling out partial mole. Post-natal clinical examination of the fetus revealed no abnormalities as also the Ultrasound examination of the neonate was normal.





DISCUSSION

Placental mesenchmal disease is a potentially misdiagnosed entity and is a relatively recently diagnosed one. The underlying cause of PMD is currently obscured but evidence suggest that it may originate from androgenic/biparental mosaicism confined predominantly to the placenta (Kaiser-Roger *et al*, 2006). This is supported by the finding of 2 separate cell lines in PMD placental tissue. In this condition, the androgenetic cell line is thought to arise from endore duplication of the haploid paternal genome, whereas the biparental cell line arises from the combination of the haploid maternal and the paternal genomes. The abnormal androgenetic cells are confined to chorionic mesoderm, membranes, and vessels, whereas the trophoblastic cells are normal with no evidence of androgenetic cells. This explains the absence of trophoblast overgrowth in PMD in contrast to complete moles in which androgenetic cells are identified in the trophoblastic cell layer. The incidence of PMD is 0.02% with a definite preponderance of females (about 1 in 500 cases (Arisawa and Nakayama 2002) referred for probable molar change. There is no specific clinical symptomatology associated with PMD. Most cases of PMD in early pregnancy are diagnosed by prenatal ultrasonography done either for routine prenatal checkup or because of an abnormal amniocentesis result. The most common abnormal laboratory test includes increased level of maternal serum alpha fetoprotein, which is thought to be of fetal origin. It is speculated that the increase in the surface transfer area because of increased placental volume and increased vessels within the stem villi may lead to increased transfer of alpha fetoprotein into the maternal circulation. The level of β-human chorionic gonadotropin is normal to slightly increased. Although most of the fetuses associated with PMD are phenotypically normal, but fetal complications such as fetal growth restriction, intrauterine death(IUFD) and Beckwith-Wiedemann syndrome (BWS) are also associated with it. Beckwith-Wiedemann syndrome (BWS) is characterized by macrosomia, exomphalos, macroglossia, omphalocele, and internal visceromegaly in addition to placentomegaly and increased susceptibility to childhood tumors whereas prematurity was explained by a potentially chronic hyoxia secondary to obstructive fetal vascular thrombosis and a decrease in maternal-fetal gas exchange as a result of a insufficient amount of normal chrionic villi and the shunting of blood from the exchange surface in chorioangiomas and dysplastic villi. Maternal complications that were associated with PMD were gestational proteinuric hypertension and polyhydramios. The main differential diagnose of PMD both clinicopathologically and sonographically is partial mole or twin pregnancy with molar placenta co-existing with a normal fetus. The sonographic features of PMD are very similar to those of partial moles. A thickened placenta with hypoechoic spaces are classical sonographic findings of both PMD and molar pregnancies. The other differential diagnoses of these ultrasonographic findings include chorioangiomas and subchorionic hematomas. Jauniaux *et al* evaluated 6 suspected cases of PMD with serial prenatal ultrasonography and Doppler imaging that showed cystic spaces located deep in the placental parenchyma and increased placental thickness early in gestation. As the pregnancy advanced, the cystic spaces moved toward the chorionic plate. The chorionic plate vessels, including both the arteries and veins, became progressively dilated and aneurysmal. No abnormal chorionic vessels were seen before midgestation. The

ultrasonographic finding of a large cystic placenta along with phenotypically well-formed fetus is highly unlikely in molar pregnancy and should raise the possibility of PMD. Grossly, the placenta is usually extremely large for gestational age, it weights of more than the 90th percentile. The gross placental findings in PMD vary with gestational age. In third trimester PMD placenta, the chorionic plate vessels are aneurysmally dilated and tortuous and show abnormal branching. The dilated chorionic plate vessels may show luminal thrombosis or can rupture giving rise to subamniotic hemorrhage, which can further compromise the growth restricted fetus. The grapelike cystic vesicles, which are similar grossly to those of molar pregnancy and are usually visible grossly. In rare cases, the vesicle formation is minimal or absent. Before 20 weeks of gestation, the chorionic plate vessels are not dilated and the normal and abnormal areas are not clearly delineated suggesting that the vascular malformations develop progressively secondary to circulatory imbalance and poor vascularization of the dysplastic villi. This could have been the cause in our patient explaining the normal double marker screening and a later pick up sonologically at 19 weeks. The main differential diagnoses of PMD, both clinically and pathologically, are partial hydatidiform moles, a twin gestation with complete mole, spontaneous abortion with hydropic changes, and confined placental mosaicism. Unlike partial moles, the placenta in PMD is almost always diploid and histologically the villi do not show proliferation of trophoblasts or stromal trophoblastic inclusions. it is difficult to determine whether the complications faced by these fetuses are true complications of this disease process or merely coincidental findings. Some of the complications seen in fetuses born with PMD are secondary to BWS without fully developed phenotypic features. The common fetal complications reported in phenotypically normal fetuses associated with PMD are prematurity, intrauterine growth restriction, and intrauterine fetal demise. Obstetrical complications such as polyhydramnios, fetal hydrops, gestational diabetes, and pre-eclampsia may be associated with a large placenta, placentomegaly in PMD, in and of itself, is thought not to be the cause of fetal complications.

CONCLUSION

In conclusion, PMD is a rare and clinically significant lesion. It is important to recognize the detailed gross and histopathological features of this rare disease entity and be able to differentiate this condition from cases presenting as possible partial hydatiform moles. Ultrasonographic findings suggestive of a molar pregnancy because of hypoechoic spaces in the placenta in the presence of a phenotypically normal fetus, a fetus with growth restriction, or a fetus with features of overgrowth should raise the possibility of PMD.

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