Placental Mesenchymal Dysplasia, a Case of Intrauterine Sudden Death

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ABSTRACT

Placental mesenchymal dysplasia (PMD) is a rare condition presenting with enlarged, multicystic placenta like molar changes. Although PMD usually features a normal fetus and the pregnancy often extends into the third trimester, PMD is clinically significant lesion with high rates of FGR, IUFD, and is associated with Beckwith-Wiedemann syndrome (BWS). We report a 30-year old woman at her first pregnancy with intrauterine sudden death at 31 weeks of gestation. The vesicular lesion in her uterus was detected at 10 weeks on ultrasound. The fetus was normal size without any anomaly on ultrasound and normal trophoblastic vascularization by Doppler study during the pregnancy. As the pregnancy advanced, the vesicular lesion decreased in size and no fetal abnormalities were detected. At 28 weeks of gestation an ultrasound detected dilated periumbilical chorionic vessels. We didn't detect severe FGR or abnormal trophoblastic vascularization. At 31 weeks of gestation an intrauterine sudden death of a normal-sized fetus without any anomaly occurred. The placenta was enlarged, and microscopic morphology confirmed a diagnosis of PMD. The chorionic vessels were cirsoid, dilated and tortuous. We determined the rupture of expanded periumbilical chorionic vessels led to fetal death.

INTRODUCTION

Placental mesenchymal dysplasia (PMD), first described in 1991 by Moscoso and colleagues[9] , is a placental vascular anomaly characterized by mesenchymal stem villous hyperplasia.[15] It is a rare condition presenting with macroscopic features of molar changes, placentomegaly and grape-like vesicles in the placenta. Its prevalence is 0.02% (seven cases among 30,758 placentas over a period of 21 years).[1] PMD appears as diffuse molar change sonographically and often has a characteristic macroscopic appearance of the delivered placenta, which is enlarged with multiple cystic structures affecting the parenchyma and prominent, dilated and tortuous chorionic vessels. Histologic findings are diagnostic, demonstrating normal terminal villi mixed with hydropic stem villi, without trophoblast hyperplasia.[4,5,7,9,11,14] Unlike molar pregnancies which are characterized by absent or malformed fetuses, PMD usually features a normal fetus and the pregnancy often extends into the third trimester.[13] Pregnancy complications are common, including preterm delivery in 75%,   intrauterine fetal growth restriction in 20% and intrauterine fetal death in 30%. In addition, approximately 20% of reported cases of PMD are associated with fetal Beckwith-Wiedemann syndrome with a marked predominance (80%) of female fetuses suggesting a possible defect in imprinting.[3] Recent data suggest that placental tissue-specific androgenetic-biparental mosaicism may be the underlying cause of the condition.[6]

We report a case of PMD with intrauterine sudden death of a normal-sized fetus without any anomaly at 31 weeks of gestation. The patient was informed that data from the cases would be submitted for publication and gave her consent.

CLINICAL CASE

A 30-year old woman at her first pregnancy visited previous hospital at 6 weeks' gestation. An ultrasound performed at 10 weeks' gestation detected a vesicular lesion in her uterus. An ultrasound screening performed at

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12 weeks showed a normal size fetus with no detectable anomalies and the Doppler study excluded abnormal trophoblastic vascularization. The vesicular lesion and normal lesion in placenta were clearly divided on ultrasound, so the previous doctor assumed that PMD caused the placental change.

An MRI at 12 weeks didn't detect multivesicular lesion in the placenta and the image was not the typical image of placenta of molar pregnancy.

As the pregnancy advanced, the vesicular lesion decreased in size and no fetal abnormalities were detected.

At 28 weeks of gestation, however, an ultrasound detected dilated periumbilical chorionic vessels. The previous doctor thought that PMD caused the vascular change in placenta and the patient was referred to our center to receive cautious pregnancy management.

We planned ultrasound scans twice a month to monitor the placenta lesions, fetal anatomy and growth.

At 30 weeks of gestation, the placental tissue was found to have the same characteristics (Fig. 1) including the dilated periumbilical chorionic vessels (Fig. 2) and the growth of the fetus was regular (estimated fetal weight 1340 g, -1.2SD) with a normal flow pattern in the umbilical and cerebral arteries.

Figure 1.
Transabdominal ultrasound study at 30 weeks' gestation. One part of placenta (circle) is filled with multiple small hypoechoic cysts.

Figure 2.
Dilated subchorionic vessels at 30 weeks' gestation. The diameter is up to 5.1 centimeters.

At 31 weeks of gestation, the patient visited our center because of a diminishing feeling of fetal movements. An intrauterine fetal death was diagnosed. The patient delivered a 1550 g female fetus with no definite anomalies at 31 weeks 4 days of gestation. The placenta was 15 cm in diameter and the weight was 490g which was greater than 95th percentile for gestation. The serum hCG level just after delivery was 6600mIU/mL. The chorionic vessels were cirsoid, dilated and tortuous (Fig. 3).
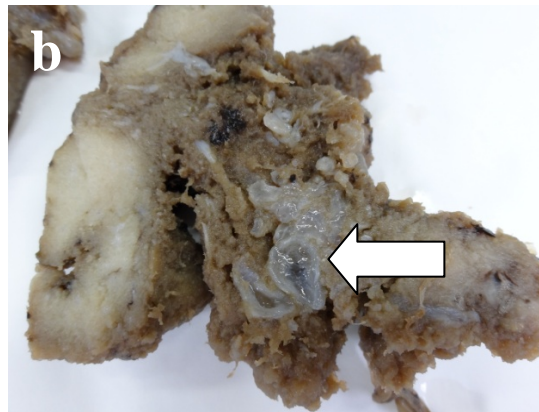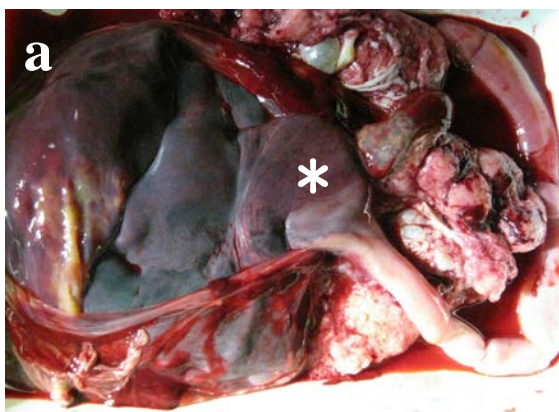

Figure 3.
Fetal surface and cross-section of placenta. a) Periumbilical chorionic vessels are markedly dilated and ruptured (asterisk). b) Closer view of enlarged, edematous villi (arrow).

We considered the rupture of expanded periumbilical chorionic vessels led to fetal death.

The microscopic examination revealed large stem villi with hydropic swelling and cistern formation interspersed with unaffected terminal villi (Fig. 4).
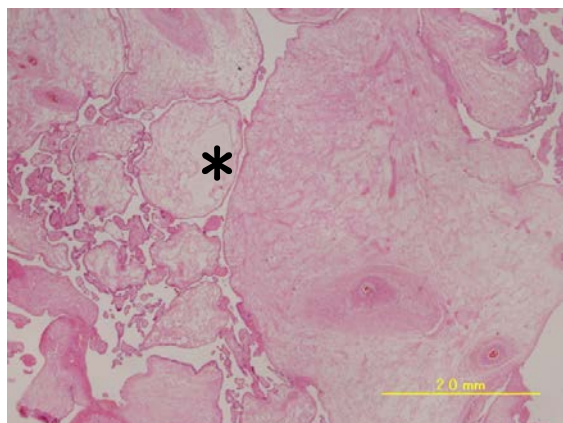

Figure 4.
Hydropic stem villi with cistern (asterisk) formation. Normal terminal villi among the hydropic stem villi.
Vessels are diminished.

The vessels were diminished and there was no trophoblastic hyperplasia. Macroscopic and microscopic morphology confirmed a diagnosis of placental mesenchymal dysplasia.

DISCUSSION

Diffuse cystic molar changes of the placenta in association with an apparently normal fetus in the third trimester is usually due to placental mesenchymal dysplasia (PMD) rather than any type of gestational trophoblastic neoplasia. There have now been around 70 well-documented cases of PMD reported, the prevalence appearing to be around 1 in 4,000 to 5,000 pregnancies.[2] Although the pathogenesis of PMD remains unknown, there may be a possible relationship with X chromosome[1] and androgenetic/biparental mosaicism[15] has recently been suggested as the underlying cause of PMD.

PMD is thought to be caused by a circular disorder.[13] The hypervascularity in the placenta may lead to fetal growth restriction or intrauterine fetal death and maternal hypertension.[12] Therefore PMD is clinically significant lesion with high rates of FGR, IUFD, and is associated with Beckwith-Wiedemann syndrome (macrosomia, exomphalos, macroglossia, omphalocele, craniofacial features, and ear anomalies).

Postdelivery PMD is characterized by a large placenta with parenchymal vesicle and dilated vessels over its surface and weight usually greater than 95th percentile for gestation. Microscopic examination of placental tissue reveals massive hydrops of stem villi and characteristic absence of trophoblastic proliferation and villous trophoblastic inclusion. Terminal villi are normal or mildly edematous.[8] Absence of trophoblastic proliferation in PMD placentas is the main histological difference from partial moles. For this reason, in PMD, levels of maternal serum hCG are normal or slightly increased throughout gestation.[12] A placenta in PMD cases looks similar to the one in a partial mole or twin pregnancy with a complete mole and surviving co-existent fetus. In fact, the sonographic features (thickened placenta with hypoechoic spaces) of PMD are very similar to those of partial moles. Unlike molar pregnancies, PMD usually features a normal fetus and we can expect better prognosis. Even though PMD is a rare condition, it is important to distinguish PMD from mimics, especially molar pregnancy, for preventing the unnecessary termination of pregnancy.

There are several reports in which polycystic lesions of the placenta during the second trimester and dilated subchorionic vessels during the third trimester are detected by sonography. In our case, polycystic lesions of the placenta by sonography revealed at 12 weeks and dilated subchorionic vessels were detected in 28 weeks. Histological findings suggested that the dilated cirsoid chorionic vessels were fragile and eventually a part of the vessels wall had ruptured, causing a hemorrhage with hematoma formation, resulting in IUFD. The vessels were diminished and there may be chorioangiomatoid change of stem villous[8] or aneurysmal dilatation of stem villous vessels.

We observed this case carefully as a high possibility of PMD, but didn't choose the option of hospitalization because severe FGR and abnormal trophoblastic vascularization by Doppler study were excluded. Sudden IUFD including rupture of the cirsoid chorionic vessels might occur at any time throughout gestation. While it is very difficult, it is nonetheless important to set the optimal time of termination of pregnancy for the case which has neither severe FGR nor abnormal trophoblastic vascularization . We will need to precisely diagnose this disease and closely consult with the patient and receive informed consent for the decision on termination of the pregnancy.

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