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A case of placental mesenchymal dysplasia with one year follow-up

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Summary

Placental mesenchymal dysplasia (PMD) is a rare placental disease with unknown etiology. Because of the malformation of the placenta, the incidence of the obstetric complications and the poor perinatal outcomes are higher than usual. PMD usually accompanies with Beckwith-Wiedemann syndrome. In this article, the authors report a case of PMD with a live delivery in 35+5 gestational weeks. The phenotype of the neonate was normal. During the one-year follow-up, the development of the child was normal.

Key words: Placental mesenchymal dysplasia; Obstetric complications; Pregnancy outcome; Beckwith-Wiedemann syndrome.

Introduction

Placental mesenchymal dysplasia (PMD) is a rare placental disease with unknown cause. The pathogenesis of PMD remains unclear. It was first reported by Jauniaux *et al.* [1] in 1997. Only approximately 100 cases of this disease have been reported so far in English literature. Zeng *et al.* [2] conducted an investigation at the Department of Gynecology and Obstetrics, McGill University of Quebec, Canada for 18 years (1991–2009). A total of 95,000 babies were delivered; however, only two cases were diagnosed with placental mesenchymal dysplasia. Thus, the incidence rate was only 0.02‰.

The sonographic and macroscopic features of PMD are similar to partial hydatidiform mole. It is difficult to distinguish these two diseases during prenatal care. Because of the malformation of the placenta, the incidences of the obstetric complications and the poor perinatal outcomes are higher than usual. Here, the authors report a case of PMD with a good outcome.

Case Report

In this case, the gravida was 23-years-old. She usually had regular menstruation. This was her first pregnancy. She did not experience nausea, vomiting, and other reactions in early pregnancy. She also had no history of vaginal bleeding, drainage, toxic substance and drug use, and radiation exposure. At 11+6 weeks, inspection card was used for regular prenatal examinations. At 13+6 weeks, the crown-rump length and NT of the fetus in the uterus were 7.46 and 1.3 mm, respectively. A honeycomb clump (8.8×4.2×7.6 cm) was attached to the uterine back wall, revealing an abnormal intrauterine echo group (suspected partial malignant mole) (Figure 1A). Down's screening in the middle stage of pregnancy (16+1 weeks) showed that the free β-hCG was 29.07 ng/ml, which indicated a normal result, and APF (MOM) was 6.22 higher than normal. The finding revealed a high risk of open neural tube defect. Targeted ultrasonic examination showed no abnormality. The repeated B-ultrasonic examinations during pregnancy showed abnormal intrauterine echo group. At 30+3 weeks, intrauterine growth restriction was found. Ultrasonic examination revealed that biparietal diameter, femur length, head circumference, and abnormal circumference of the infant were 7.0, 5.1, 25.9, and 22.6 cm, respectively. The patient was only asked to improve nutrition without special drug treatment. No abnormalities were revealed in the remaining pregnancy inspection. An increase in gravida body weight of approximately 10 kg was found.

At 35+5 gestational weeks, premature rupture of membranes occurred and the patient was immediately admitted to the hospital. After admission, the patient gave birth to a live premature female infant. The premature baby weighed 1,800 grams and measured 43 cm in length without obvious deformity. The Apgar score of the newborn was 10-10-10. The placenta measured approximately 23×20×4 cm and weighed 760 grams. Intact placenta and membranes were observed. Approximately 0.2–1 cm beaded blisters were directly found distributed in the maternal placenta. Approximately 8×7 cm area of the beaded structure was found clustered in the maternal placenta (Figure 1B). No abnormalities in the navel cord, which adhered to the placenta paracentralis, were found.

On the second postpartum day, blood examination showed ThCG 4335.0 mIU/ml. The blood ThCG on the 18th postpartum day was reduced to 7.5 mIU/ml, which was within the normal range.

On the first day after birth, the baby was admitted in the Neonatology Department because of pathological jaundice and treated for five days. No abnormalities were found in the appearance, phenotype, and all blood examination indicators of the newborn. Since the birth of the baby one year later, no abnormalities were also observed in her growth and development.

Postpartum placental pathology showed less mature single placenta, placental mesenchymal dysplasia (Figure 1C), and placental focal infarction accompanied by calcification, mild chorioam-

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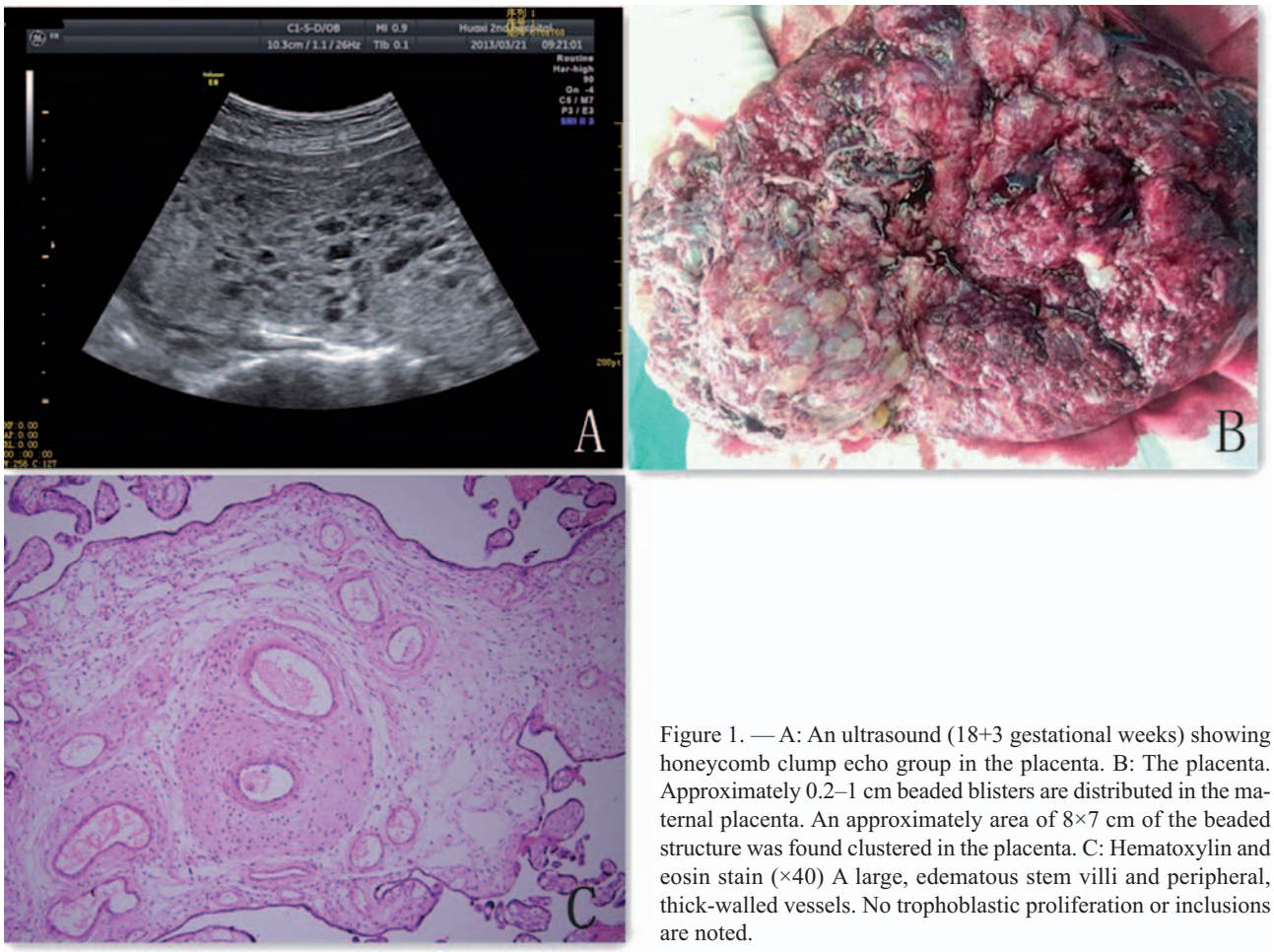

Figure 1. — A: An ultrasound (18+3 gestational weeks) showing honeycomb clump echo group in the placenta. B: The placenta. Approximately 0.2–1 cm beaded blisters are distributed in the maternal placenta. An approximately area of 8×7 cm of the beaded structure was found clustered in the placenta. C: Hematoxylin and eosin stain (×40) A large, edematous stem villi and peripheral, thick-walled vessels. No trophoblastic proliferation or inclusions are noted.

nionitis, Wharton's Jelly mild edema, and normal umbilical cord.

Discussion

PMD can easily be misdiagnosed as partial mole on imaging because the disease is difficult to distinguish. This disease is mostly diagnosed through postnatal pathological examination. Its main pathological features are cystic expansion manifested in the placental stem, but no trophoblastic hyperplasia. The hCG of the patient without trophoblastic hyperplasia is similar to that in normal pregnancies. The hCG in this case was sharply reduced after operation and then restored to normal on the 18th postpartum day.

Most of the patients diagnosed with PMD have normal karyotype [3, 4]. However, some studies report that PMD may be related to abnormal karyotype, such as androgenetic-biparental mosaicism [5] and cytogenetic ploidy [6]. The incidence of the disease is also found more in females than males [6]. The newborn described in this paper was female. Unfortunately, chromosome testing was not conducted because of the disagreement from the patient and her family.

According to a current literature, placental echo abnormality is found in most PMD patients at the middle stage of pregnancy or 13–20 weeks, showing honeycomb performance without fetal malformation [7]. During this time, the disease can be distinguished from partial mole through blood hCG inspection, amniocentesis, or umbilical cord puncture [3]. Doctors always inform patients of abnormal findings during prenatal check-ups. Some pregnant women will opt to terminate the pregnancy [4], whereas some will choose to continue the pregnancy. However, other pregnant women do not undergo relative inspection to continue the pregnancy because of the lack of knowledge on this disease. The patients who continue the pregnancy are more at risk of suffering from obstetrical complications, such as severe preeclampsia [8], oligohydramnios [8], intrauterine fetal growth restriction [3], premature birth [9], hemangioma [9], and in severe cases, fetal death [6, 8]. Pregnant patients rarely continue the pregnancy until the late stage and have uncomplicated delivery of live newborn babies [9]. In this paper, placental echo abnormality was initially found at 13+6 weeks of pregnancy. The patient had a normal hCG. No fetal chro-

mosomal abnormalities were found and no abnormalities were also observed in the remaining inspections except for fetus growth restriction. When pregnancy reached 35+5 weeks, her weight increased approximately ten kg, meeting the weight increase standard. However, the fetus growth was restricted, indicating that PMD may affect maternal–fetal nutrient exchange.

Approximately 20%–50% of placental mesenchymal dysplasia in newborns is accompanied with fetal Beckwith–Wiedemann syndrome [4, 6], which is a congenital overgrowth. Infants may develop this disorder before birth [10]. After birth, newborns may have umbilical hernia, macroglossia, visceral mast, adrenocortical cell hypertrophy, hypoglycemia, and other diseases. The case in this paper did not have the aforementioned abnormalities after birth. During one year follow-up, no abnormalities in growth and development were found. The authors inferred that the influence of PMD to the fetus only existed in the uterus. When the baby was born, there was no influence on the growth of the normal phenotype baby.

Conclusion

PMD is a rare malformation of placenta. It is difficult to distinguish from the partial mole during prenatal care and diagnose definitely. If the prenatal examination indicates an abnormal placental echo, doctor should be aware of this disease and perform related examinations, such as karyotype and hCG. More frequent prenatal care is needed to find the obstetric complications, treat the complications in a timely manner, and improve the outcome of pregnancy.

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References

[1] Jauniaux E., Nicolaides K.H., Hustin J.: "Perinatal features associated with placental mesenchymal dysplasia". *Placenta*, 1997, *18*, 701.
[2] Zeng X., Chen M.F., Bureau Y.A., Brown R.: "Placental mesenchymal dysplasia and an estimation of the population incidence". *Acta Obstet. Gynecol. Scand.*, 2012, *91*, 754.
[3] Kotani T., Sumigama S., Tsuda H., Mano Y., Yamamoto E., Iwase A., *et al.*: "A case report of placental mesenchymal dysplasia with an increased VEGF-D expression". *Placenta*, 2012, *33*, 888.
[4] Agarwal R., Khatuja R., Sharma L., Singh A.: "Placental mesenchymal dysplasia: a case report". *Case Rep. Obstet. Gynecol.*, 2012, *2012*, 202797.
[5] Kapur R.P., Cole B., Zhang M., Lin J., Fligner C.L.: "Placental mesenchymal dysplasia and fetal renal-hepatic-pancreatic dysplasia: androgenetic-biparental mosaicism and pathogenesis of an autosomal recessive disorder". *Pediatr. Dev. Pathol.*, 2013, *16*, 191.
[6] Cohen M.C., Roper E.C., Sebire N.J., Stanek J., Anumba D.O.: "Placental mesenchymal dysplasia associated with fetal aneuploidy". *Prenat. Diagn.*, 2005, *25*, 187.
[7] Ohira S., Ookubo N., Tanaka K., Takatsu A., Kobara H., Kikuchi N., *et al.*: "Placental mesenchymal dysplasia: chronological observation of placental images during gestation and review of the literature". *Gynecol. Obstet. Invest.*, 2013, *75*, 217.
[8] He Lijuan, Gao Qiu, Ye Guangming, Li Dongzhi: "A case of placental mesenchymal dysplasia". *J. Diagn. Pathol.*, 2010, *2*, 158.
[9] Qichang W., Wenbo W., Liangkai Z., Hui K., Xiaoqin H., Li S., Yasong X.: "Pregnancy with concomitant chorioangioma and placental mesenchymal dysplasia: a rare placental abnormality". *Case Rep. Obstet. Gynecol.*, 2013, *2013*, 591956.
[10] Hikita R., Kobayashi Y., Tsuji M., Kawamoto T., Moriyama K.: "Long-term orthodontic and surgical treatment and stability of a patient with Beckwith-Wiedemann syndrome". *Am. J. Orthod. Dentofacial. Orthop.*, 2014, *145*, 672.

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