Case Report

Placental Mesenchymal Dysplasia Complicated by Hydrops Fetalis and Fetal Death: A Case Report

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Abstract

Placental mesenchymal dysplasia is a rare condition of the placenta and its true incidence and underlying cause has remained unknown till now due to its rarity. Its accurate diagnosis is essential, because placental mesenchymal dysplasia is usually compatible with a good fetal and maternal outcome. A precise ultrasonographic evaluation can contribute to the identification of characteristic features, particularly to discriminate it from partial hydatidiform mole, its main differential diagnosis. We report an early third-trimester pathologically- diagnosed case of placental mesenchymal dysplasia. It was complicated by fetal hydrops and death.

Keywords: Hydrops fetalis, partial mole, placenta, placental mesenchymal dysplasia

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Introduction

Placental mesenchymal dysplasia (PMD) was first described in 1991 by Moscoso and colleagues.[1] Clinically and pathologically, PMD has differential diagnosis with partial hydatidiform mole, twin pregnancy with complete mole, spontaneous miscarriage with hydropic changes, as well as confined placental mosaicism.[2] On pathologic examination, the placenta is enlarged with tortuous and dilated chorionic vessels on the fetal surface.[1,3] Other characteristic features are stem villous hydrops without trophoblastic proliferation, villous chorangiosis, and chorangimatosis.[1] The fetus usually has normal karyotype with female predominance and is associated with intrauterine growth retardation (IUGR), stillbirth, and Beckwith-Widemann syndrome (BWS), or normal fetal morphology.[1,3]

Case Report

A 26-year-old lady, gravid 1 with the gestational age of 28 weeks, was referred to the obstetrician with chief complaint of decreased fetal activity for the previous week. Ultrasonographic evaluation showed a single dead fetus, with marked tissue edema, ascites, and pleural effusion, and a large hyperechoic mass measuring 118 x 80 mm in the placenta (Figure1). The findings were suggestive of hydrops fetalis (Figure 2). She delivered a single 1500 g female dead fetus with hydrops fetalis. Both mother and the fetus were RH+. The parents of the fetus were not relatives. On autopsy, there was a massive tissue edema, pleural and pericardial effusion, and ascites. No gross anomaly was identified. The placenta was enlarged, weighing 750 g (> 90th percentile for gestational age), and measuring 20 cm in the greatest diameter and 8 cm in maximal thickness. The fetal surface showed dilated and tortuous chorionic vessels (Figure 3). The maternal surface was pale. The cut sections revealed a relatively well-defined unencapsulated brown-black spongy mass at the periphery measuring 11 × 7 × 4 cm. Also, some grape-like vesicles measuring 0.2 to 1.5 cm were present in the parenchyma (Figure 4). The umbilical cord was too long measuring 100 cm in length and 2 cm in diameter with membranous insertion. The cut section showed three vessels.

Microscopic examination showed dilated chorionic vessels with endothelial damage, and hemorrhage into the vessel wall, some of which with luminal thrombosis. Some stem villi showed cystic changes without trophoblastic proliferation (Figure 5). Terminal villi showed maturation compatible with gestational age. An area of chorangiomatosis was identified with proliferation of large vessels resembling cavernous hemangioma in the enlarged stem villi. Numerous nucleated RBCs were present in the vascular lumens (Figure 6).

Discussion

PMD is characterized by placentomegaly, mostly more than 90th percentile,[1,4] and dilated and tortuous vessels on chorionic plate being prominent in the third trimester.[1,3,5] These dilated vessels are complicated by luminal thrombosis or rupture leading to subamniotic hemorrhage.[1,3] In the placental parenchyma, multiple vesicles are seen especially in the first and second trimesters. These vesicular changes grossly and ultrasonographically resemble partial mole measuring 0.3 to 2.5 cm.[1–6] In microscopic examination of the placenta, stem villous hydrops and cistern formation are evident, but without trophoblastic proliferation, stromal pseudoinclusion, or scalloping of the villi which is differentiated from partial mole. Terminal villi are unaffected.[1,3,4]

Umbilical cord anomaly such as excessive length, hypercoiling, and single artery are reported in PMD.[3,7] The vessels of villi show various changes ranging from normal to chorangiomatous or myxoangiomatosis. Discrete chorangiomas and chorangimatosis are identified in some cases,[1,4,8] and in some of them these vascular changes are associated with the presence of nucleated fetal erythrocytes and extramedullary hematopoiesis.[4,5]

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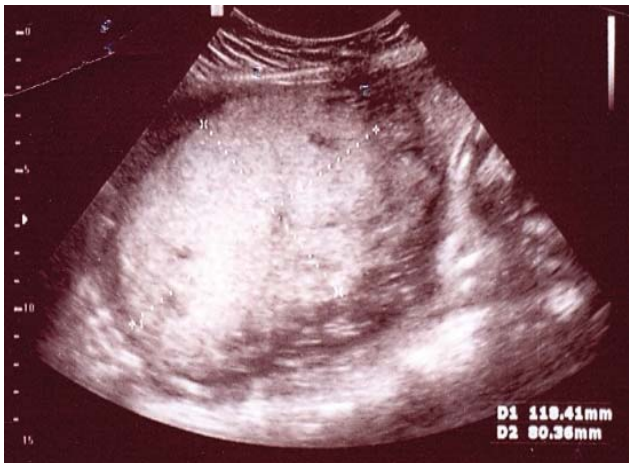

Figure 1. A large hyperechoic mass in the placenta measuring 118 x 80 mm.

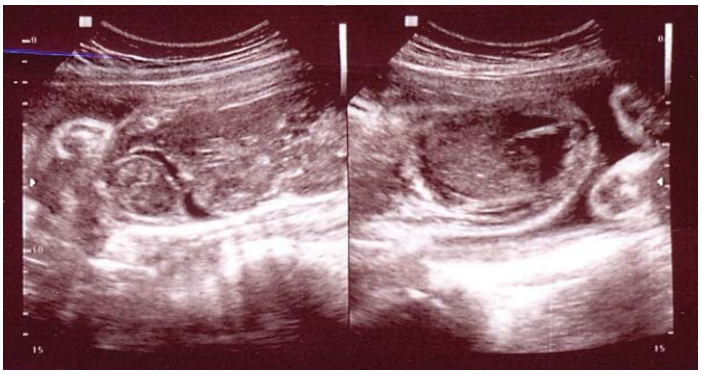

Figure 2. Ascitic fluid in the abdomen of the fetus.

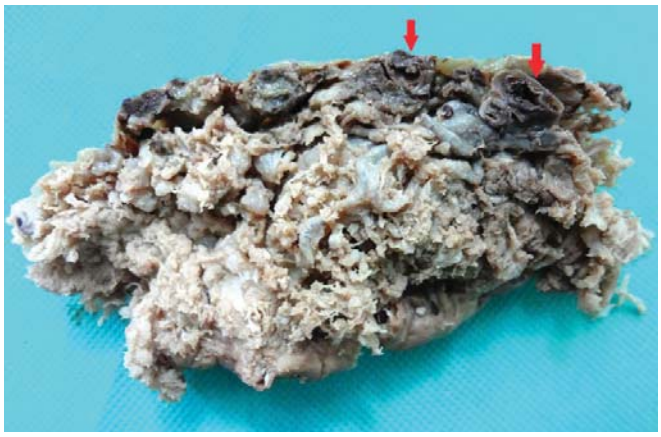

Figure 3. Cut surface of the placenta shows dilated, tortuous, and thrombosed chorionic vessels.

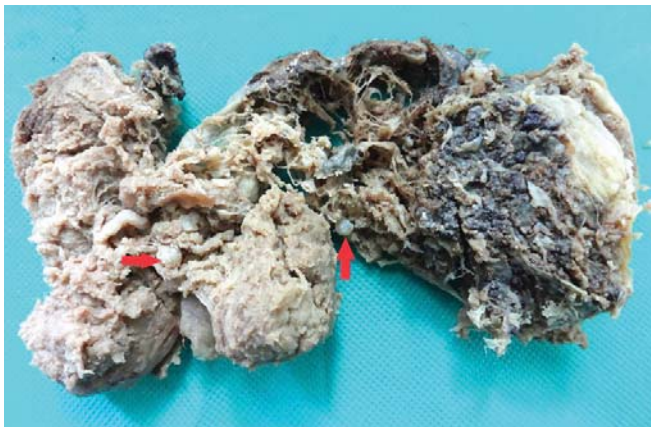

Figure 4. Well-defined dark- brown area at the periphery of the placenta. Some vesicles are evident (arrows).

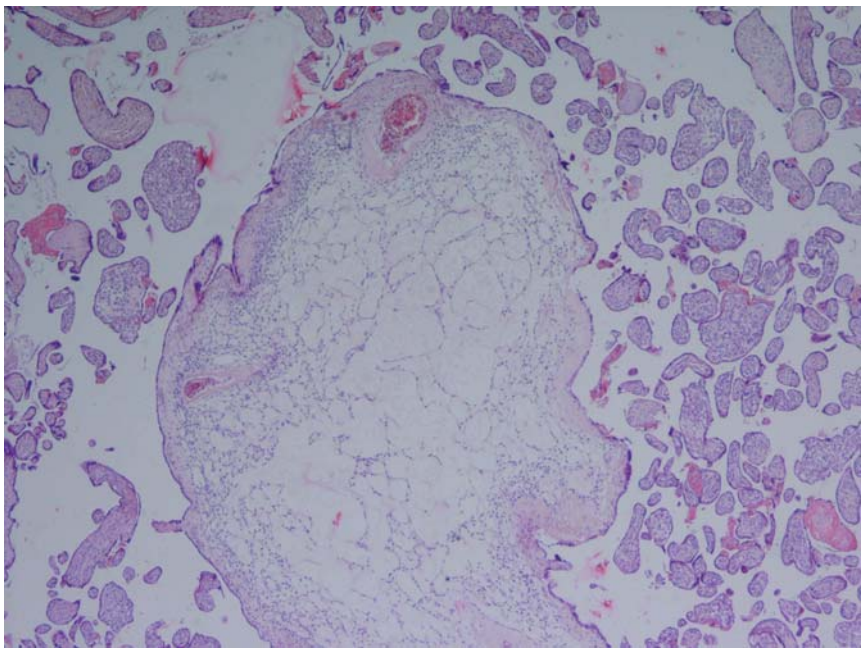

Figure 5. Markedly edematous stem villous and some terminal villi with chorangiosis (H&E x100).

PMD has individual clinicopathologic features and is usually associated with normal fetus; unlike hydatidiform mole, the pregnancy extends to the third trimester. In about 25 % of cases, fetus presents with BWS that is characterized by macrosomia, visceromegaly, hemihyperplasia, macroglossia, omphalocele, and adrenal cytomegally.[1,7,9] In phenotypically normal fetus, prematurity, IUGR, and intrauterine fetal death are reported.[1,4,10] In all cases of PMD, a female preponderance is noted.[1,3–8,10]

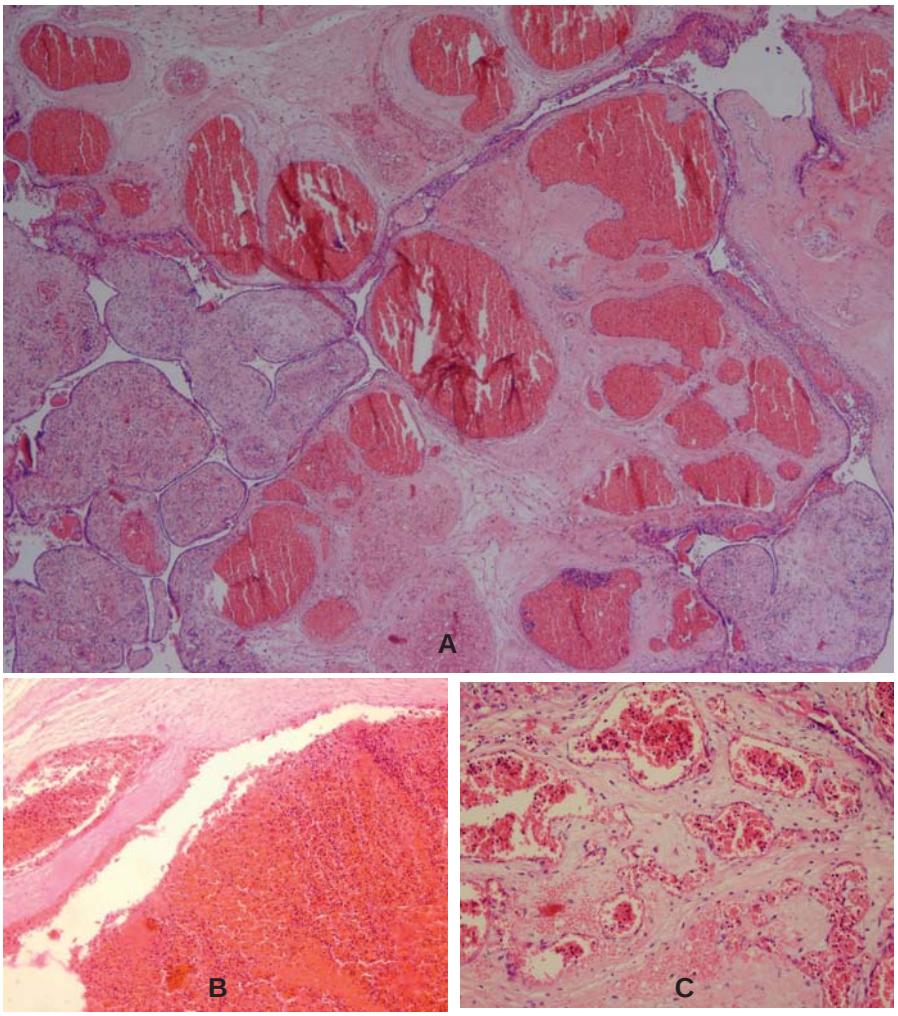

Figure 6. A) Proliferation of large- sized vessels resembling cavernous hemangioma, packed with red blood cells in the stem villi and proliferation of small -sized vessels in the intermediate villi (H&E x 100). **B and C)** Nucleated RBC in vessels (H&E x 400).

The underlying cause of PMD is unclear. Some genetic anomalies are considered as etiology of PMD. Due to the high association of PMD with BWS, abnormal expression of imprinting gene on chromosome 11p15.5 is one of the theories.[1] Hypoxia and hypoperfusion with unknown cause may lead to vascular abnormality seen in PMD due to production of vascular endothelial growth factor (VEGF) by villous macrophages.[1] VEGF-D gene is located on chromosome Xp22.31 and has been proposed for the development of PMD.[1,10] Androgenic/biparental mosaicism is another theory suggested as the cause of PMD. Kaiser-Rogers, et al. identified two separate cell lines in PMD tissue.[1,7,11] They explained that failure of reduplication of the maternal genome before the first cleavage with normal reduplication and segregation of paternal genome caused to produce two types of daughter cells, one with haploid paternal genes and the other with normal biparental genes. Endoduplication of haploid paternal-only daughter cell bear diploid androgenic line.[11] Since the androgenic cell line was originated from duplication of haploid paternal genome and 46, YY cell line is incompatible with life, female fetus predominance in PMD can be explained by androgenic/biparental mosaicism.[1,7,10]

Herein, we presented a case of PMD reported ultrasonographically as a placental mass with intrauterine fetal death and hydrops fetalis at the early third trimester. Previous ultrasonographic reports were not available. To the best of our knowledge, placental mass associated with PMD has not been reported in English articles. Pathologic examination showed placentomegaly with a well-defined mass, long umbilical cord (100 cm) with membranous insertion, and vascular changes in the large chorionic vessels. Vascular changes resembling cavernous hemangioma were seen in the stem villi at the area of placental mass (chorangimatosis). There was enlarged stem villi with cistern formation without trophoblastic proliferation. In the reported cases of PMD, the proliferated vessels were small sized, resembling capillary hemangioma. In our dead fetus, massive tissue edema, pleural and pericardial effusion, and ascites were detected. Placental chorangioma is reported as a rare etiologic factor of nonimmune hydrops. It may cause cardiac decompensation due to the right to left shunt and increased heart output.[12,13] The fetal death in this case might be due to the marked vascular changes in the placenta leading to cardiac decompensation, hydrops fetalis, and intrauterine death.

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References

1. Parveen Z, Tongson-Ignacio JE, Fraser CR, Killeen JL, Thompson KS. Placental mesenchymal dysplasia. *Arch Pathol Lab Med*. 2007; **131:** 131 – 137.
2. Robertson M, Geerts LT, de Jong G, Wainwright H. Mesenchymal dysplasia in a monochorionic diamniotic twin pregnancy with review of the differential diagnosis of cystic changes in the placenta. *J Ultrasound Med.* 2007; **26:** 689 – 693.
3. Pham T, Steele J, Stayboldt C, Chan L, Benirschke K. Placental mesenchymal dysplasia is associated with high rates of intrauterine growth restriction and fetal demise. *Am J Clin Pathol.* 2006; **126:** 67 – 78.
4. Paradinas FJ, Sebire NJ, Fisher RA, Rees HC, Foskett M, Seckl MJ, et al. Pseudo-partial moles: placental stem vessel hydrops and the association with Beckwith-Wiedemann syndrome and complete moles. *Histopathology*. 2001; **39:** 447 – 454.
5. Yuen FC, Sampson A. Placental mesenchymal dysplasia: A report of four cases with differentiation from partial hydatidiform mole. *Aust N Z J Obstet Gynaecol.* 2003; **43:** 475 – 479.
6. Gizzo S, Gangi SD, Patrelli TS, Saccardi C, Antona D, Nardelli GB. Placental mesenchymal dysplasia: can early diagnosis ensure a good materno-fetal outcome? A case report. *Arch Gynecol Obstet.* 2012; **286:** 15 – 17.
7. Umazume T, Kataoka S, Kamamuta K, Tanuma F, Sumie A, Shirogane T, et al. Placental mesenchymal dysplasia: a case of intrauterine sudden death of fetus with rupture of cirsoid periumbilical chorionic vessels. *Diagnostic Pathology*. 2011; **38:** 1 – 7.
8. Woo GW, Rocha FG, Oishi MG, Bartholomew ML, Thompson KS. Placental mesenchymal dysplasia. *Am J Obstet Gynecol.* 2011; **205(6):** e3 – e5.
9. Gibson BR, Padilla JM, Champeaux A, Suarez ES. Mesenchymal dysplasia of the placenta. *Placenta.* 2004; **25:** 671 – 672.
10. Heazell AEP, Sahasrabudhe N, Grossmith AK, Martindale EA, Bhatia K. A case of intrauterine growth restriction in association with placental mesenchymal dysplasia with abnormal placental lymphatic development. *Placenta*. 2009; **30:** 654 – 657.
11. Kaiser-Rogers KA, McFadden DE, Livasy CA, Dansereau J, Jiang R, Knops JF, et al. Androgenetic/biparental mosaicism causes placental mesenchymal dysplasia, *J Med Genet.* 2006; **43:** 187 – 192.
12. Shafqat G, Iqbal F, Rizvi F. Chorioangioma of the placenta with hydrops foetalis. *J Pak Med Assoc.* 2009; **59:** 411 – 412.
13. Sivaslı1 E, Tekşam O, Haliloğlu M, Güçer S, Orhan D, Gürgey A, et al. Hydrops fetalis associated with chorioangioma and thrombosis of umbilical vein. *Turk J Pediatr.* 2009; **51:** 515 – 518.