Asian Journal of Pharmaceutical and Health Sciences
www.ajphs.com

Placental mesenchymal dysplasia - a differential diagnosis of molar pregnancy: A case report

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ARTICLE HISTORY

Received: 17.09.2015

Accepted: 12.10.2015

Available online: 30.12.2015

Keywords:

Mesenchymal dysplasia, stillborn, placenta praevia

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ABSTRACT

Placental mesenchymal dysplasia (PMD) is a rare placental abnormality characterised 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. We present a case of a stillborn who had a placenta praevia radiologically, & was seen to have mesenchymal dysplasia on histological examination.

INTRODUCTION

First described in 1991 by Moscoso and colleagues,[1] placental mesenchymal dysplasia (PMD) is a rare lesion. Sonographically, PMD shows findings similar to those of partial hydatidiform mole, such as enlarged placentas with multicystic, anechoic regions (giving a moth-eaten appearance[2-6]), and widely distributed, large, edematous villi as seen under gross examination. PMD has distinct clinicopathologic features. Unlike molar pregnancies, characterized by absent or malformed fetuses, PMD usually features a normal fetus and the pregnancy often extends into the third trimester. About 20% of the fetuses with PMD also have Beckwith-Wiedemannsyndrome ,a condition characterized by macrosomia, visceromegaly, hemihyperplasia (hemihypertrophy), macroglossia, omphalocele, and adrenal cytomegaly that is recognizable prenatally or in infancy.

In all reports of mesenchymal dysplasia so far, fetal death or IUGR has been reported [2,7,8]. No exception was noted in our case.

CASE REPORT

A 22 year female with a post caesarean pregnancy, presented with dribbling per vagina for about 7 days, around 27 weeks of gestation. She was a 2nd gravida and had an uneventful pregnancy 2 & 1/2 years back. The child born after her 1st pregnancy was healthy. The mother is non diabetic, non hypertensive.

On clinical examination, her os was closed, & no bleeding per vagina was seen. Fetal heart sounds were normal.USG abdomen showed a placenta praevia(low lying posterior type, 3 cm away from external os)& polyhydramnios.Fetal abdomen showed some cysts. The cardiac activity was good & so was the amniotic fluid volume. Doppler study was also normal. The patient was admitted &stabilised. In her 32nd week of gestation, the patient complained of sudden pain. On examination there was effacement of os & blood with meconium was seen per vagina. An emergency lower uterine caesarean section was undertaken, & a stillborn baby was delivered. The baby`s abdomen was distended,& placenta was seen to contain many cysts. The mother recovered uneventfully.

We received the large placenta for histological examination. Grossly, it weighed around 850 gm and measured . The umbilical cord was seen to be abnormal with branched umbilical cords and dual insertions. There were some areas resembling a normal placenta & some cystic areas resembling a molar pregnancy.[Picture 2]Cut surfaces showed heterogeneous areas of abnormal tan and gelatinous and normal red-brown and spongy villous tissues [Picture 3]. The tissue was extremely friable. Histology showed, the umbilical cord having 3 vessel lumens but the vessels were markedly dilated, thick-walled with fibromuscular hyperplasia up to 3.0 cm in diameter, and

thrombosed. The vascular walls were extensively degenerated. [Picture 4].These changes have been described by Sander[9] as "hemorrhagic endovasculitis" and by Redline et al[10] as "villous stromal-vascular karyorrhexis." These changes were found most often in cases with IUFD.Villous chorangiosis (hypercapillarization), defined as more than 10 terminal villi showing 10 or more capillaries per villus, was identified diffusely throughout the placental parenchyma uninvolved by dysplastic changes.[11][Picture 5].

The placental parenchyma showed typical features of mesenchymal dysplasia. The stem villi were abnormally enlarged and often surrounded by excessive fibrinoid material. They consisted of a myxofibroblastic proliferation of spindle and stellate mesenchymal cells without cytologic atypia or prominent mitotic activity [Picture 6].{The abundant extracellular matrix supporting the cells consisted primarily of hyaluronic acid, as demonstrated by a loss of alcian blue staining after hyaluronidase digestion.} Villous stromal cystic degeneration, forming cisterns simulating those in molar villi, was present focally. Trophoblastic hyperplasia and inclusions were absent. Central thick-walled blood vessels with constricted lumens and scattered peripheral small capillaries were observed. Some vessels were thrombosed. Nucleated RBCs were identified.[Picture 7].

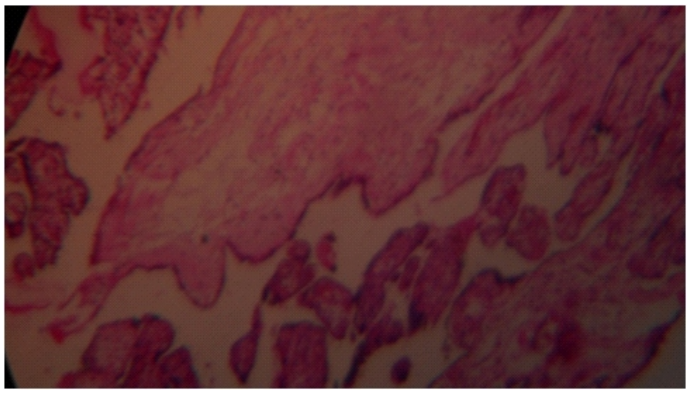

Fig 1 : Eedematous villus L.P. VIEW

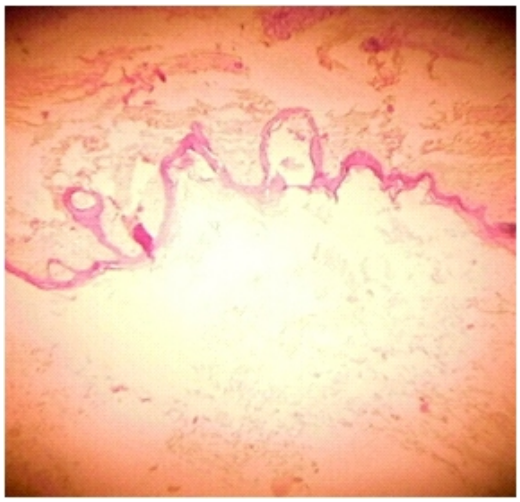

Fig 2

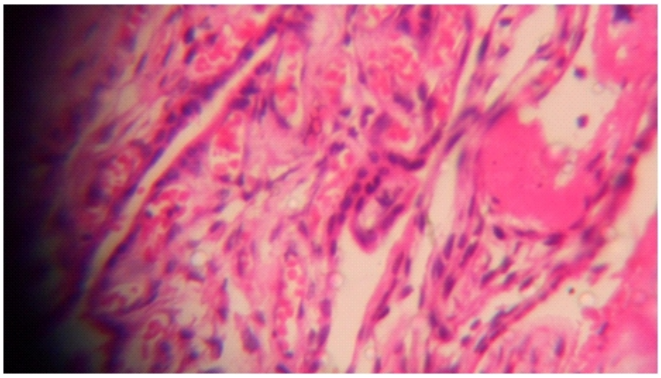

Fig 3 : Increased number of vessel in villi

DISCUSSION

Paradinas et al[12] estimated the rate of 1 Placental mesenchymal dysplasia in 500 cases referred for probable molar pregnancies. Arizawa and Nakayama[13] calculated an incidence of 0.02%.So this condition is rather rare. In a study by Pham et al, cases having abnormal umbilical cords, alongwith mesenchymal dysplasia usually had IUGR or IUFD [7]. A high rate of fetal and neonatal death was found; about 40% [7]. IUGR and fetal death are caused by a variety of genetic and environmental insults.[14-17]

No significant maternal illnesses are reported in association with Placental mesenchymal dysplasia. However, the chorionic vessel thrombosis raises the question of whether there might have been cryptic maternal thrombophilia .Maternal causes include various thrombophilias such as antiphospholipid syndrome, MTHFR, prothrombin and factor V mutations, and systemic lupus erythematosus. Maternal illness, such as hypertension and diabetes mellitus, and narcotic drug use also contribute to fetal morbidity and mortality. Fetal causes include numeric and structural chromosomal abnormalities. The placenta has a significant role in the health of the fetus because it is the sole

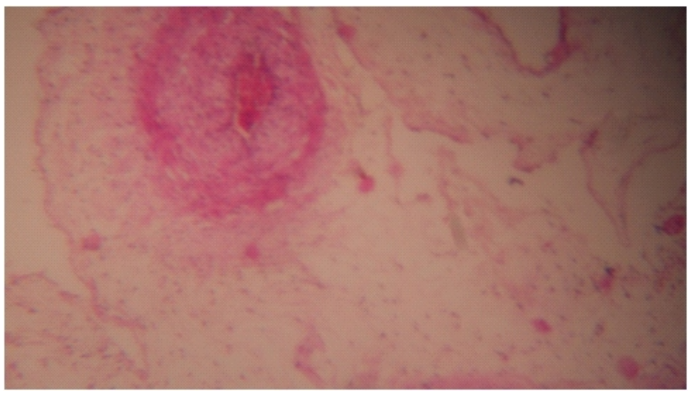

Fig 4 : Thick walled thrombosed umbilical artery L.P. VIEW (3)

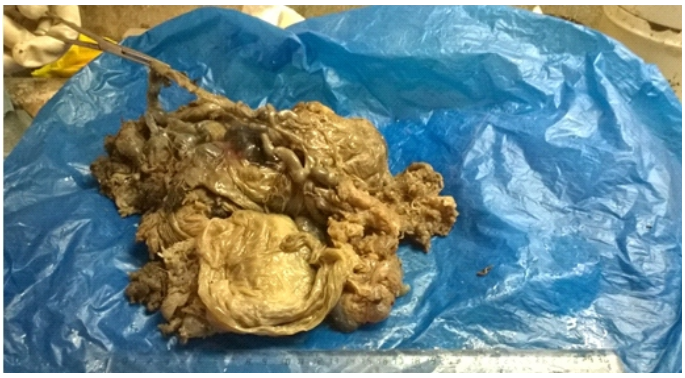

Fig 5

source of nutrients for the fetus and is the first line of defense from the external world. Many placental diseases, including large chorioangiomas (8-17 cm in greatest dimension), maternal and fetal vascular thrombosis, maternal floor infarction, infection, and umbilical cord abnormalities, result in fetal growth restriction and even fetal death.[18,19]

In cases of Placental mesenchymal dysplasia, IUGR and IUFD may be explained by a potentially chronic hypoxia secondary to obstructive fetal vascular thrombosis and decreased maternal-fetal gas exchange due to an insufficient amount of normal chorionic villi and shunting of blood from the exchange surface in chorioangiomas and dysplastic villi. One prominent feature is thrombosed chorionic vessels. Sander[9] has shown that arteries and veins are affected. The marked dilation of these vessels may have caused significant distortion of the vascular channels and altered the flow dynamics, leading to endothelial injury and thrombosis. Redline[10]has noted that umbilical cord defects can cause complete obstruction of venous return and lead to fetal death. Hypothetically, if the cord problems are sublethal and persisting, the resulting fetal hypoxia may stimulate placental remodeling such as hypervascularity (chorangiosis and chorioangiomas), increased erythropoiesis (excessive number of nucleated RBCs), and stromal hyperplasia (dysplastic villi).

The pathogenesis of Placental mesenchymal dysplasia is unclear. Ultrasonographic findings are confused with a molar pregnancy because of hypoechoic spaces in the placenta in the presence of an apparently normal fetus, a fetus with growth restriction, or a fetus with features of overgrowth.Although Placental mesenchymal dysplasia may mimic partial molar pregnancy sonographically and by gross examination, a key difference between partial molar and normal placentas is their genetic composition. Partial hydatidiform moles are 70% to 80% triploid, often with 2 sets of paternal genes and 1 of maternal genes as a result of dispermyfertilization[20]. In PMD, apparently normal karyotypes often were found.Also, the important diagnostic features of PMD include the absence of trophoblastic proliferation, stromal trophoblastic inclusions, and scalloping of the villous surface, which are characteristics of a molar pregnancy[21].

Spontaneous abortion with hydropic change may have vesicle formation and can be confused with early Placental mesenchymal dysplasia. The vesicles in hydropic spontaneous abortion, if present, are usually small and are not diffuse. Second, the histology of spontaneous abortions shows degenerative changes without the classic histopathologic features of PMD.[22]

CONCLUSION

PMD is a rare and clinically significant lesion with high rates of IUGR, IUFD, and neonatal death. Promising results can be expected from molecular and genetic studies. Awareness of this entityits sonographic similarity to partial hydatidiform mole, unique pathologic features, and high rate of IUFDis important for prenatal counselling and monitoring. Given its association with IUGR, close postnatal monitoring also is warranted. Although a definitive association between acquired or inherited thrombophilia and PMD has not been identified, evaluation for such conditions may prove beneficial. The cooperation among pathologist, radiologist, neonatologist, and obstetrician will facilitate diagnosis.

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