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Case Report

Placental mesenchymal dysplasia- A case report

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ABSTRACT

Placental mesenchymal dysplasia is an underdiagnosed and a rare vascular anomaly of the placenta. It is characterized by the presence of grape-like vesicles, which, on ultrasonography gives the appearance of a partial mole. Histologically it can be differentiated by partial mole by the absence of trophoblastic proliferation. It is essential to make a correct diagnosis as the management and outcome of both these entities are vastly different. We present this case of placental mesenchymal dysplasia, not just because of its rarity but to highlight its radiological, gross and histopathological features to keep it in mind as a differential diagnosis and in making the correct diagnosis.

1. Introduction

Placental mesenchymal dysplasia is an underdiagnosed, unrecognized, rare vascular anomaly of the placenta.[1] It has been reported to have an incidence of only 0.02%.[2] The true incidence of this entity is largely unknown because it often goes misdiagnosed. It is characterized by the presence of grape-like vesicles, which, on ultrasound give the appearance of a partial mole.[1] It is usually accompanied by the presence of a foetus that may or may not have karyotypic anomalies. Histologically it can be differentiated by partial mole by the absence of trophoblastic proliferation.[3] Since the management and outcome of both these entities are vastly different it is important to make a correct diagnosis. Fewer than 130 cases of this disease have been reported in the literature.[4]

2. Case Report

A 27-year-old Gravida 3, presented for a dating scan with 2 months of amenorrhea and pregnancy confirmed by a urine pregnancy test. Ultrasound revealed a single live foetus with an approximate period of gestation of 8 weeks and 3 days. There was also a 5 ml collection of fluid along the inferior part of the gestation sac which was suggestive of a subchorionic haematoma. The patient was advised to follow up. Level 1 scan revealed a single live foetus with a gestational age of approximately 13 weeks and 2 days with mild molar changes in the placenta. Following this patient was referred to our centre where an ultrasound showed a thickened placenta with multiple cystic spaces. Beta HCG was advised which came out to be normal for the period of gestation. MRI showed a bulky uterus with the foetus. The placenta appeared heterogeneous and the majority appeared to be replaced by numerous tiny variably sized altered signal intensity cystic spaces which were hypointense on T1W and hypointense on T2W and a diagnosis of placental anomaly suggesting H mole was given. Medical termination of pregnancy was performed at the patient's request and the

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placenta was sent for histopathology.

A placenta measuring 13.5 cm x 8 cm x 4 cm and weighing 180 gm was received for histopathology. The cord was attached centrally and measured 11 cm. Membranes were intact and the maternal surface appeared normal. On the cut, the parenchyma shows patchy, cystically enlarged villi, along with grossly normal-appearing villous tissue. Few dilated tortuous vessels were also seen on the foetal side. The cord on the cut showed three vessels. Microscopy revealed numerous dilated and enlarged villi along with a few normal-looking villi. Few of the dilated villi showed central cistern formation and myxomatous areas.(Figure 1 a,b) Also seen were numerous thick-walled vessels which showed fibromuscular hyperplasia, some of which were dilated and occluded by thrombus with varying degrees of luminal obliteration. An overgrowth of fibrous areas was identified. (Figure 1 c.d) Trophoblastic proliferation was conspicuously absent. On immunohistochemistry, the abnormal-looking villi stained strongly positive for vimentin, and weakly for Desmin. (Figure 2 a.b) Staining for smooth muscle actin was absent in these abnormal villi while it was present in the normal-appearing villi. Alcian blue staining was performed and it was positive in the ground substance. Based on radiological and pathological characteristics the diagnosis of placental mesenchymal dysplasia was confirmed. The patient had an uneventful hospital stay following termination and beta HCG returned to normal within 2 weeks.

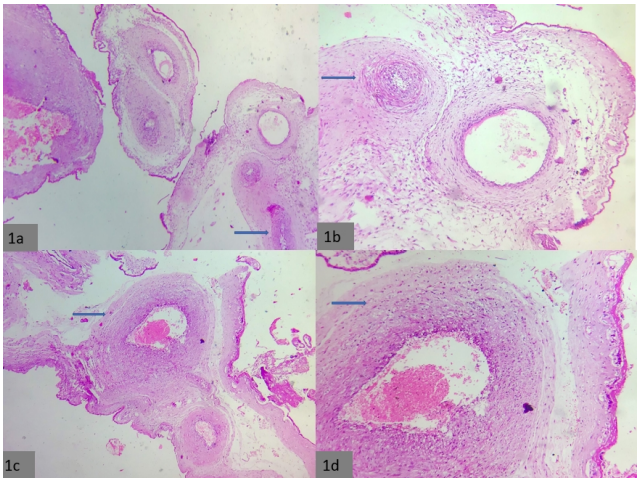

Fig. 1: a,b): Stem villous is markedly enlarged and hydropic with myxoid stroma and development of a centralcistern along with thick-walled vessels (arrows). H and E stain,100 X,100X; **c,d**): The large villous shows thick-walled vessels (arrows) which distinguish it from other molar gestations. There is no evidence of trophoblastic hyperplasia seen. H and e stain,100x 400x

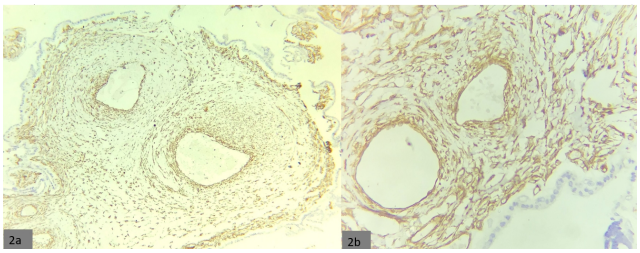

Fig. 2: The large villous shows thick-walled vessels an the surrounding stroma shows positivity for Vimentin and desmin, 100X

3. Discussion

Placental mesenchymal dysplasia (PMD) was first described by Moscos in 1991 as stem villous hyperplasia with raised alpha-fetoprotein and resemblance to the partial mole on ultrasound.[3] The low incidence may be attributed to misdiagnosis of the disease and the majority number of cases has been diagnosed after pathology examination. Our case was initially diagnosed as partial H- mole and finally correctly diagnosed by histopathological examination as placental mesenchymal dysplasia. This highlights the importance of subtle radiological clues to keep as differential diagnoses of this disease, although a definite diagnosis is made only after microscopic histological examination. The differentials to be kept for this entity include Partial hydatidiform mole, chorangioma, intervillous hematoma, non-specific hydropic changes, infections, etc.[1] Neyari et al. found that most common radiological features are enlarged and cystic placenta with dilated chorionic vessels.[5] In this case, for the diagnosis, however, ultrasound is sufficient in the past. PHM is associated with increased beta HCG levels and a triploid fetus and, can be excluded by a genetic workup. Color Doppler can be used and PMD shows a stained-glass appearance suggesting abundant blood flow in PMD while CHM shows little to no blood flow.[4] Recently MRI is used as an instrumental modality to distinguish CHM with twin co-existent fetuses from PMD.[6] As it may help in the location of the cystic placental tissue within or outside the fetal “sac” and CHM outside and inside in PMD within), and aid in the distinction of these two entities 9 PMD from molar gestation can be differentiated by the lack of trophoblastic hyperplasia in the PMD and normal level of maternal serum bHCG level, except elevations in maternal serum alpha-fetoprotein (AFP).

The exact cause of PMD is not yet known. However, it is postulated that congenital malformation of mesoderm and/or androgenetic mosaicism with endoreduplication of the paternal genome might be responsible.[7]It is also seen to be associated with Beckwith Weidmann syndrome in around 23% of cases and molecular disruption of chromosome 11.[8] In our case, no anomaly was found in

the foetus on an ultrasound. PMD is associated with the hypertensive disease during pregnancy including gestational hypertension, eclampsia, and HELLP syndrome.[5] In this case patient had an uneventful history. PMD is also related to adverse outcomes in the foetus. In an analysis of 61 cases of PMD, it was found to lead to IUGR (33%), IUFD (13%), and preterm delivery (52%).[5]There have been a few case reports of favourable outcomes as well, but this requires prompt diagnosis, knowledge of the disease and facility to handle pre term births and infections.[9]Gross examination generally has been found to reveal a large placenta with numerous dilated cystic spaces which are more towards the foetal side. The maternal side appears to be normal.[10] In third-trimester pregnancies the blood vessels are seen to be grossly dilated with the presence of tortuosity.[1]

Microscopically PMD is seen to have violated villi with central cisterns and fibrous overgrowth and peripheral thickened vessels. The dilation of the stem villi and trophoblastic proliferation are not seen.[1] The dilated villi stain for vimentin and Desmin, are negative for smooth muscle actin. This signifies a diminished differentiation of the stroma. The fibroblasts in the dilated villi fail to acquire myofibroblastic characteristics. Villi are also seen to stain positive for Alcian blue which indicates the presence of large amounts of mucopolysaccharides.[1] Cytogenetic studies have also been found to be useful in these cases but the majority of the cases are diploid. These tests do not help in distinguishing between complete moles or spontaneous abortions with hydropic change.[1] Cytogenetic and molecular testing was not done in the present case.

4. Conclusion

Placental mesenchymal dysplasia should be kept in mind as a differential diagnosis upon encountering an enlarged cystic placenta. Differentiation from molar pregnancy is also important and may be aided by histopathology and beta HCG levels. The role of ultrasound in guiding the diagnosis cannot be ignored. Presently this condition is largely underdiagnosed since the features are not very well recognised by pathologists and radiologists. A correct diagnosis requires a careful evaluation of the radiological findings, correlation with beta HCG and serum alpha-fetoprotein levels and other routine investigations so that a diagnosis can be made in the early gestation period and the decision about termination can be made soon.

5. Source of Funding

None.

6. Conflict of Interest

None.

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