Multiple chorangiomas of placenta: a case report

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Introduction

Chorangioma is a benign villous capillary lesion of the placenta. It involves the large stem villi and is characterized by a nodular proliferation of capillaries and intervening perivascular stroma surrounded by trophoblasts. Most chorangiomas are small and clinically incidental. Large lesions (>4-5 cm in size) can result in significant clinical complications. Here we describe a case of extensive placental involvement by multiple chorangiomas associated with polyhydramnios and foetal hydrops diagnosed on pathological examination following emergency Caesarean section.

Case report

A 38 year old woman in her second pregnancy attended antenatal clinic at our institution at 33 weeks' gestation. She had undergone an ultrasound scan by a private provider which reportedly showed polyhydramnios. Her past medical history included a clinically stable prolactinoma treated with bromocriptine, which was discontinued at 6 weeks' gestation. Significant obstetric history included recurrent gestational diabetes mellitus. There were no significant findings on antenatal blood tests or foetal anomaly scan.

Foetal cardiotography at presentation showed recurrent decelerations with reduced variability. The patient was admitted to the labour ward for monitoring. Ultrasound assessment showed severe polyhydramnios, foetal cardiomegaly and diffuse cystic changes of the placenta.

Emergency lower segment Caesarean section was performed at 33 weeks and 5 days' gestation and delivered a female baby with a birth weight of 1740g. The Apgar score was 1 at 1 minute, 5 at 5 minutes and 6 at 10 and 20 minutes after birth. Umbilical cord blood gas showed metabolic acidosis with pH 7.08 and BE -10. A large amount of liquor was noted at operation. The placenta and umbilical cord were fragile and delivered piecemeal with the placental disc involved by abundant friable lesional tissue.

Pathological findings

Macroscopic examination of the placenta after formalin fixation found a singleton placenta in a fragmented state. The specimen weighted 626 g. The umbilical cord measured 27 cm in length and the placental disc tissue measured 23 x 17 x 2.5 cm in aggregate. The completeness of the cotyledons could not be ascertained. The maternal surface of the placenta was diffusely involved by multiple discrete nodular lesions ranging from several millimetres to several centimetres in size. These nodules were well-demarcated from the adjacent normal-looking placental parenchyma and had uniform firm whitish cut surfaces.

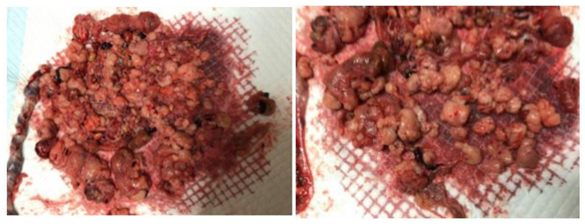

Figure 1

Figure 2

Figures 1 and 2: Macroscopic appearance of the placenta in the fresh state following emergency Caesarean section

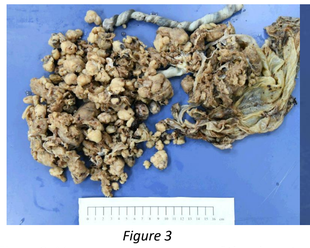

Figure 3

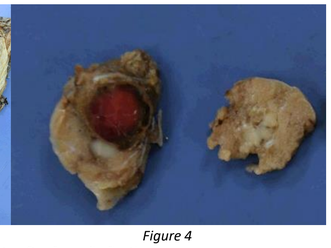

Figure 4

Figure 3: Macroscopic appearance of the placenta after formalin fixation
Figure 4: Smooth interface of nodular lesion with normal-looking placental tissue

Microscopic examination of the placenta showed multiple well-circumscribed vascular lesions each rimmed by a layer of attenuated trophoblasts. The vascular lesions were composed of multiple closely packed small capillaries and infrequent medium-sized muscularized blood vessels. The capillaries were lined by attenuated to plump endothelial cells with minimal cytological atypia. A few areas showed occasional mitotic activity. The capillaries were surrounded by a fibroblastic stroma which showed variable oedema, hyalinization and karyorrhexis. No atypical trophoblastic proliferation or malignancy was identified.

Immunohistochemical staining with CD31 highlighted the endothelial cells which were surrounded by a layer of pericytes immunoreactive to actin.

The final pathological diagnosis was that of multiple chorangiomas.

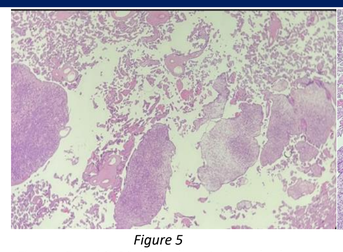

Figure 5

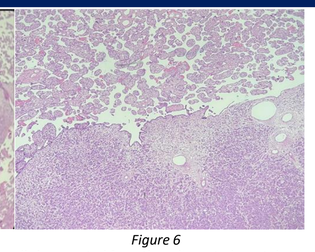

Figure 6

Figures 5 and 6: Multiple discrete vascular lesions well-demarcated from the adjacent placental tissue

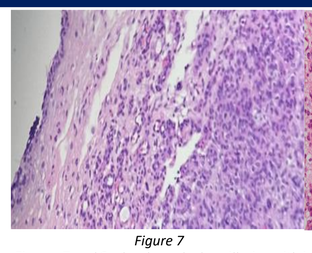

Figure 7

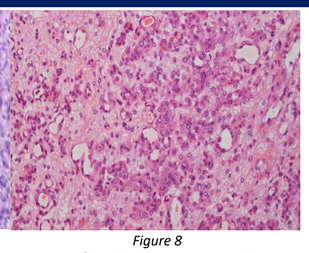

Figure 8

Figures 7 and 8: closely packed capillaries with intervening fibroblastic stroma rimmed by trophoblasts

Clinical outcome

Physical examination of the neonate found marked peripheral oedema consistent with hydrops foetalis. Initial blood tests showed mild anaemia (Hb 11.2 g/dl, reference range 13.5-22.5 g/dl) and thrombocytopenia (platelet count 46 x 10^9/L, reference range 150-500 x 10^9/L). The blood film showed no reticulocytosis. The baby responded well to supportive treatment including blood product and inotrope infusions and was discharged from hospital on Day 34.

Discussion

A classification of placental lesions[1] incorporating the 2014 Amsterdam Placental Workshop Group Consensus Statement[2] places chorangiomas within a subgroup of foetal stromal-vascular lesions. A retrospective study of over 20,000 placenta specimens[3] found an incidence of 0.61%. Large chorangiomas (>4-5 cm in diameter), have a prevalence of 1 in 9000 to 1 in 15000 and are associated with significant clinical complications. These include foetal anemia, polyhydramnios, hydrops and growth restriction[4]. A systematic review[5] reported increased rates of intrauterine death (8.2%), low birth weight (24%) and pre-term delivery (34.1%). There was a high incidence of perinatal death (40.5%) in cases presenting with foetal hydrops.

Chorangiomas may be hamartomatous lesions or result from reactive angiogenesis due to hypoxia. In support of the latter hypothesis, it has been noted that most clinically significant chorangiomas present in the early third trimester. One theory proposes that trapping of red blood cells and platelets within the proliferating capillaries during this period of villous maturation results in haemolysis, anaemia and foetal oxygen deprivation, leading to cardiac failure in the foetus.

Our case presented with features consistent with clinically significant chorangioma. The mother presented in the early third trimester with polyhydramnios. There were other features compatible with cardiac failure in the foetus, namely cardiomegaly and hydrops. Cardiotography and umbilical cord blood gas reflected associated physiological compromise. The neonate had anaemia and thrombocytopenia and a birth weight in the 10th percentile for gestational age, in keeping with intrauterine growth restriction.

Another notable feature in this case was the diffuse involvement of the placental disc by multiple discrete chorangiomas, suggestive of the rarely described phenomenon of multiple chorangioma syndrome. A recent review published in 2021 found 13 cases described in the literature[6]. Of these, only 5 of 13 newborns survived and 4 of 13 cases were recurrences.

In conclusion, clinically significant chorangiomas are relatively uncommon but may result in serious foetal and neonatal complications. They present late in pregnancy and may not be recognized before birth. In our case, pathological examination of the placenta after delivery was essential to the diagnosis.

References

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Acknowledgements

Dr Wai-Lam Chan, Department of Obstetrics and Gynaecology, Kwong Wah Hospital, for providing the clinical information in this case report.