Case Report

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Giant Chorioangioma with Extraplacental Presentation: A Rare Case Report

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ABSTRACT

Chorioangiomas are non-trophoblastic placental tumours, commonly located near the cord insertion and are characterized by excessive vascular proliferation within the chorionic villi. Small chorioangiomas are common but giant chorioangiomas are quite rare and present with complications like preterm delivery, polyhydramnios, hydrops, growth restriction, thrombocytopenia and fetal death. The pathogenesis of various feto-maternal complications can be attributed to this indolent entity. So, early diagnosis, close fetal monitoring and timely intervention are crucial to prevent feto-maternal morbidity and mortality. Here, we report a case of giant chorioangioma that led to intrauterine fetal death.

Keywords: *Chorioangioma, Extraplacental, Non-Trophoblastic Placental Tumours, Intra-Uterine Fetal Death*

Introduction

Chorioangiomas are non-trophoblastic placental tumours, commonly located near the site of the umbilical cord insertion and are characterized by excessive vascular proliferation within the chorionic villi. Although, small intra-placental chorioangiomas are common accounting for approximately 1% of all pregnancies, giant extraplacental chorioangiomas are quite rare with only few cases reported in literature.[1] These giant chorioangiomas have adverse effects on both, the mother and the fetus, leading to complications like preterm labour, polyhydramnios, hydrops, fetal growth restriction, fetal anemia, thrombocytopenia and fetal death.[1,2] Here, we report a case of giant extraplacental chorioangioma that led to intrauterine fetal death.

Case Report

A 28years old, G4P3L3, presented at 31 weeks of gestation with labour pain and passage of clots. She delivered a stillborn female fetus. The patient was an unregistered case and had no previous ante natal check-ups.

We received a female fetus weighing 1700gm for fetal autopsy, along with a placenta measuring 22x18x4cm, weighing 900gm and, a separate irregular mass measuring 15x11x4cm, weighing 500gm. On gross examination of the fetus, petechiae (Fig.1) were noted all over the body. Rest of the autopsy findings were normal. The placenta appeared intact and complete with attached membranes. The external surface of the irregular mass showed areas of congestion with the cut surface appearing solid, grey brown and homogenous (Fig 2A & B).

On histopathological examination, fetal organs and umbilical cord showed normal histology. Placenta and membranes showed normal histology with focal calcification (Fig 3A & B). Microscopic examination of the mass revealed many thin walled capillaries and small blood vessels lined by plump endothelial cells along with focal areas of calcification, hemorrhage and hemosiderin pigment deposition (Fig 4A & B). Based on these findings a diagnosis of extraplacental chorioangioma was made.

Discussion

Chorioangiomas are the most common tumours of the placenta, with the majority being small, encapsulated and intra-placental, and are likely to be missed during gross pathological examination, unless the placenta is carefully sectioned.[3] An increased incidence of chorioangioma is associated with maternal age, diabetes, female sex of the fetus, premature labour and multiple deliveries.[4] Although small chorioangiomas are common, giant chorioangiomas are rare with a prevalence of one in 9000 to one in 50000 pregnancies which are arbitrarily defined as measuring more than 5 cm in diameter.[3] These chorioangiomas are

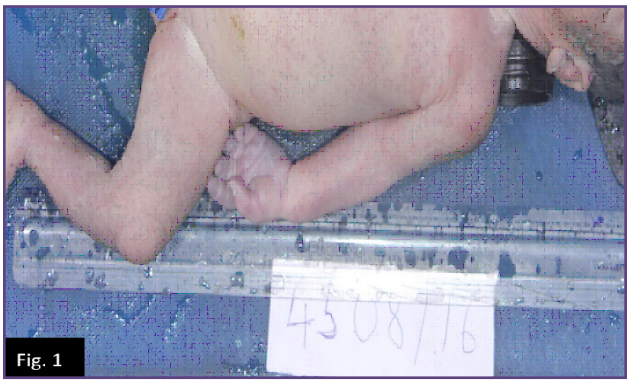

Fig. 1: Shows petechiae all over the body on gross examination.

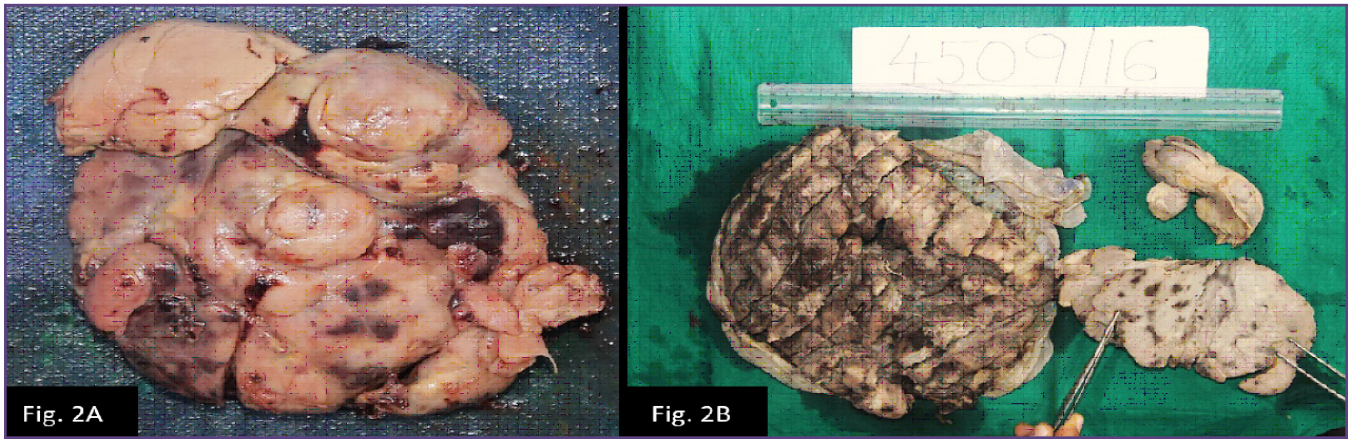

Fig. 2: (A) Shows an irregular mass separate from the placenta with the external surface showing areas of congestion. (B) Depicts the cut surface of the mass after fixation which appears solid, grey-brown compared to the maternal surface of the placenta.

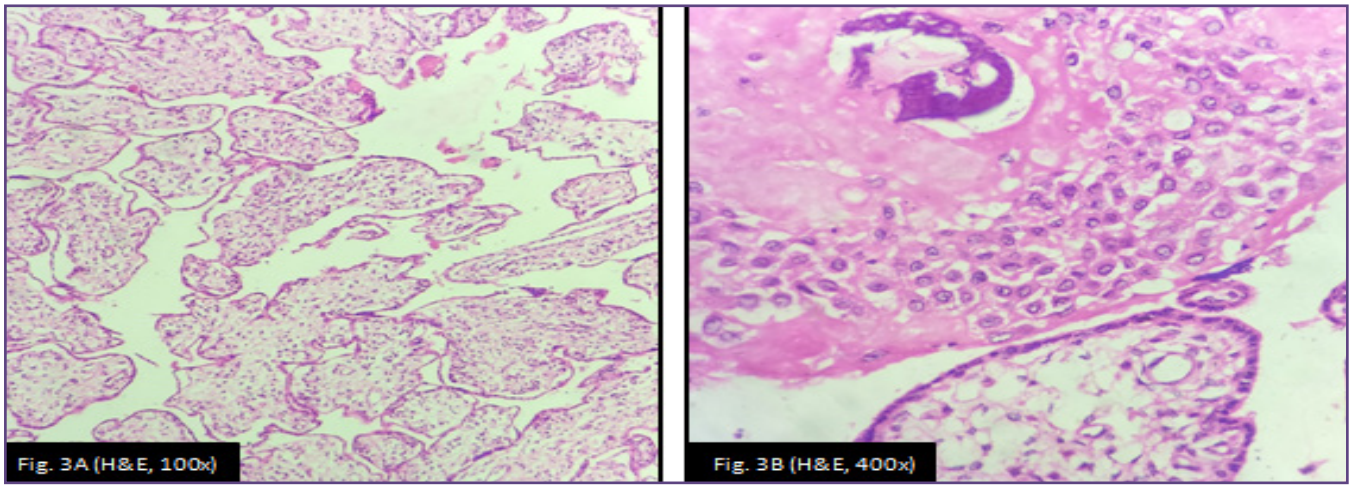

Fig. 3: (A) depicts the normal placental tissue showing chorionic villi lined by cytotrophoblast and synctiotrophoblast layer and a primitive mesenchymal core. (B) shows focal calcification in the placenta.

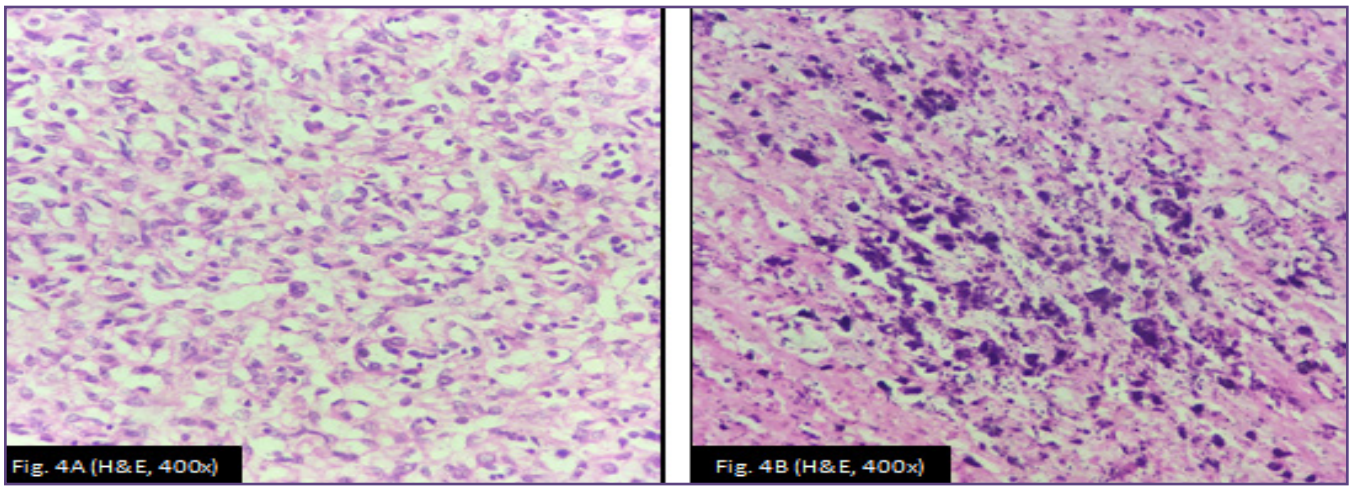

Fig 4: (A) depicts the microscopic appearance of the mass showing many thin walled capillaries and small blood vessels lined by plump endothelial cells in a scant connective tissue stroma. (B) shows focal areas of calcification within the tumour.

the ones associated with a number of fetal complications and thus have a high perinatal death rate.[5,6]

Chorioangiomas can grossly be confused with degenerated myoma, subamniotic hematoma, placental teratoma and placental infarction. [7,8] So, a thorough histopathological examination and clinico-radiological correlation is necessary for identifying chorioangiomas.

Microscopically, there are three histological patterns of chorioangioma: angiomatous, cellular and degenerative type. The most common type is angiomatous, which is composed of numerous proliferative blood vessels, surrounded by placental stroma. The tumour cells show immunoreactivity for CD31, CD34, factor VIII, GLUT-1 and Cytokeratin-18 which suggests an origin from the blood vessels of chorionic plate and anchoring villi. [9,10] In the present case, angiomatous type of the tumour was noted along with features of degeneration.

Ultra-sonographic findings of chorioangioma include a well-defined, complex, echogenic mass protruding into the amniotic cavity which appears different from rest of the placenta and is usually situated near the insertion of the placenta. Doppler helps to differentiate placental teratoma, leiomyoma, blood clot and chorioangioma. [7] In the present case, the patient gave no history of prior antenatal checkups or an ultrasonogram.

A variety of complications such as polyhydramnios, premature delivery, premature placental separation and placenta previa are commonly associated with large chorioangiomas. Impaired fetal circulation is noted in large chorioangiomas which is due to arteriovenous shunts that cause an increase in the venous return to the heart, resulting in tachycardia, cardiomegaly, hypervolemia and congestive cardiac failure. Anemia and thrombocytopenia may also be seen in the neonate. [9] One possible mechanism for these complications can be attributed to microangiopathic hemolysis. [3] Chorioangiomas show high expression of angiopoietin-1 & 2 leading to numerus, narrow, tortuous and partially thrombosed blood vessels which can cause erythrocyte injury and trap platelets resulting in hemolytic anemia and thrombocytopenia.[2,11] In the present case, petechiae noted on gross examination of the fetus, could be attributed to fetal thrombocytopenia.

Treatment modalities available for chorioangiomas are endoscopic surgical devascularization, alcoholic ablation, interstitial laser coagulation, endoscopic suture with bipolar electrosurgery, microcoil and enbucrilate embolization. [3,9]

Conclusion

Due to the high fetal mortality rate associated with large chorioangiomas, early diagnosis is necessary so that fetal surveillance can be instituted. Examination of placenta is crucial in complicated pregnancies as the pathogenesis of various feto-maternal complications can be attributed to this tumour. So, in all such cases, a differential diagnosis of Chorioangioma should be considered.

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