ABSTRACT

Background: Chorangiomas are the most common, benign placental tumors. The size of these tumors has an impact on the clinical outcome. Giant chorangiomas, more than 5 cm in diameter, are rare tumors, and are frequently associated with a variety of gestational complications and a poor perinatal outcome.

Case report: We report the case of a 26-year-old pregnant female, who presented with sonography findings of an extraplacental mass and polyhydramnios. She was taken up for emergency cesarean section in view of anteptum hemorrhage and intrauterine fetal death (IUFD). The placenta with an attached circumscribed, lobulated, firm, extraplacental mass was sent for histopathological evaluation. On microscopy, the mass was diagnosed as a myxoid degenerate chorangioma.

Conclusion: Giant chorangiomas are associated with an unfavorable prognosis. Timely antenatal diagnosis, monitoring, and management of associated complications are quintessential to ensure a favorable outcome.

Keywords: Intrauterine fetal death, Myxoid chorangioma, Placenta, Polyhydramnios.


INTRODUCTION

Chorangiomas are the commonest benign placental tumors, accounting for approximately 0.6 to 1% of all pregnancies. They arise from the nontrophoblastic primitive chorionic mesenchyme. Giant chorangiomas, more than 5 cm in diameter, are rare tumors, with prevalence ranging from 1:9,000 to 1:50,000 of placentas. They are frequently associated with a variety of fetomaternal complications. The overall fetal mortality rate of giant chorangiomas is 30%.

CASE REPORT

A 26-year-old female, second gravida with one living issue, was referred to us at 26 weeks of gestation, with sonographic findings of a chorangioma cyst. On review ultrasound, a single fetus with normal biparietal diameter was noted. There was no evidence of obvious anomalies, intrauterine growth retardation (IUGR), hydropic changes, or cardiac failure. The placenta was low lying and there was severe polyhydramnios (amniotic fluid index of 50). On gray scale, an isoechoic mass was seen over the anterior surface of the placenta, protruding into the amniotic cavity (Fig. 1A). On color Doppler, the mass showed vascularity and the pulse rate in the vascular channels of the mass was the same as the fetal heart rate. The patient was taken up for emergency cesarean section during the same period, in view of anteptum hemorrhage, with the outcome of a still-born female fetus. The placenta, with an attached extraplacental mass, was sent for histopathological evaluation.

Grossly, the specimen weighed 550 gm. The placental disk measured 16 × 16 × 2.5 cm. Attached umbilical cord and membranes were unremarkable. A lobulated, firm mass weighing 60 gm and measuring 6 × 5.5 × 3.5 cm was attached to the fetal surface of the placenta via a pedicle (Fig. 1B). Cut surface of the mass revealed firm gray-white to spongy tan-red areas (Fig. 1C). Histopathological examination of the mass confirmed the diagnosis of myxoid degenerate chorangioma. The lesion had a trophoblastic lining (Fig. 2A) and was composed of diffuse proliferation of predominantly capillary sized vessels, along with a few large caliber muscular vessels embedded in a loose, myxoid stroma (Figs 2B and C). Focal areas of hyalinization, calcification (Fig. 2D), and neutrophilic infiltrate were seen. Sections from the pedicle confirmed two blood vessels, without Wharton’s jelly-like parenchyma. Microscopy of the main placental disk revealed late second trimester villi, many of which were edematous, with increased Hofbauer cells (Figs 2E and F).
DISCUSSION

Chorangiomas, or placental hemangiomas, are the most common benign, placental tumors, characterized by excessive vascular proliferation, arising from the non-trophoblastic primitive chorionic mesenchyme. Most authors classify chorangioma as neoplasms; however, few consider them as placental hamartomas. 4,5 They are often small, inconsequential, and diagnosed incidentally on antenatal sonography or postpartum, by microscopy. Minute and single tumors may be missed if meticulous sectioning of the placenta is not performed, suggesting that the true prevalence is probably unknown. 5 Giant chorangiomas are frequently associated with a variety of fetomaternal complications. Maternal complications include pregnancy-induced hypertension, preeclampsia, antepartum hemorrhage, and preterm labor. Fetal complications include anemia, thrombocytopenia, cardiomegaly, heart failure, polyhydramnios, nonimmune hydrops fetalis, IUGR, IUFD, congenital anomalies, and chromosomal abnormalities. 6 Our case was associated with polyhydramnios, abruptio placenta, and IUFD. Chorangiomas act as a physiological dead space, resulting in chronic hypoxia leading to IUGR and IUFD. 7 Polyhydramnios may be caused by fetal heart failure, transudation of fluid from the large surface area of the chorangioma, transudation due to mechanical obstruction of blood flow due to large chorangiomas located close to the cord insertion, or shunting of blood to the tumor vascular bed, leading to vascular insufficiency. 4,7 Antepartum hemorrhage may be due to premature separation of the placenta or rupture of the vascular pedicle. 6,8 Chorangiomas associated with polyhydramnios lead to high perinatal morbidity and mortality. 1,2

Ultrasonography (USG), Doppler, and magnetic resonance imaging (MRI) are often helpful in prenatal diagnosis and follow-up. Gray-scale USG typically shows a single or multiple, complex echogenic mass, different from the rest of the placenta. It usually protrudes into the amniotic cavity, near the cord insertion. On color Doppler, the chorangial vascular channels are in continuity with fetal circulation, thus distinguishing it from sonographically similar lesions, such as placental teratoma, placental hematoma, partial mole, deceased twin, and degenerated myoma. 1,6 T2 images
of MRI are similar to hemangioma, and thereby aid in diagnosis. Our case showed a vascular mass embedded in the placenta, with pulse rate of the tumoral vascular channels being same as the fetal heart rate.

Microscopically, chorangiomas are divided into three main subtypes: angiomatous, cellular, and degenerative. The angiomatous pattern is the commonest, characterized by abundant capillaries and blood vessels surrounded by placental stroma. The cellular pattern has abundant endothelial cells within the stroma. The degenerate pattern is characterized by calcification, necrosis, or hyalinization. Chorangiomas do not have malignant potential. Histopathological differentials include chorangiosis, chorangiomatosis, and
Chorangiocarcinomas (misnomer; chorangioma associated with trophoblastic proliferation). Chorangiosis is diagnosed by the presence of ten or more vascular channels per villi, in at least ten terminal villi, in ten or more areas of three or more random, noninfarcted areas, under low-power magnification. Chorangiomatosis is characterized by a nonexpansile vascular proliferation in otherwise normal stem villi. Atypical chorangiomas are characterized by mitotic count more than 7 per 10 high-power fields, cytological atypia, and necrosis. 

Our case showed a chorangioma with myxoid degeneration, which is a very rare form of the degenerate subtype. 

Management options comprise of serial fetal transfusions through cordocentesis, fetoscopic laser coagulation of vessels supplying the tumor, endoscopic surgical devascularization with use of suture ligation or bipolar cautery, and absolute alcohol chemosclerosis. Therapeutic amniocentesis and maternal indomethacin therapy are carried out for polyhydramnios. Steroid administration is indicated before 34 weeks for accelerating fetal lung maturity. 

**CONCLUSION**

Giant chorangiomas are associated with significant fetomaternal morbidity. They carry a grim prognosis, including fetal death. Timely antenatal radiological diagnosis, monitoring, and management, including that of complications, are quintessential to ensure a favorable outcome.

**REFERENCES**