

Chorioangioma: An Unpredictable Riddle

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Abstract

Chorioangioma is a non-malignant vascular placental tumor, appears as a well-defined mass contained within the placenta or on its fetal surface. On color doppler scanning variable amount of blood flow is seen. Chorioangiomas >2 cm in diameter with arteriovenous shunting may result in fetal complications like fetal congestive heart failure, hydrops, hemolytic anemia, fetal thrombocytopenia, cardiomegaly, growth restriction and maternal complications like preeclampsia, preterm labour, placental abruption, and polyhydramnios. Prenatal intervention options are ultrasound guided laser coagulation of vessels within the tumor, fetal blood transfusions in case of fetal anemia and amnio-drainage for symptomatic polyhydramnios. Chorioangiomas >2 cm in diameter with evidence of intralesional arterial flow requires frequent biophysical testing and serial evaluation of fetal growth, cardiac function, and amniotic fluid to identify a fetus at risk and requiring early delivery to avoid increased peri natal morbidity or mortality risks. We report a case of large chorioangioma referred to us at 31⁺⁵ weeks of gestation with raised MCA PSV doppler and severe polyhydramnios. Close surveillance was done for maternal-fetal wellbeing. Counselling done for available options of treatment and conservative approach versus early delivery. The timely intervention resulted in good maternal and perinatal outcome.

Keywords: Chorioangioma, Polyhydramnios, Fetal anemia, MCA- PSV doppler

Introduction

Chorioangioma is a non-malignant vascular placental tumor, appears as a well-defined mass contained within the placenta or on its fetal surface. Chorioangiomas (chorioangiomas) occur in 1 of every 3500–9000 pregnancies.¹ Placental chorioangioma is the most common benign tumor of the placenta.²

It was first described by Clarke in the year 1978.³ They were more seen in multiple pregnancies and in female babies. Chorioangioma is believed to arise by 16th day of fertilization, although there is no documentation of tumor in first trimester.⁴

It consists of a benign angioma arising from chorionic tissue. Three histological patterns of chorioangiomas have been described by Marchetti⁵: angiomatous, cellular, and degenerate. The angiomatous is the most common, with numerous small areas of endothelial tissue, capillaries, and blood vessels surrounded by placental stroma.⁵

On ultrasonography it appears as hypo- or hyperechoic, well-circumscribed mass, which is usually located underneath the chorionic plate near the umbilical cord insertion, and often protrudes into the amniotic cavity. Color Doppler demonstrates large vascular channels around and within the tumor.⁶

Chorioangiomas >2 cm in diameter with arteriovenous shunting may result in fetal complications like fetal congestive heart failure, hydrops, hemolytic anemia, fetal thrombocytopenia, cardiomegaly, growth

restriction and maternal complications like preeclampsia, preterm labour, placental abruption, and polyhydramnios.¹

Fetal anemia and thrombocytopenia occur due to sequestration of red blood cells and platelets by the tumor, and fetal heart failure, hydrops placentomegaly occur due to hyperdynamic circulation as a result of arteriovenous shunting. Polyhydramnios results due to direct transudation into the amniotic fluid and due to fetal polyuria, secondary to the hyperdynamic circulation and maternal mirror syndrome due to generalized fluid overload and preeclampsia.⁶

Chorioangiomas >2 cm in diameter with evidence of intralesional arterial flow requires frequent biophysical testing and serial evaluation of fetal growth, cardiac function, and amniotic fluid to identify fetus at risk and requiring early delivery to avoid increased peri natal morbidity or mortality risks.⁶

We report a case of large chorioangioma referred to us at 31⁺⁵ weeks of gestation with raised MCA PSV doppler and severe polyhydramnios.

Case Report

A 31-year-old, G5P2L2A2, literate patient, from an average socioeconomic background, in a non-consanguineous marriage for 13 years, referred at 31+6 weeks gestation for polyhydramnios with large placental chorioangioma with hyperdynamic fetal circulation (MCA PSV>1.5 MOM) and suspected fetal anemia.

Patient blood Group: O Negative, husband blood group- O positive. She had previous 2 uneventful full term vaginal delivery and received Inj. Anti D as per baby blood group. Patient had history of prior 2 abortions, history of D and E once and received Inj. Anti-D that time (once).

Patient conceived naturally each time, including present pregnancy. She booked at nearby centre. Pregnancy confirmed by ultrasonography and was diagnosed with diabetes and hypothyroidism. She was started on folic acid, thyroid supplements and Inj. Insulin both intermediate acting and long acting as per sugars. NT/NB scan done was normal. No complaints of spotting PV, bleeding PV, hyperemesis, fever with rashes in first trimester.

Quickening felt at 5th month of amenorrhea. Anomaly scan and fetal echo done at 20-21 weeks showed- no gross anomalies, posterior low-lying placenta and normal fetal echo. She was started on iron and calcium supplements.

She started perceiving fetal movements at 6-7 months of gestation. Growth scan done at 29-30 weeks showed single live intrauterine gestation of 29-30 weeks in cephalic presentation, well defined hypoechoic homogenous mass lesion measuring 6.1x5.7 Cm, seen in upper margin of placenta. Vascularity seen within lesion on color doppler study, suggestive of large placental chorioangioma. No obvious fetal defects noted. Fetal growth is on 95th centile, EFW-1830gms, FHR-148bpm, AFI-13 Cm, polyhydramnios, MCA doppler normal. Cervical length normal(3.1cm), no funneling. Indirect Coombs test done- negative.

Admitted at nearby private medical college, managed conservatively, received antenatal Anti-D prophylaxis and antenatal steroiding.

Patient developed epigastric discomfort and decreased urine output for one week. No history of any high BP recordings.

Repeat scan done after 2 weeks, showed single live intrauterine gestation of 31-32 weeks in breech presentation with increase in size of chorioangioma from 6.1x5.7cms to 8.2x7.0 Cm, fetal growth above 95th centile, EFW-2390gms, FHR-151bpm, AFI-39 Cm, gross polyhydramnios. Fetal MCA PSV >1.5 MoM, suggestive of fetal anemia, cervical length normal (3.1cm).

Patient was referred to our tertiary care center in view of large placental chorioangioma with raised MCA PSV doppler and severe polyhydramnios. Patient had complaint of difficulty in breathing for 2-3 days, gradually increasing day by day.

Patient admitted at our center and monitored for vitals, urine output, GRBS, FHR and uterine contractions. Repeat scan showed: single live intrauterine gestation of 31+6 weeks, 83x60 Cm chorioangioma, EFW-2063gms, FHR-151bpm, AFI-43 Cm, gross polyhydramnios. Fetal MCA PSV-71Cm/Sec (1.66 MoM) suggestive of hyperdynamic circulation (Figure 1 &2).

Couple explained regarding the risks and complications associated with patient's condition, need for close maternal and fetal monitoring.

Discussed regarding option of continuation of pregnancy with conservative management, amniocentesis for associated maternal discomfort versus early delivery in view of raised MCA-PSV and hyperdynamic fetal circulation leading to risk of fetal hydrops, cardiac failure and intrauterine death. Option of Cordocentesis and estimation of fetal hemoglobin followed by intrauterine transfusion if required in view of raised MCA -PSV and procedure related difficulties and complications with severe polyhydramnios discussed. Risks related to polyhydramnios, risk of PPROM, cord prolapse, fetal malpresentation, placental abruption, risk of maternal mirror syndrome- pre-eclampsia explained. Risk to baby related with prematurity, maternal diabetes, risk of respiratory distress, need for NICU care if required explained. Couple opted for early delivery. Discussed regarding option of vaginal delivery versus LSCS in view of preterm, polyhydramnios with malpresentation, risks involved with each explained in details. Couple opted for early delivery by LSCS. Risks during Cesarean section explained. Chances of heavy bleeding, risk of PPH and need for its management explained. Increased risks of maternal and perinatal morbidity, mortality explained.

Relevant blood investigations done. Rescue dose of steroids given, sugar monitoring done and Inj. Insulin dosage adjusted as per sugars. Injection Magnesium Sulfate given for fetal neuro-protection. Informed high risk consents obtained, blood and blood products arranged. Patient was taken for LSCS under spinal anesthesia. Intraoperative 5.5 L of amniotic fluid drained (Figure 3). Patient delivered female baby of birth weight of 2.32 Kg. APGAR at birth 7 and 8 at 1 minute and 5 minutes respectively. Placenta with membranes and the chorioangioma alongside it, was expelled spontaneously. Patient had no intraoperative complications.

On examination large placental mass seen near cord insertion site below amniotic membrane on fetal surface attached to main placental mass through communicating large vascular bundle (Figure 4). Placenta weighed 695 gm in total, with chorioangioma weighing 140 gm separately. Placenta with chorioangioma sent for HPE, confirmed chorioangioma histo-pathologically (Figure 5).

Relevant investigations done for baby at birth. Baby blood group-O negative, DCT- negative. Hemoglobin -15.8 gm%, suggested raised fetal MCA PSV >1.5 MOM was related to hyperdynamic fetal circulation related to large placental chorioangioma, not fetal anemia. Baby blood group-O negative, hence Inj. Anti-D not given. Post operatively patient monitoring continued and started on oral hypoglycemics for raised blood sugars. Further course in hospital for both mother and baby was uneventful, discharged in stable condition on postoperative day 5.

Discussion

Dong et al.⁸ examined 56 cases of placental chorioangioma (≥ 2.2 cm in diameter), the incidence of polyhydramnios, preeclampsia, fetal distress, preterm birth, fetal loss or induced abortion was 16.1%,

8.9%, 8.9%, 23.2%, and 10.7%, respectively.

In a systematic review and meta-analysis including twenty-eight studies and 161 pregnancies, the size of the mass and presence of fetal hydrops were found to be the main determinants of perinatal outcome in pregnancies complicated by chorioangioma.⁹

Three-dimensional color Doppler acquisition and placental MRI has recently been reported as adjunct to placental imaging, which can offer more information about the tumor and its vascular distribution.¹⁰ In the 10 years from August 2008 to December 2018 by Ma et al.⁷ 175 cases (0.17%) were identified as placental chorioangioma histologically and 44(0.04%) of them were large chorioangiomas. Nearly one-third of cases with large chorioangiomas were associated with severe maternal and fetal complications or required prenatal intervention. Although one-fifth of fetuses/newborns complicated with large chorioangiomas were lost perinatally, the long-term prognosis for surviving fetuses was generally good. In their study further statistical analysis revealed that tumor size and location affect prognosis.

Treatment of complications depends on gestational age and maternal-fetal symptoms.⁷

Termination of pregnancy is considered for worsening maternal condition, or late-onset complications with viable fetuses.⁷

If severe complications occur before the fetus is viable, prenatal intervention should be carried out to extend the gestation, which is classified as supportive and definitive treatment.⁷

The choice of supportive treatment is based on the specific type of complication, such as intrauterine transfusions for anemic fetuses and hydropic fetuses, amnioreduction and oral indomethacin for polyhydramnios with respiratory embarrassment and/or pre term labor, transplacental digoxin intervention for fetal cardiomegaly.⁷

Definitive treatment refers to the treatments to block arteriovenous shunting including alcohol injection, microcoil embolization, endoscopic laser coagulation, radiofrequency ablation, and interstitial laser therapy.⁷

To date, there is still no consensus on whether to choose supportive or definitive treatment, and whether to choose a single treatment modality or combined therapy for a given condition.⁷

We discussed regarding option of continuation of pregnancy with conservative management, amniocentesis for associated maternal discomfort versus early delivery in view of raised MCA-PSV and hyperdynamic fetal circulation leading to risk of fetal hydrops, cardiac failure and intrauterine death. Option of Cordocentesis and estimation of fetal hemoglobin followed by intrauterine transfusion if required in view of raised MCA -PSV and procedure related difficulties and complications with severe polyhydramnios discussed. As our case was approaching gestational age of 32 weeks, we proceeded with termination of pregnancy after discussion with couple.

Conclusion

As till date there is no single definitive treatment modality, individualized care and counselling after taking consideration of gestational age, size of chorioangioma, associated complications and patient preferences plays key role in deciding the option of treatment for a particular case. Our case signifies the importance of counselling according to available options at a particular setting.

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Figures:



Figure 1: Large Placental Chorioangioma (measuring 60x83mm) on Color Doppler showing vascular channels around and within the tumor.

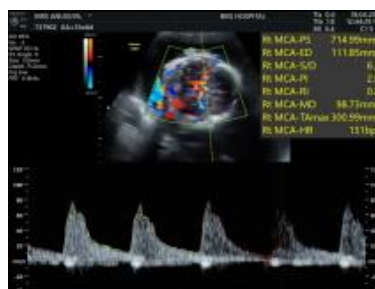


Figure 2: Fetal Middle Cerebral Artery Doppler showing hyperdynamic fetal circulation (MCA-PSV-71Cm/Sec- 1.66 MoM)

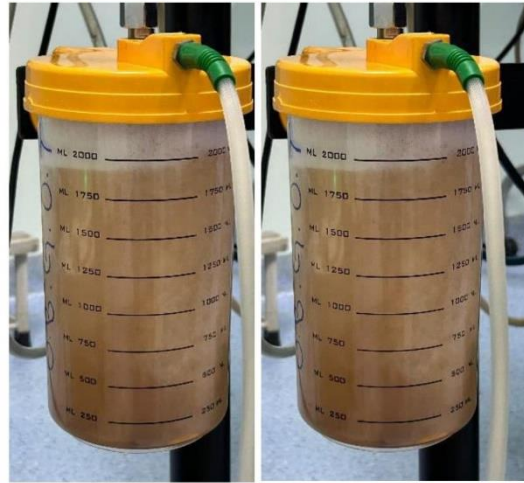


Figure 3: Intra-operative drainage of a total of 5.5 L amniotic fluid.



Figure 4: Placenta and membranes with large placental choriangioma: Large placental choriangioma seen attached to main placental mass through communicating large vascular bundle, near cord insertion site below amniotic membrane on fetal surface.

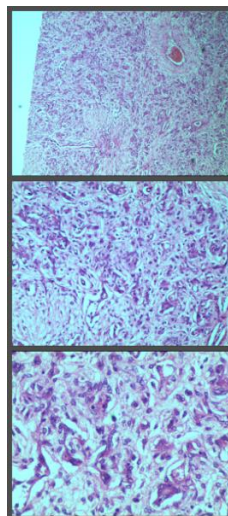


Figure 5: Histopathology of Placental choriangioma - Back-to-back arranged chorionic villi and trophoblast, with fibrovascular stroma and presence of prominent capillaries. a) 40x b) 100x c) 200x