Chorioangioma Syndrome: Co-relation of Color Doppler Ultrasound and Pregnancy Outcome
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Abstract

Large chorioangiomas are rare and associated with significant fetal and maternal risks. A case of chorioangioma syndrome is presented with polyhydramnios, pre-eclampsia, preterm labor and feto-maternal hemorrhage. Antenatal diagnosis is possible with ultrasound, and the prognosis for pregnancy outcome can be predicted by evaluating the vascularity of the tumor on color doppler sonography.

Key Words
Placental chorioangioma, Chorioangioma syndrome, Hydramnios, Color Doppler

Introduction

Although chorioangiomas are the commonest non-trophoblastic tumours of the placenta, majority of these are small and of little clinical significance. Tumors larger than 5cm are rare and have a high incidence of associated feto-maternal complications. The triad of polyhydramnios, pre-eclampsia and preterm labour associated with placental chorioangiomas has been described as chorioangioma syndrome. A case of large chorioangioma is presented highlighting the association of tumour vascularity detected by colour Doppler ultrasound with pregnancy complications.

Case Report

A 29-year-old primigravida was referred to the All India Institute of Medical Sciences, New Delhi at 30 weeks pregnancy with acute polyhydramnios and pre-eclampsia. Her pregnancy was uneventful till 25 weeks when her blood pressure increased from 110/70 to 140/90mm of Hg. Over the next four weeks she noticed rapid increase in the size of her abdomen for which she was referred to our center. Sonography revealed a single live fetus with normal growth parameters, the liquor was excessive with an AFI of 45. The placenta was located on the posterior wall and contained 8x8x6cm solid mass near the cord insertion, consistent with chorioangioma of the placenta (Fig 1). On color flow doppler, the tumor showed peripheral as well as central vascularity. There were no features suggestive of hydrops fetalis and the umbilical artery doppler was normal.

The patient was admitted to the antenatal ward with a blood pressure of 160/110mm Hg. Her haemogram, glucose tolerance test, kidney & liver function tests were normal. She was started on antihypertensive therapy and her BP was maintained between 140/90-140/100mm Hg on a combination of three drugs including methyldopa 1.5g, nifedipine 40mg and atenolol 50mg per day. Therapeutic amnioreduction was done twice to relieve maternal discomfort. Fetal surveillance was performed

Fig. 1 Ultrasound scan showing chorioangioma of the placenta
with twice weekly biophysical profile. A course of betamethasone was administered at 31 weeks.

At 33 weeks of pregnancy, patient developed high leak and uterine contractions after four hours. The cervix however failed to dilate after eight hours of good uterine contractions. A course of betamethasone was administered at 31 weeks. At 33 weeks of pregnancy, patient developed high leak and uterine contractions after four hours. The cervix however failed to dilate after eight hours of good uterine contractions. A slow amniocentesis was performed in an attempt to ameliorate the dysfunctional labor and reduce the risk of placental abruption. However after initial 200 ml of clear liquor, blood stained liquor was obtained. An immediate cesarean section was performed suspecting placental abruption. The liquor was blood stained. A male baby weighing 1.87 kg, appropriate for gestational age, with Apgar of 9 and 9 at 1 and 5 minutes was delivered. The placenta exhibited an 8x8cm tumor. The newborn was admitted to neonatal intensive care unit for observation. The baby was severely pale with general anasarca. His PCV was 25% and peripheral smear showed evidence of microangiopathic haemolytic anaemia. A diagnosis of chronic compensated anaemia possibly due to feto-maternal bleed consequent to placental chorioangioma was made. This was supported by a positive Kleihauer-Betke count of 5%. Blood transfusion was withheld, as he was well compensated and stable at room air. The baby was transferred to the mother on the third day, and was discharged from the hospital at 14 days of life with a PCV of 35%.

The patient made an uneventful recovery; her blood pressure gradually declined to normal and antihypertensive medication was withdrawn on the tenth post-partum day.

Discussion

Chorioangiomas are probably hamartomas derived from the primitive chorionic mesenchyme; what triggers their development however is not clear. It has been observed that placenta of women residing at high altitudes (more than 3600m) have a very high incidence of chorioangiosis and chorioangiomas (1), as high as 22% reported from Ukraine (2). Hypobaric hypoxia in these conditions probably induces abnormal vascular proliferation in the placenta to compensate for the tissue anoxia. The clinical course of pregnancies associated with chorioangiomas is variable, depending largely on the vascularity of these tumors. The commonest maternal complication reported is hydramnios in about 30% pregnancies, others are preeclampsia, preterm labor and placental abruption. As a result of a large volume of blood from the fetal circulation diverted through the arteriovenous shunts in the vascular channels of the tumor, fetal hypoxia occurs leading to congestive heart failure, cardiomegaly and hydrops fetalis. Fetal anemia, thrombocytopenia and fetomaternal hemorrhages have also been described. Our patient had acute hydramnios, severe preeclampsia, preterm premature rupture of membranes, placental abruption and fetomaternal hemorrhage. The neonate had hemolytic anemia at birth but escaped serious complications probably because it was delivered before cardiac failure could set in.

The first ultrasound diagnosis of a placental tumor was reported by Asokan et.al. in 1978 (3). However, grayscale sonographic appearances of chorioangiomas may be at times indistinguishable from those of placental hemorrhages. Color flow doppler can provide additional information by demonstrating increased blood flow within the tumor, thereby differentiating it from other placental masses. Besides diagnosis, color doppler is also of value in identifying pregnancies at greater risk of developing complications. In a recently reported series of nine patients, where color flow doppler was used for the prenatal assessment of chorioangiomas, three tumors appeared avascular and pregnancies were uneventful (4). Two had only few peripheral vessels and four had numerous vessels. All the six were complicated by polyhydramnios, preterm, labour non-immune hydrops in one. In our patient too, the tumor showed both peripheral and central vascularity and was associated with several maternal complications. On the basis of ultrasound and color doppler imaging, pregnancies with placental tumors which need intensive surveillance can be pre-selected. Patients with vascular chorioangiomas should be followed up with serial ultrasound and doppler assessment to detect hydramnios, early hydropic changes and fetal distress.

References