Placental chorioangiomatosis – a high risk pregnancy

R. Jaffe, A. Siegal, L. Rat, J. Bernheim, A. Gruber and M. Fejgin

Department of Gynecology A, and Department of Pathology, Meir General Hospital and Sackler School of Medicine, Tel Aviv, Israel

Summary: A case of diffuse chorioangiomatosis leading to fetal hydrops, disseminated intravascular coagulopathy with massive umbilical vein thrombosis and fetal death is described. Although rare, this benign mesenchymatous malformation of the placenta should be kept in mind as a possible cause of neonatal morbidity. Prenatal diagnosis could prevent fetal death.

Introduction

Chorioangioma is the most common neoplasm of the placenta. Although generally small, being only a few cm in diameter, it occurs with a frequency of up to 1% of all deliveries (Fox, 1978), and is clinically insignificant. However, larger chorioangiomata are rarer and can lead to symptoms. Hydramnios, toxaemia, fetal distress, anaeemia, fetal hydrops and neonatal death have been associated with large chorioangioma in one of 3500–9000 deliveries. An even rarer manifestation, chorioangiomatosis, is defined as a diffuse proliferation of the placental capillaries.

We present a rare case of chorioangiomatosis leading to the infant’s death with hydrops and massive umbilical vein thrombosis, associated with disseminated intravascular coagulopathy.

Case report

A thirty year old P³ G₄ woman was admitted in the 39th week of her pregnancy because of anæmia. She had gained 22kg during the pregnancy. Previous pregnancies were normal. Her abdomen was very distended, and blood pressure was 110/70 mmHg. She was not in active labour. Ultrason examination revealed polyhydramnios with enlarged fetal heart and liver. The placenta was hypertrophic, and over 5 cm thick. Fetal echography showed spontaneous decelerations of type II. An emergency caesarean section was performed and a female infant was delivered. The mother was discharged on the eighth postoperative day in good general condition. The infant was a 2950 g girl, Apgar scores were 6 and 9 at 1 and 5 min respectively. In the first hour she developed progres-

![Image](https://example.com/image.png)

Figure 1 Masses of endothelial-lined vascular channels with scanty stroma (H.E. × 40).

Correspondence: R. Jaffe, M.D., Dept. of Gynecology A, Meir Hospital, Kfar Sava 44281, Israel.

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Fig. 2  Haemangiomatous proliferation in chorionic villi (left) adjacent to normal appearing villi (right) (H.E. x 40)

stroma. In between these diffuse haemangiomatous areas, mature and normal chorionic villi were seen without syncitio-trophoblastic proliferation (Figure 2). At autopsy the entire length of the umbilical vein was occupied by a thrombus penetrating into the liver. Microscopically, the umbilical vein showed no signs of organization. There was prominent extramedullary haematoipoiesis in the liver. The lungs were partly aerated with a small amount of amniotic fluid and oedema in the alveoli.

Discussion

Chorioangioma, first described by Clarke in 1798, is the commonest neoplasm of the placenta. Most are small and clinically insignificant. Much rarer, large lesions can be associated with significant maternal and fetal complications. They are composed of benign proliferations of the villi’s haemangiomatous tissue. Three histological types are described: vascular (mature), cellular (immature), and degenerative types with myxomatous changes of the stroma. They are thought to be a hamartomatous malformation of the primitive angioblastic tissue of the early placenta. The diagnosis is generally made after the delivery of the placenta, as in our case; however, a few reports have been published of antenatal ultrasonic diagnosis (Asokan et al., 1978; Dao et al., 1981; Liang et al., 1982 O’Malley et al., 1981). More or less echogenic masses were found without variation of size during later stages of pregnancy and variable echogenicity in the degenerative areas. Placentomegaly is common. The associated maternal and fetal complications described (Wallenburg, 1971) are oligoamnios, premature labour, toxoaemia of pregnancy, fetal microangiopathic haemolytic anaemia, low birth weight, and congenital malformations. Polyhydramnios was reported in 22% of these cases (Fox, 1978; Wallenburg, 1971). McInroy & Kelsey (1954) suggested that the large angioma deviates fetal blood into its vascular network and this is not oxygenated, thus causing fetal anoxia (Knoth et al., 1976) and stimulating the increased excretion of waste products of metabolism; this increases the osmotic pressure of amniotic fluid and facilitates the passage of greater amounts of fluid into the sac probably from the placenta, uterus and fetus. Other attractive hypotheses have been proposed, namely, local compression of the umbilical vein by the neoplasm; transudation of fluid from vascular channels on the fetal surface; and the haemangioma acting as a peripheral arterio-venous shunt with fetal fluid imbalance caused by congestive cardiac failure.

Polyhydramnios can be associated, as in our case, with hydrops fetalis. In addition, this case was complicated by umbilical vein thrombosis as part of the symptomatology of disseminated intravascular coagulopathy (DIC). Thrombosis of umbilical vessels is an extremely rare condition. Fetal distress and death often occurs, but is most probably due to the underlying disease rather than to the thrombosis itself. Placental chorioangioma has rarely been associated with DIC (Jones et al., 1972) but large haemangioma elsewhere in the body can be.

The diffuse haemangiomatous proliferation was responsible for the severe maternal and fetal complications in this present case. Altshuler (1984) studied 1350 placentas of abnormal pregnancies all with neonatal morbidity, or and mortality. 'Chorioangiomas', namely more than 5 vessels in each chorionic villi, was found in 5.5% and significantly associated with fetal pathology. This case demonstrates the need for awareness of the presence of such lesions, and emphasizes the importance of antenatal diagnosis confirmed by pathological examination of the placenta immediately after delivery.

References


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