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Prenatal diagnosis of a large abdominal cyst – Recommendations and management

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ABSTRACT

This case report describes a twin fetus diagnosed in the third trimester with an enlarging abdominal mass that was confirmed on fetal magnetic resonance imaging (MRI) to be a hepatic mesenchymal hamartoma (HMH) without involvement of the placenta. Serial ultrasonography demonstrated progressive enlargement of the mass and infant was delivered at 33 + 6 weeks gestational age following maternal complications. On the fifth day, at laparotomy, a huge mass connected to the lower portion of the liver was completely resected. Microscopic evaluation confirmed a hepatic mesenchymal hamartoma. Postoperative recovery was uneventful and the infant was discharged at 4 weeks. The antenatal diagnosis of the hepatic mesenchymal hamartoma by fetal MRI and subsequent follow-up by serial ultrasonography emphasizes the importance of combining these 2 modalities for optimal management of the pregnancy to allow a favorable outcome.

1. Introduction

Hepatic mesenchymal hamartoma is a benign tumor which can grow rapidly intrauterine and cause polyhydramnios, hydrops and fetal demise [1,2]. Maternal and newborn serum alpha fetoprotein and beta human chorionic gonadotrophin maybe elevated, however these markers are not specific to HMH [3]. HMH is strongly associated with placental abnormalities mainly mesenchymal stem villous hyperplasia of the placenta [4,6]. A demonstration of translocation between chromosome 11 and, most commonly, chromosome 19 has been identified in HMH and is associated with an increased risk of subsequent malignancies [7,8]. Diagnosis of a fetal abdominal mass can ensure proper antenatal management and possibly prevent fetal complications and fetal demise. We describe a twin infant with a perinatal diagnosis of abdominal mass.

2. Case report

A 27 year old gravida 2 para 1 woman was admitted to hospital at 29 weeks gestational age (GA) following vaginal bleeding and preterm uterine contractions. The bi-chorionic, bi-amniotic twin pregnancy had followed treatment with in vivo fertilization (IVF). The first pregnancy four years previously had been uneventful.

On admission in preterm labor, she was managed with intravenous fluids and antenatal betamethasone and a prenatal US was performed. In the first twin the prenatal US demonstrated polyhydramnios and a large abdominal mass occupying almost the entire abdomen (Fig. 1). Heterogeny (solid and cystic characteristics) and proximity to the liver was noted and the lesion had irregular borders. The mass displaced all the intra-abdominal organs laterally and the diaphragm was elevated bilaterally. No blood flow was demonstrated on Doppler studies and therefore an hemangioendothelioma was unlikely. There was no evidence of fetal ascites. The mass measured 60 × 55 mm. Fetal echocardiography was normal and no other malformations were identified. Preterm delivery was prevented and the mother was discharged home for outpatient follow-up.

At 30 weeks' GA, a further US demonstrated a significant growth in the size of the mass. To determine the identity of the fast growing mass a fetal MRI was performed. MRI demonstrated an abdominal mass connected by a stalk to the liver (Fig. 2). The mass had high signal

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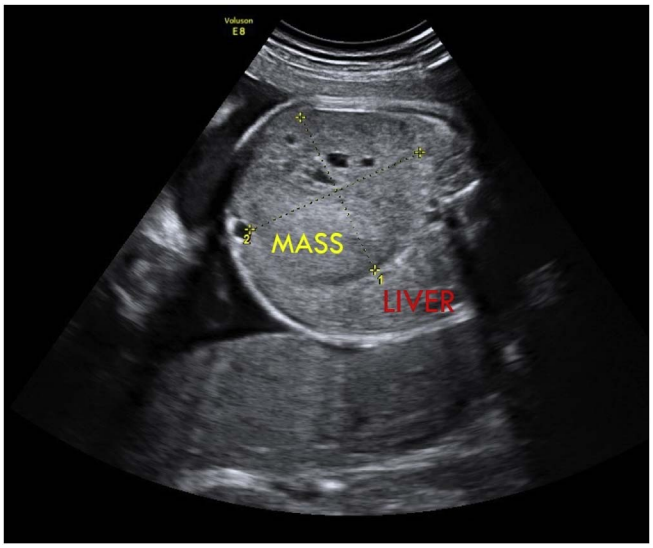

Fig. 1. Prenatal ultrasound demonstrated polyhydramnios and a large abdominal mass occupying almost the entire abdomen.

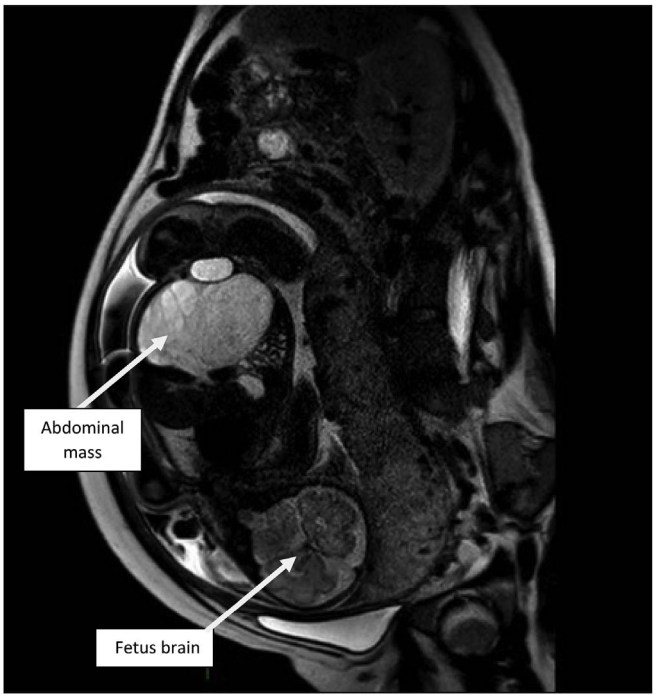

Fig. 2. Fetal MRI Images demonstrating mesenchymal hamartoma.

heterogenicity with multiple septae, and the T2 sequence showed numerous focal lesions within the mass that were suspected to be hemorrhagic lesions or calcifications. Besides the liver, the mass was not attached to any other abdominal organs and a provisional diagnosis of a cystic hepatic mesenchymal hamartoma was made. At 32 weeks GA although the mass had enlarged there was no change in the morphology or any evidence of fetal decompensation.

At GA 33 + 6, there was significant vaginal bleeding from placenta previa and the pregnancy was terminated by Caesarean section. The first twin (male) weighed 2475 g and had an APGAR score of 2 and 7 at 1 and 5 min respectively. Physical examination demonstrated a markedly distended abdomen without skin discolouration. A large smooth mass was palpable occupying the whole abdomen with clear borders. The infant was intubated and transferred ventilated to the neonatal intensive care unit where an umbilical artery catheter and a

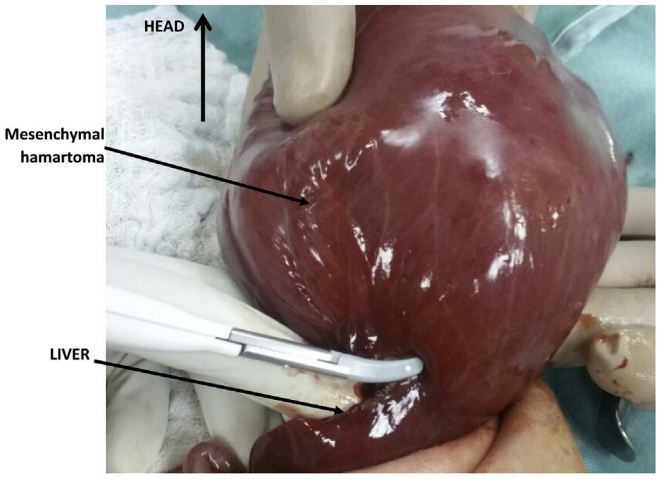

Fig. 3. Removal of hepatic mesenchymal hamartoma by open laparotomy.

percutaneous intravascular central line were inserted. The infant required mechanical ventilation and treatment with inotropic agents. An echocardiogram demonstrated poor cardiac contractility. Head and renal US were normal. Full blood count, coagulation screen and alpha fetoprotein were normal, liver transaminases were mildly elevated although direct bilirubin and gamma GT were within the normal range.

Initial abdominal US demonstrated a huge solid mass with peripheral cystic lesions occupying the entire abdomen and pelvis. The liver was displaced to the right upper quadrant and the diaphragm was elevated. A needle aspirate was performed under computer tomography (CT) guidance in order to obtain cytology prior to surgery and a small amount of gelatinous material was extracted with no pathological diagnosis. An exploratory laparotomy was planned once cardiovascular stability was obtained. At surgery, the mass, which had a macroscopic appearance of a hamartoma, was exposed and carefully removed from the lower border of the liver (Fig. 3). Intra-operative ultrasound was used to assess the quality of the surgical resection. Pathology and cytologic studies confirmed the diagnosis of HMH (Fig. 4). The infant made a complete post operative recovery, gained weight and was discharged home on full enteral feeds.

3. Discussion

Abdominal masses frequently present in the fetus and the diagnosis is often unclear [1]. We describe a case of hepatic mesenchymal hamartoma (HMH) in a twin infant following an IVF induced pregnancy. The prenatal diagnosis was made initially by fetal ultrasound and confirmed by MRI at 30 weeks gestation. There was no indication for fetal intervention as no signs of fetal compromise were demonstrated. The infant was born at 34 weeks gestation and the large mass was removed completely during laparotomy. Pathology confirmed the diagnosis and that the excision of the lesion was complete with clear borders.

HMH is a rare benign tumor usually presenting in childhood as a rapidly growing asymptomatic mass [9]. There have been 14 reported cases of HMH suspected prenatally following the diagnosis of a fetal abdominal mass, however only in 4 cases was the diagnosis of HMH confirmed prior to delivery [10]. The diagnosis might be suspected if abnormalities are noted in the placenta. Mesenchymal stem villous hyperplasia is a diffuse edematous expansion of the stem villi in the placenta that has been associated with fetal anomalies including HMH and Beckwith Wiedeman syndrome [4]. Kitano et al. suggested that this placental pathology might share a common pathogenesis with HMH [5]. Laberge considered the placental pathology was secondary to umbilical vein obstruction by the fetal liver tumor [6].

A fetal abdominal mass demonstrated on fetal ultrasound demands