Fetal hepatic mesenchymal hamartoma: a case report

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Case report

A 38 years-old woman, gravida 2, para 1 came to the Artemisia Prenatal Diagnosis Centre at 36 weeks of gestation referred from another institution for the evaluation of a fetal mass in the upper-right abdomen. Previous ultrasound scanning performed at 18 and at 22 weeks of gestation were negative for fetal abdominal masses. Amniocentesis performed at 18 weeks of gestation was negative and the alfa fetoprotein in the amniotic fluid was 11243 (n.v. < 26000).
The ultrasound scan of the fetus demonstrated a multicystic, well-encapsulated mass in the liver of 7×5×5 cm in size (Fig.s 1, 2). No calcifications were observed into the mass; there were both echolucency and echodense areas. The Doppler flow investigation of the mass demonstrated a relatively low-absent vascularisation. Others organ near the neoplasm were all normal. No hydrops was observed. The only remarkable sign was the elevated amniotic fluid index (75° centile). After a multidisciplinary counselling a caesarean section was planned at 37 weeks-of gestation and a strict control of the fetus with US was performed until the delivery date. Controls were all negative and polyhydramnios was equal in all measures.
The CS was performed at 35 weeks of gestation due to uterine contractions, giving birth to a 2.9 kg female infant. Apgar scores were 7 and 8 at 1 and 5 minutes respectively. At birth, the abdomen was extremely distended and the baby was referred to the Bambino Gesù Children's Hospital that is a tertiary care centre with the diagnosis of acute abdomen. At physical examination on arrival the abdomen was distended and tender. An abdominal X-ray showed an elevated diaphragm and virtually no gas in the bowel. Blood tests were unremarkable. On the hypothesis of a neonatal volvolus secondary to an intestinal atresia an emergency laparotomy was carried out. At surgery, a huge mass was found taking up to three quarters of the abdomen, with the bowel compressed but not necrotic. The mass originated from the anterior margin of the liver and was pedunculated. The peduncle was suture ligated and the mass resected. Immediately after the removal of the mass, acute anaemia occurred (Haemoglobin level 5 g/dl), probably from blood sequestration into the mass itself, leading to profound bradycardia and cardiac arrest, successfully treated with prompt cardiopulmonary resususcitation and blood transfusion. At final histology, the mass was compatible with a mesenchimal amartoma of the liver. Postoperative course was complicated by chylothorax which

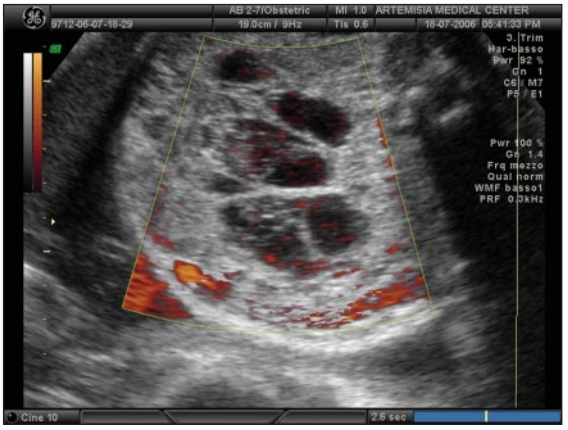

Figure 1

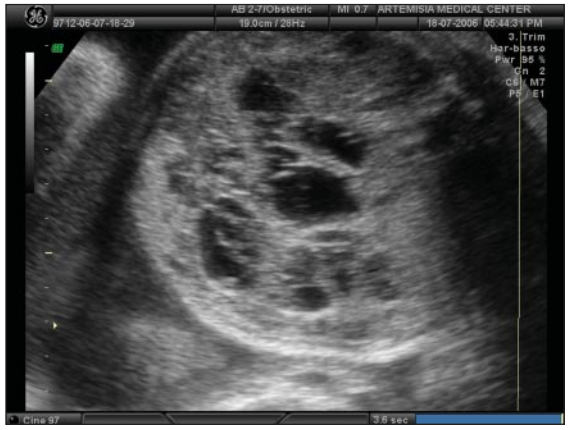

Figure 2

required total parenteral nutrition for two weeks and Octreotide for 7 days, and when enteral nutrition was resumed, a medium chain triglicerides milk was initially used to reduce the risk of recurrent chylothorax. During the post-operative course, neurological evaluation and serial head US were performed which showed no hypoxic brain damage, other mutations causing cystic fibrosis were excluded by specific genetic testing, and the girl was discharged on post-operative day 33 in good clinical.

Discussion

The incidence of liver masses in general represent approximately 5% of all congenital tumors (1) and they can arise from the mesenchymal or endodermal tissues giving a wide variety of both benign and malignant masses. A mesenchymal hamartoma of the liver is the second tumor of the hepatic tissue after the hemangioblastoma (2) and is a benign neoplasm composed of large, fluid-filled cyst surrounded by mesenchymal membranes containing small bile ducts (3), with a rapidly growly mass.

As most of these tumors are detected in the first year of life (4) and only 15 % in the neonatal period (5) diagnosis “in-utero” is very rare and the differential diagnosis is very difficult to do because the natural history of the tumor is still not know. Thus only pathological findings after surgical resection is the cornerstone in the definition of the mass.

In this group of fetuses issuing concern the diagnosis early in pregnancy is of utmost importance due to the possibility of organs failure related to the growing of the mass (Hirata et al.) leading to fetal demise (6-8).

The differential diagnosis of MHL with other abdominal masses poses challenging problems to the obstetrics because many malformation both of fetal urinary system (multicystic renal malformations) and gastrointestinal tract (congenital atresias and stenosis) could be exchanged for a MHL (9). Moreover, no laboratory tests for liver function are helpful in the early diagnosis of the MHL (10) and AFP arising in the maternal blood could be normal the amniotic fluid (11).

Others fetal conditions must be considered in the differential diagnosis after these discussed above. This includes bowel obstructions subsequently to a mechonium ileum or neurological disease (9). A good visualization of the gastrointestinal system can minimize the error in the diagnosis.

Due to the rarity of this condition the management cannot rely on robust and evidence-based data. We managed our case expectantly in the prenatal period with serial ultrasound examinations given the lack of complications

Today is widely accepted the benign nature of the MHL and the primary endpoint is to reduce the size of the mass (12). Hence the early diagnosis of abdominal masses is important because they have the potential to cause complications such as intestinal ischemia by their mass effect infect among 6 cases before 31 wks of gestation only 2 were alive and 1 of these had cyst aspiration at 27 (13). Although they may displace adjacent structures, obstruction of the bowel, vessels, or ureters is never been described. In general, large cystic masses require surgical excision and when the diagnosis of MHL is near to term the prognosis seems to be better and the postnatal surgical treatment is recommended (14).

We believe that an early antenatal diagnosis and successive follow-up could help to established the time for delivery making the expectant management an opportunity for this condition.

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