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 alpha 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 resuscitation 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
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required total parenteral nutrition for two weeks and Oc-
trotide for 7 days, and when enteral nutrition was re-
sumed, a medium chain triglycerides 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 hy-
poxic brain damage, other mutations causing cystic fi-
brosis 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 ap-
proximately 5% of all congenital tumors (1) and they can
arise from the mesenchymal or endodermal tissues giv-
ing a wide variety of both benign and malignant masses.
A mesenchymal hamartoma of the liver is the second tu-
mor 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 grow-
ning mass.

As most of these tumors are detected in the first year of
life (4) and only 15% in the neonatal period (5) diagno-
sis “in-utero” is very rare and the differential diagnosis is
very difficult to do because the natural history of the tu-
mor is still not know. Thus only pathological findings af-
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 abdomi-
nal 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 ex-
cluded for a MHL (9). Moreover, no laboratory tests for
liver function are helpful in the early diagnosis of the
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cluded for a MHL (9). Moreover, no laboratory tests for
liver function are helpful in the early diagnosis of the
mass (10) and AFP arising in the maternal blood could
be normal the amniotic fluid (11).

Others fetal conditions must be considered in the differ-
etial diagnosis after these discussed above. This in-
cludes bowel obstructions subsequently to a mechoni-
um ileum or neurological disease (9). A good visualiza-
tion of the gastrointestinal system can minimize the er-
ror in the diagnosis.

Due to the rarity of this condition the management can-
t not rely on robust and evidence-based data. We man-
aged our case expectantly in the prenatal period with
serial ultrasound examinations given the lack of compli-
cations.

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 im-
portant because they have the potential to cause complica-
tions such as intestinal ischemia by their

mass effect infect among 6 cases before 31 wks of ges-
tation only 2 were alive and 1 of these had cyst aspira-
tion at 27 (13). Although they may displace adjacent
structures, obstruction of the bowel, vessels, or ureters
is never been described. In general, large cystic mass-
es 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 suc-
cessive follow-up could help to established the time for
delivery making the expectant management an opportu-
nity for this condition.

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