Case Report

Fetal hepatic mesenchymal hamartoma. A case report

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Abstract

Hepatic tumors accounted 5% of congenital neoplasms. Mesenchymal hamartoma of the liver is a rare benign childhood tumor, whose definitive diagnosis during the fetal period remains difficult, despite advances in antenatal imaging. In this paper, we report a case of hepatic mesenchymal hamartoma diagnosed prenatally with ultrasound scan showing a multicystic mass in the left upper abdomen accompanying polyhydramnios and complicated by a preterm labor. The newborn died on the first day of life due to respiratory distress and neonatal jaundice. The diagnosis was confirmed histologically post-delivery.

Key words
Hepatic mesenchymal hamartoma, fetal ultrasound scan, prenatal diagnosis.

Introduction

Mesenchymal hamartoma of the liver (MHL) is an extremely rare benign childhood tumor, whose etiology remains unclear. Described for the first time in 1956 by Edmondson [1], MHL is an uncommon benign hepatic tumor that in 80% of cases is detected during the first two years of life [2]. Despite advances in antenatal imaging have allowed accurate and earlier diagnosis of MHL during the fetal period, a definitive diagnosis of this tumor in utero remains difficult. In fact, including the current case, there have been only 20 reported cases of MHL diagnosed or detected prenatally by ultrasound examination [3]. Although this lesion is histologically benign, its rapid growth to enormous size may result in perinatal complications such as fetal hydrops, maternal toxemia, preterm labor and intrauterine fetal death.

We report a case of hepatic mesenchymal hamartoma diagnosed prenatally with ultrasound scan showing a multicystic mass in the left upper abdomen accompanying polyhydramnios and complicated by a preterm labor. The diagnosis was confirmed histologically after delivery.

Case report

A 34-year-old pregnant women, gravida 3 para 2, was referred to our unit at 33 weeks of gestation for management of her pregnancy. An antenatal ultrasound showed a highly vascularized intra-abdominal anechoic cyst, occupying two-third of the abdominal cavity and pumping up the intestine associated with polyhydramnios. Since cyst was compressing other organs and distending abdomen, a termination of pregnancy has been proposed but refused by the couple. At 34 weeks of gestation, the pregnancy was complicated by a preterm labor and a male newborn of 2600 g was vaginally delivered. The examination of the newborn showed an abdominal firm lump. Postnatal X-chest radiograph showed a compression of both lung fields by the abdominal mass. The newborn died on the first day of life due to respiratory distress and neonatal jaundice. Postmortem abdominal ultrasound noted an anechoic cyst, measuring 13 cm, in close relation to the left lobe of liver. A complete autopsy was performed and the external examination showed a male neonate anatomically of 34-35 weeks having

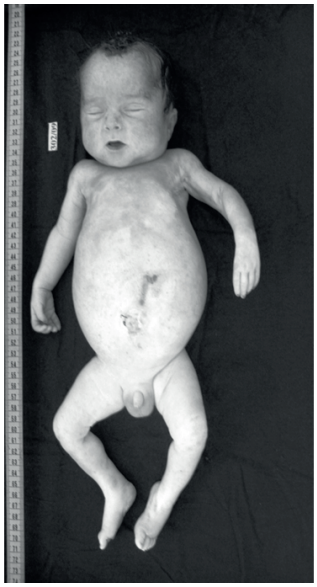

Figure 1. External examination: male neonate anatomically of 34-35 weeks with an abdominal dilatation and a venous collateral circulation (with permission from his parents).

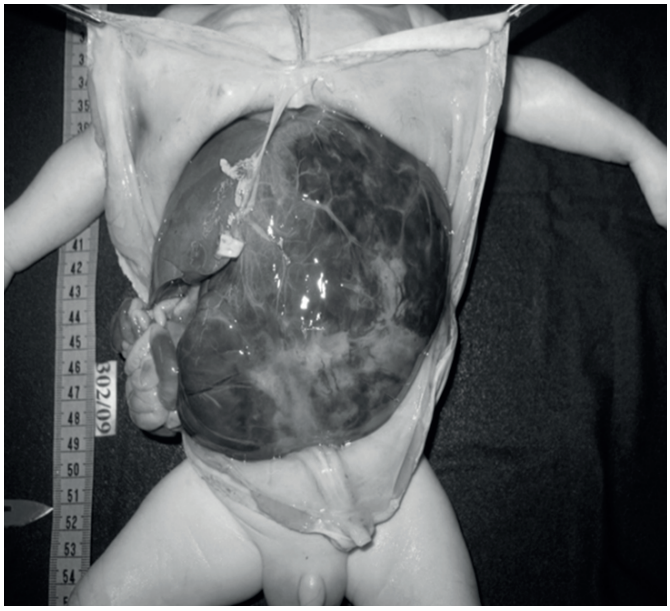

Figure 2. Macroscopic examination: hepatic tumor of the left lobe showing bleeding reshuffle (with permission from his parents).

an increased periumbilical diameter, an abdominal dilatation and a venous collateral circulation (Figure 1). Fetal dissection noted an ascites, a splenomegaly, a pulmonary hypoplasia, a pleuro-pericardial effusion, and a cardiomegaly. The liver weighted 383.7 g (normal: 60 g) and was the seat of a well limited tumor developed in the left lobe and measuring 14 cm, with hemorrhage and necrosis (Figure 2). Histologically, the tumor showed a mixture of normal liver tissues with blood or lymphatic vessels, bile ducts within an abundant edematous and myxoid stroma (Figure 3). The histopathology description confirmed the diagnosis of a mesenchymal hamartoma of the liver. The histological examination of the placenta was not done.

Discussion

Primary hepatic tumors are rare in children, where they account for about 5% of all intra-abdominal masses and represent between 0.5% and 2% of all pediatric neoplasms [4]. Mesenchymal hamartoma of the liver (MHL) is the second most common benign hepatic tumor in children [4], defined as an excessive focal overgrowth of mature normal cells and stroma native to the liver [5] and presents as a large, rapidly growing mass during early infancy [6]. A strong female predisposition for fetal MHL was reported, and this is in contrast with the male preponderance for postnatal MHL [2]. Microscopically, MHL consists of spindle cells in a myxoid background, with occasional areas of extramedullary hematopoiesis, all in a disordered arrangement of mesenchyme, malformed bile ducts, and cords of normal-appearing hepatocytes [4]. Cytogenetically, these tumors are characterized by translocations involving 19q13.4 [4].

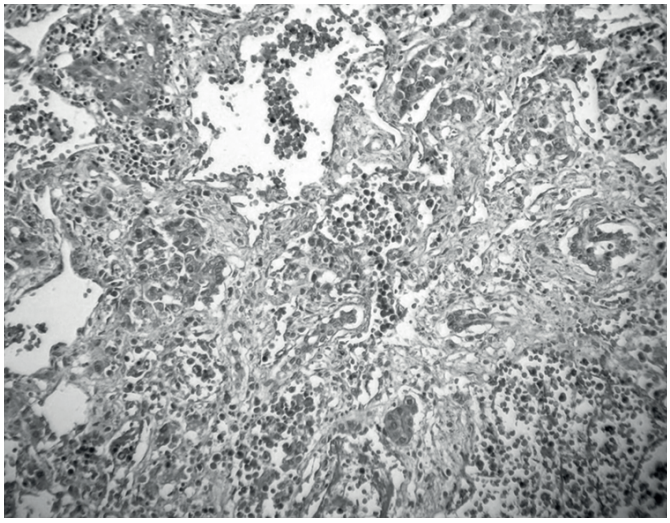

Figure 3. Histopathology examination: mixture of normal liver tissues with blood or lymphatic vessels, bile ducts within an abundant edematous and myxoid stroma.

Even with advance prenatal diagnostic tools, prenatal diagnosis of MHL remains challenging because the hepatic origin can be difficult to be specified in ultrasound examination. MRI can be useful to identify liver tissue, which is the only organ to produce a hyperintense signal on T1 imaging [7, 8]. In our case, the prenatal diagnosis of MHL was not made because the origin of the cystic intra-abdominal masse was uncertain. Usually, prenatal ultrasound detects MHL during the last trimester of pregnancy with a mean gestational age at 35 weeks [4, 9]. The MHL presented prenatally as multicystic and hypovascular masses, but mixed

and solid lesions were also described. In the largest series by Isaacs [9], 45 cases of mesenchymal tumors are reported over a period of 35 years. However, only 14 cases were prenatally diagnosed in this series and most common presentation was abdominal cyst with a mean gestational age of 35 weeks. Since the differential diagnosis is very difficult to do because the natural history of the tumor is still not known, only pathological findings after surgical resection is the cornerstone in the definition of the mass [10].

Although this lesion is histologically benign, associations with congenital heart disease, intestinal malrotation, biliary atresia, omphalocele, myelomeningocele, Beckwith-Wiedemann syndrome, and abnormalities of chromosome 19 have been reported [2, 4, 5]. While the MHL has generally a good prognosis in childhood, the outcome is much worse when diagnosed in the prenatal period with a mortality rate of 35% [2, 9, 11]. In our case, the liver tumor was diagnosed at 33 weeks of gestation and was associated with a polyhydramnios and a compression of the lungs. Prenatal occurrence of these tumors is associated with adverse outcome mainly due to its mass effects. Poor prognostic factors are mainly represented by the early onset of presentation, the rapidly progressing tumor, and the polyhydramnios [6, 7, 12]. Congestive cardiac failure reported in case of MHL is due to the compression of the inferior vena cava and umbilical vein [11]. The risk of hydrops is increased by the loss of fluid to the cysts and the reduced liver production of fetal albumin [11]. Polyhydramnios is associated with upper intestinal tract obstruction and elevation of the diaphragm poses the fetus at risk for pulmonary hypoplasia.

Invasive antenatal procedures remain controversial and a balanced consideration in a multidisciplinary team is mandatory in each individual patient. In the literature, some authors proposed intrauterine cyst drainage [7] since the risks of simple or repeated needle aspiration appear minimal compared to the consequences of a large fetal abdominal mass. Tsao et al. [6] suggested that the goal of the treatment with repeated aspirations was to reduce the lesion to a manageable size providing adequate decompression to allow for proper placental and fetal organ development and safe vaginal delivery. However, other authors demonstrated that the drainage of the cyst fluid do not reduce the production of cyst fluid, firstly because the fluid would have reaccumulated in the cyst [13, 14] and, secondly, because multilocular cysts frequently have no communication with each other [13, 15]. Thus, antenatal therapy is only decompressive and does not reduce the need for postnatal surgical resection which offers definitive diagnosis and treatment. The surgical treatment should have as purpose to reduce the production of cyst fluid using a complete resection when possible or partial resection and cauterization of the cyst wall adherent to hepatic parenchyma if complete excision of the tumor is dangerous or invasive [11].

Conclusion

Despite technological advancements and efforts toward early diagnosis, the prognostic of the MHL remains poor due to mass effects and associated fetal malformations. Actually, no firm recommendations can be made about the mode of delivery of the fetus with a presumed MHL, and the invasive antenatal procedures remain controversial. Thus, several publications are still needed to establish a consensus for optimum management.

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