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ORIGINAL PAPER

# A Prenatally Detected Case of Congenital Hepatoblastoma

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**Abstract** Hepatoblastoma is a rare tumor of childhood and its incidence in the first year of life is about one in a million. Forty-two congenital hepatoblastoma cases were reported so far. Among 42 congenital hepatoblastoma patients, only seven cases have been detected in the prenatal period. Here we report a rare case diagnosed before birth and confirmed by postmortem autopsy.

**Keywords** Hepatoblastoma · Prenatal diagnosis · Ultrasound

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## Introduction

Hepatoblastoma is a rare tumor of childhood. The incidence of hepatoblastoma in the first year of life is about one in a million. The mean time of its onset is 14 to 24 months. [1] Forty-two congenital hepatoblastoma cases were reported so far. Among 42 congenital hepatoblastoma patients, seven cases have been detected in the prenatal period. Only one out of seven cases detected in the prenatal period has been diagnosed as hepatoblastoma [1, 2]. The etiology of hepatoblastoma is unknown. However, it has been shown to be associated with prematurity, low birth weight, hepatitis B, familial adenomatous polyposis, Beckwith-Wiedemann syndrome and hemihypertrophy.

In this report, we report a rare case of hepatoblastoma detected before birth and confirmed by postmortem.

## Case Report

Thirty years old primipar pregnant woman, who could not receive adequate prenatal follow up and care, has been admitted to our hospital for labor at 34th weeks of gestation. Family history and pregnancy were unremarkable. Both parents were healthy and nonconsanguineous. There was no history of maternal diabetes or exposure to any teratogenic agent during the pregnancy. Maternal hepatitis B surface antigen was negative. Prenatal ultrasound revealed a big, solid and hyperechogenic mass located over the right lobe of fetal liver (Fig. 1). Tumor borders were clearly visible, seperated from surrounding tissues and strongly suggested the diagnosis of hepatoblastoma. No hydrops fetalis was demonstrated.

The boy was delivered by the spontaneous vaginal route. Aggressive positive pressure ventilation was used due to

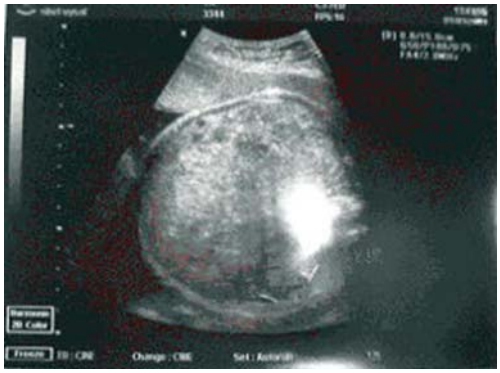

**Fig. 1** Prenatal ultrasound revealed a big, solid and hyperechogeneous mass located over the right lobe of fetal liver

the lack of spontaneous respiration. Apgar scores for the first and the fifth minutes were 2 and 5, respectively. Due to respiratory distress, he was intubated and supported by mechanical ventilation in the newborn intensive care unit. Birth weight was 2,800 g (75th percentile), length was 47 cm (50th percentile), occipitofrontal head circumference was 33 cm (50th percentile). He was dramatically pale and hypotonic. He had central cyanosis and apparent abdominal distention. The rest of the physical examination was unremarkable (Fig. 2).

Laboratory findings revealed marked anemia (Hb: 5.4 g/dL), mild thrombocytopenia (96,000/mm$^3$), a normal white blood cell count (12,100/μL). His liver function tests were normal (AST 45 U/L, ALT 20 U/L) but serum alfa-feto protein level was quite high (366,000 ng/mL). Computerized tomography images of the chest and abdomen showed a giant mass originating from the right liver lobe, which was 11×7 cm in size (Fig. 3). The mass had cystic areas and a rich vascular supply. Minimal ascites was detected and no metastatic lesions were demonstrated.

The patient developed tachycardia during the follow up. Blood oxygen saturation remained low and red blood cell transfusion was administered. Despite symptomatic treatment and aggressive cardiopulmonary resuscitation, the baby died at the 10th hour of birth. Postmortem partial autopsy confirmed the diagnosis of hepatoblastoma of mixed type. (Figs. 4 and 5).

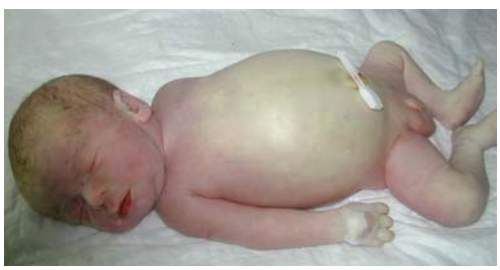

**Fig. 2** Infant had apparent abdominal dystension, central cyanosis and he was dramatically pale and hypotonic

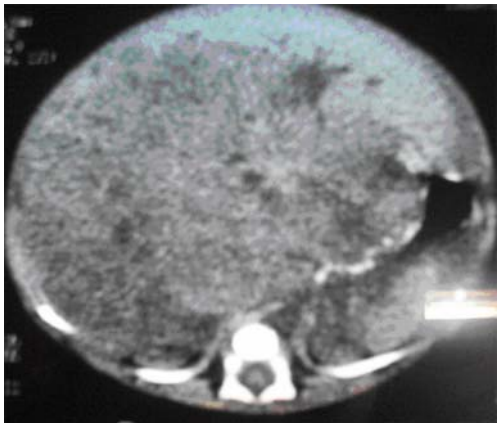

**Fig. 3** Computerized tomography images of abdomen showed a giant mass originated from right liver lobe

## Discussion

Hepatic tumors constitute 0.5% to 2% of pediatric neoplasms, but are rare in the newborn period. Forty three percent of the cases with primary hepatic tumors have been shown to be hepatoblastoma, the most common type of hepatic tumors of the neonatal period [1]. Hepatoblastomas are well-defined, solid, echogenic lesions, which may have a spoked-wheel appearance. A pseudocapsule gives the lesions a characteristic well-demarcated appearance [3]. More than 60% of the cases arise from the right lobe of the liver and in lesser extent from both lobes [4-7]. Only one out of seven cases of hepatic masses detected in the prenatal period had been diagnosed correctly as hepatoblastoma [1, 2]. The differential diagnosis of other intrahepatic echogenic fetal liver masses includes metastatic neuroblastoma and benign entities such as hemangioma, mesenchymal hamartoma, adenoma, and focal nodular hyperplasia [8]. [1] reported a case who was diagnosed as congenital hepatoblastoma by intrauterine doppler ultrasound.Vascular characteristics of the tumor is easily identified by Doppler ultrasound and can be differentiated

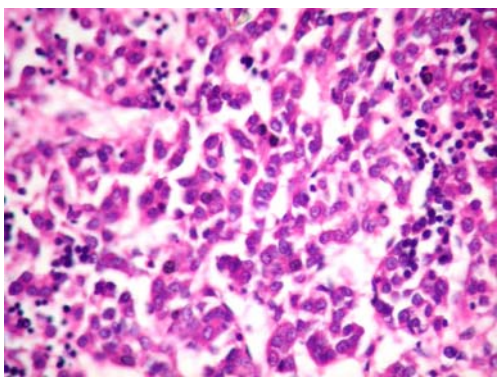

**Fig. 4** Trabecular areas and hematopoetic focus in mixed type hepatoblastoma (H&E, ×20)

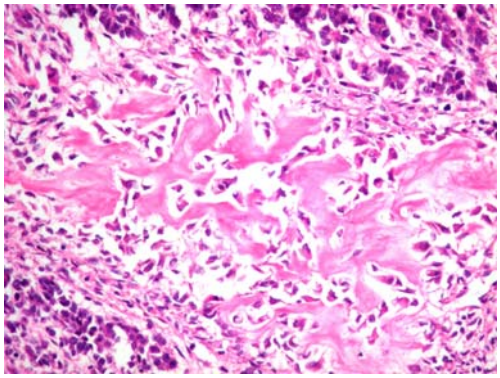

**Fig. 5** Mesenchimal component including osteoid material in mixed type hepatoblastoma (H&E, ×10)

from other benign cystic lesions [9]. Doppler ultrasound could not be performed in our case. However gray-scale ultrasound images showed giant well-defined echogenic solid mass and contrast enhanced CT revealed that the mass was hypervascular and originated completely from the right lobe of the liver.

Alpha fetoprotein which is mainly produced by fetal liver cells and yolk sac can be detected high levels in the blood of patients with hepatoblastoma, hepatocellular carcinoma, germ cell tumors and teratocarcinoma. The ability to synthesize alpha fetoprotein of the tumor suggests that it is of fetal origin. Embryonic tumors secrete less alpha fetoprotein than fetal tumors do. The high level of alpha fetoprotein in our case supports the fetal origin.

Differential diagnosis of hepatic tumors of the newborn includes hepatoblastoma, hepatocellular carcinoma, mesenchymal hamartoma, infantile hemangioendothelioma; however, rare pathologies such as metastatic neuroblastoma, leukemia and lymphomatous infiltration should also be considered [10-12]. Unlike older children, hepatoblastoma cases diagnosed in the first months of life demonstrate tumors of purely fetal histological type. Those cases have a higher risk of metastases and poorer prognosis [2]. In the present case, macroscopic and microscopic studies showed no bleeding in or outside of the tumor. Histologic examination revealed mixed type hepatoblastoma with both epithelial and mesenchymal component. Bone marrow investigation could not be performed. Computerized tomography of the chest and abdomen showed no metastases.

Due to the increased risk of rupture, cesarean section (C/S) delivery of the baby is recommended in prenatally detected cases of hepatoblastoma [13]. Seven of the congenital cases were detected in utero; in only two of them, a C/S was performed and there was not any tumor rupture in those patients. On the other hand, in four of the five cases born via the vaginal route, the tumors were ruptured [2]. In the present case, an abdominal mass was detected in the intrauterine period and the diagnosis of hepatoblastoma was confirmed by autopsy. Despite vaginal delivery, the tumor was not ruptured in our case.

Intrauterine diagnosis provides the time of counselling and planning of the postnatal intervention for neonatologist and the pediatric surgeon. Perioperative chemotherapy makes it easier to resect the larger tumors and total resection results long term survival [14]. Chemotherapy and surgery is difficult and bears a high risk in the newborn patients. Therefore, five of 15 congenital hepatoblastoma cases alone were given chemotherapy before surgery in literature [15]. Neither chemotherapy nor surgery was used in the present case since the delivery had begun on admission, and the baby died at the 10th hour of birth.

The prognosis of cases with congenital hepatoblastoma is quite poor. Polyhydroamniosis has been reported in most cases and non immune fetal hydrops may develop. The pressure of the mass may result in respiratory distress. It is reported that, six of the 7 cases examined by intrauterine ultrasound died within the first 15 days of life [2]. Severe respiratory distress caused by the mass and anemia of unexplained origin contributed to the poor condition of the present case. The patient's condition deteriorated despite mechanical ventilation support and adequate symptomatic treatment including blood transfusion.

The origin of anemia could not been determined in this case (Hb: 5.4 g/dL). Involvement of the bone marrow by the tumor might have been suggested, but the platelet and white blood cell counts of the patient were within normal limits. A complete autopsy and bone marrow examination could not be performed because of the disapproval of the parents. Macroscopic and microscopic studies showed no rupture of the tumor. Regular attendance to follow up visits and effective application of ultrasound and doppler ultrasound during pregnancy would lead to early diagnosis and may result in a better prognosis.

In conclusion, even though hepatoblastoma is peaked at 16th month of life in childhood, it may be detected in the prenatal period. A careful attention should be payed for mass images during routine intrauterine ultrasound studies and when detected, further investigation should be planned for such masses.

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