Mesenchymal Hamartoma of Liver: A Case Report

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Mesenchymal hamartoma of the liver (MHL) is an oncological entity occurs almost exclusively in infancy and childhood. Fewer than 200 cases have been reported. We present a case of a 7-month-old female infant, who was admitted to our institution with abdominal distension. Computed tomography (CT) showed a 12-cm multiloculated mass in the left lobe of liver. Angiography demonstrated a hypovascular mass with stretching of the left intrahepatic vessels. The patient underwent surgical enucleation of the tumor. The pathological report confirmed a mesenchymal hamartoma of the liver. There had been a long time since the heat of the 90's related to hamartomatous lesions of the liver. Little reports in the radiological field were announced ever since.

Key words: Liver, mesenchymal hamartoma; liver, computed tomography; liver, angiography

Hepatic mesenchymal hamartoma is an uncommon benign lesion arising from the mesenchyme of the portal tract and represents a developmental abnormality rather than a true neoplasm [1]. Malignant transformation was reported [2]. High risk of mesenchymal hamartoma associated with undifferentiated embryonal sarcoma was announced before [3-4]. Edmondson first concentrated mesenchymal hamartoma in 1956 when he recognized the similarity among synonyms of lymphangioma, bile cell fibroadenoma, hamartoma, cavernous lymphangiomatoid tumor, pseudocystic mesenchymal tumor, and cystic hamartoma [1]. The hamartoma usually presents within the first 2 years of life, abdominal distension is usually the predominant clinical features [1,5,6]. When tumor is bulky, mass effect may cause respiratory distress and lower extremities edema [7]. Few cases of adult mesenchymal hamartoma were also reported [8-12].

CASE REPORT

A 7-month-old female demonstrated abdominal fullness for three weeks. Poor appetite, activity, and urine output were noticed since then. She came to local hospital for help. Sonography showed a huge mass compressing the right kidney. She was transferred to our institute for help. Laboratory data were all within normal limits. An enhanced computed tomography (CT) demonstrated a 12x11x9 cm multiloculated mass in the left lobe of the liver (Fig. 1A and 1B). Angiography revealed a hypovascular mass in the left lobe of the liver with stretching of the intrahepatic vessels (Fig. 2). The patient was operated under the impression of mesenchymal hamartoma. Enucleation of tumor mass was done. Pathological examination showed proliferation of loose mesenchymal tissue with interspersed cystic spaces and some are lined by bile ductal epithelium. Many cysts contain proteinacious
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fluid (Fig.3). The final diagnosis was mesenchymal hamartoma of liver (MHL). She was discharged after one month and no tumor recurrence for more than 4 years.

**DISCUSSION**

In current concept, MHL is viewed as a congenital process for which the histogenesis is not well understood. Whether it represents developmental anomaly, a reaction to biliary obstruction, or even the result of regional ischemic process rather than a true neoplasm has become a topic under discussion [13-15]. Nevertheless, this lesion has been regarded as benign entity without potential for malignancy. Among hepatic masses in the pediatric patients, MHL is more frequent than focal nodular hyperplasia or adenoma, as frequent as undifferentiated embryonal sarcoma (UES), and less frequent than infantile hemangiendothelioma, hepatoblastoma, or hepatocellular carcinoma [5].

Clinical symptoms of MHL usually emerge between ages 4 months and 2 years [1], except with a few exceptions in newborn and adult patients [1, 8-12]. There is a slight male predominance [16], but not for this patient. The tumors usually grow slowly. Rapid increase in size may signify accumulation of fluid within the cyst [1, 5, 6]. Most of the mesenchymal hamartoma affects the right lobe of the liver [5, 6].

Figure 1A. Non-enhanced computed tomography shows a 12x11x9 cm multiloculated mass in the left lobe of the liver. Different density of the fluid within the locules in the mass is demonstrated. 1B. Contrast enhancement computed tomography shows enhancement of the septations within the mass.

Figure 2. Angiography reveals a hypovascular mass in the left lobe of the liver with stretching of the intrahepatic vasculature.

Figure 3. Photomicrography shows proliferation of loose mesenchymal tissue with interspersed cystic spaces and some are lined by bile ductal epithelium. Many cysts contain proteinacious fluid (H & E stain, X100).
However, this patient had a huge multiloculated mass in the left lobe of the liver. The most common clinical presentation is that of a large abdominal mass in an otherwise asymptomatic infant [17]. Vomiting, fever, constipation, diarrhea, weight loss, and respiratory compromise may occur occasionally [5]. Liver function tests are usually within normal limit [18].

Grossly, mesenchymal hamartoma is a large, usually cystic mass that is 12-15 cm in diameter on the average. It may be either predominantly cystic (multiloculated cystic appearance), as in this patient, or predominantly mesenchymal (solid appearance). The lesion may be encapsulated or pedunculated. Histologically, a disordered arrangement of primitive fluid filled mesenchyme, bile ducts, and hepatic parenchyma is presented [18].

On ultrasonography, CT or MRI, a mesenchymal hamartoma usually appears as a large, predominantly cystic lesion with internal septations as shown by CT in this case [7]. The angiography can demonstrate totally avascular mass with mass effect on major vessels in cystic predominant type as this case, to a hypovascular tumor with fine tumor vessels surrounding avascular zones in cases with more solid component [16].

Complete surgical resection is the treatment of choice and can be curative if the mass is completely resectable. Other options include enucleation, as in this patient, or marsupilization into the abdominal cavity [18]. A definite diagnosis is usually made pathologically by the gross and microscopic appearance when the mass is resected. Fine-needle aspiration cytology also plays a role in the diagnosis of mesenchymal hamartoma before surgery [19]. The prognosis is favorable, and the lesion usually can be resected completely.

Mesenteric lymphangioma, duplication cyst, cystic leiomyoma, and enteric cyst all can be excluded from list of differential diagnosis in cystic mass in the pediatric group due to its hepatic origin. Other cystic lesions of hepatic origin, such as teratomas and parasitic or nonparasitic cysts, usually show calcification. Other entities that may manifest as a single multiloculated hepatic mass are abscesses and the rare cystic hepatoblastoma. The former is associated with fever and the later, with elevated α-fetoprotein [16].

UES and malignant mesenchymoma were considered malignant entities of mesenchymal hamartoma in the recent literature [20-23]. Translocation involving 19q13.4 has been identified in both MHL and UES suggesting a histogenetic link between the two tumors [21,24-27]. UES can arise within an MHL [21].

In conclusion, recent advancement of the genetic research has challenge the benign entity of the mesenchymal hamartoma and it is related genetic defect of t(19q)(13.4). Previous separate entity of disease of UES may be actually malignantly transformed from MHL or belongs to the same family. ♦

REFERENCE

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間質缺陷瘤：病例報告

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間質缺陷瘤是一罕見的小兒腫瘤，主要發生在嬰兒及兒童身上，少於二百個病例被報告過。在這篇文章中，我們報告一例間質缺陷瘤的臨床症狀及影像表徵。於 80 年代期間，間質缺陷瘤已被放射界先進回顧過，但 90 年代鮮有進一步的報告，我們希望利用此一病例報告將最近文獻做一個回顧。

關鍵詞：肝臟，間質缺陷瘤；肝臟，電腦斷層攝影；肝臟，血管攝影