

Primary Malignant Fibrous Histiocytoma of the Liver—  
Report of A Case and Review of the Literature

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Abstract

Primary malignant fibrous histiocytoma (MFH) of the liver is a very rare tumor. We present a patient with surgically confirmed primary malignant fibrous histiocytoma of the liver. A 72-year-old man was admitted owing to abdominal fullness and right upper quadrant (RUQ) pain for 1 month. Both computed tomography (CT) and ultrasonography showed a huge tumor located at the right lobe of the liver with central necrosis. Liver biopsy under sonographic guidance revealed a sarcoma of the liver. Histological examination of the resected tumor showed bundles of spindle cells with focal storiform patterns and scattered multinucleated giant cells, which were compatible with MFH. Recent reports in the literature about MFH were reviewed. ( J Intern Med Taiwan 2002;13: 94-98 )

Key Words : Liver, Malignant fibrous histiocytoma

Introduction

Malignant fibrous histiocytoma (MFH) is a common sarcoma of the soft tissue, including primary in extremities, retroperitoneum, head and neck and abdominal cavity<sup>1</sup>. However, MFH of the liver is extremely rare and only 27 cases had been reported since 1987<sup>2,3</sup>. We present a patient with surgically confirmed MFH of the liver and reviewed reports in the literature.

Case Report

A 72-year-old male patient was admitted with abdominal fullness and right upper abdominal pain for about 1 month. Fever and jaundice developed few days prior to admission. He had history of diabetes mellitus for 2 to 3 years with regular management and liver abscess operation 30 years ago. Physical examination on

admission disclosed his body temperature was 37.5°C; icteric sclera and pale conjunctiva were noted. A tender liver was felt five finger breadths below the right costal margin. Laboratory data showed white cell count was 13830/cumm, hemoglobin was 8.7g/dl, serum albumin was 3.0g/dl (reference range (RR), 3.7-5.3g/dl), total bilirubin was 4.6mg/dl (RR, <1.6mg/dl), serum aspartate aminotransferase (AST) was 414U/L (RR, 5-35U/L), serum alanine aminotransferase (ALT) was 230U/L (RR,0-40U/L), lactate dehydrogenase (LDH) was 1840U/L (RR,90-205U/L), alkaline phosphatase was 208U/L (RR,42-128U/L), and gamma-glutamyl transferase (GGT) was 188U/L (RR, 3-60U/L). The hepatitis B surface antigen and antibodies for the hepatitis C virus were negative.

Alpha-fetoprotein was within normal limits. Abdominal sonography showed a heterogenous mass about 10 cm in diameter with peripheral hypoechoic ring over S5-6 (Figure1). Abdominal computed tomography (CT) showed a 10x10x10 cm low attenuation density mass over the right lobe with high attenuation area in the center of the tumor and post contrast peripheral enhancement. The mass was adjacent to the inferior vena cava and right portal vein without retroperitoneal lymph node (Figure 2). The initial differential diagnosis included liver abscess, so antibiotics were prescribed initially and the fever subsided. Sonographic guided liver biopsy and pathological findings revealed liver sarcoma. We consulted a surgeon for surgical intervention. At exploratory laparotomy, a mass measuring 16x10x10 cm, gray in color with capsule located at the posterior aspect of the liver with diaphragm and mesocolon invasion was noted. In addition, many blood clots impacted in the capsule were also noted. No other tumor was found in the abdominal cavity. Hepatic segmentectomy of S5, 6, 7, cholecystectomy and choledochotomy were performed.

The tumor was ill-defined, yellowish brown, and measured 16x10x10cm with focal hemorrhage and necrosis. The main bile duct was free of tumors (Figure 3).

Microscopically, the tumor revealed hypercellular proliferation of fibroblast-like and histiocyte-like cells with scattered osteoclast-like giant cells (Figure 4). Cellular pleomorphism, focal necrosis and increased mitoses were seen. Immunohistochemical study results showed that tumor cells were positive for CD-68 and vimetin stains, and negative for cytoke-ratin, CD-34, alpha-fetoprotein, HHHF-35 and S-100. The pathological diagnosis was MFH of the liver.

The post-operative course was uneventful and adjuvant chemotherapy with mesna, ifosfamide, epirubicin and dacarbazine was performed. The patient is alive and received regular follow-up examinations at the outpatient department for 6 months.

#### Discussion

Malignant fibrous histiocytoma is a rare neoplasm first described by O'rien and Stout 4 in 1964. In previous reports, hepatic MFH was a tumor that occurred during middle

to late adult life, similar to the same tumors that originated from other organs 5. Clinically symptoms included chest pain, right upper or diffuse abdominal pain, weight loss, anorexia, fever, jaundice, malaise, and palpable abdominal mass 6. The size of the tumor at diagnosis ranged from 7 to 24 cm in diameter<sup>3</sup>. Some patients had leukocytosis and abnormal liver transaminases and alkaline phosphatase<sup>7,8</sup>. Abdominal sonography of patients with MFH varied from hypoechoic, mixed echoic or anechoic pattern depending on the pathological changes such as necrosis, hemorrhage or myxoid degeneration. Some tumors had peripheral hypoechoic rings. The CT features of hepatic MFH included low-attenuation mass with poorly defined borders on precontrast scanning. In addition, the solid component showed variable enhancement after contrast injection<sup>2,3,7,9,10</sup>. Magnetic resonance imaging (MRI) of primary hepatic MFH showed heterogeneous intensity on both T1 and T2 weighted image. As extrahepatic MFH, 88% of such tumors had intermediate signal intensity (equal to surrounding muscle) on T1 weighted image and 92% of cases had high signal intensity (equal to or higher than subcutaneous fat) in T2 weighted image<sup>7,9,11</sup>. All tumors with irregular areas of extremely high and low signals represented areas of hemorrhage and necrosis which were heterogeneous on T2-weighted images<sup>7,8,14</sup>. Angiography of hepatic MFH showed a hypovascular to avascular lesion. It is difficult to differentiate from cholangiocarcinoma, metastatic tumor or liver abscess; other differentiated diagnosis may be taken into account including HCC, angiosarcoma and fibrolamellar hepatocarcinoma. Histopathologically, the characteristics of MFH include varying degree of pleomorphism, spindle cells arranged in storiform pattern, mononuclear cells, and multiple nucleated giant cells. Five variants of MFH have been recognized including storiform pleomorphic, myxoid, giant cell, inflammatory and angiomatoid<sup>12,13</sup>. Immunocytochemical stain showed positive results for vimentin, actin, alpha-1, antichymotrypsin and alpha-1 antitrypsin indicated presence of sarcomatous, fibrous, myofibroblastic and histiocytic differentiation. The absence of antigens characteristics for epithelial markers, such as keratin, desmin, alpha-fetoprotein, CEA, S-100, indicated against the origin of epithelial cells<sup>4,6</sup>. However, no markers for epithelial cells were detected in the tumor cells and positive stains for vimentin and CD-68 were in favor of MFH in our case. Surgical treatment alone or combined with chemotherapy has been the mainstay of management for primary MFH of the liver. The prognosis is poor with a 2-year-survival of 60% and recurrence rate of 44%<sup>1</sup>. Mcgrady et al. reported a recurrent patient with survival in good health of 9 years after the initial diagnosis<sup>14</sup>. Fukayama reported a patient with a 4-year survival following hepatectomy, whose tumor showed a relatively marked inflammatory reaction, suggesting that the presence

of inflammation may thus be a sign of better prognosis in MFH of liver 12. Hamasaki et al. reported a patient with MFH of the liver who received liver transplantation and multiple lung metastasis was detected 8 months later, although no evidence of metastasis was found during the initial evaluation. Because micrometastasis is hard to detect, recurrence after liver transplantation frequently occurred 6.

In conclusion, hepatic MFH is a very rare malignant mesenchymal tumor. Its clinical presentations were related to the rapid growth and necrosis of tumor. However, variable features of images make the diagnosis difficult. Pre-operative needle biopsy may be necessary. MFH should be considered when encountering large liver lesions especially for patients with lack of history of chronic viral hepatitis.

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原發性肝惡性纖維組織細胞瘤：  
一病例報告及文獻回顧

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摘 要

肝臟惡性纖維組織細胞瘤為一罕見之原發性肝腫瘤，我們報告一位七十二歲男性，因為腹脹及右上腹痛一個月而住院。於住院期間，腹部電腦斷層及超音波檢查顯示肝右葉有一巨大腫瘤併有中心壞死。實驗室檢查肝指數有異常升高；施予超音波導引切片檢查顯示為肝肉瘤。經手術切除腫瘤，經病理檢查顯示有成束紡錘狀細胞併有局部旋渦狀排列以及散在的多核巨細胞，而確定診斷為肝臟惡性纖維組織細胞瘤。病患於術後接受化學治療，目前仍門診追蹤治療中。



Fig.1. Sonography shows a huge heterogenous mass, about 10 cm in diameter with a peri-peral hypochoic margin over S5-6.

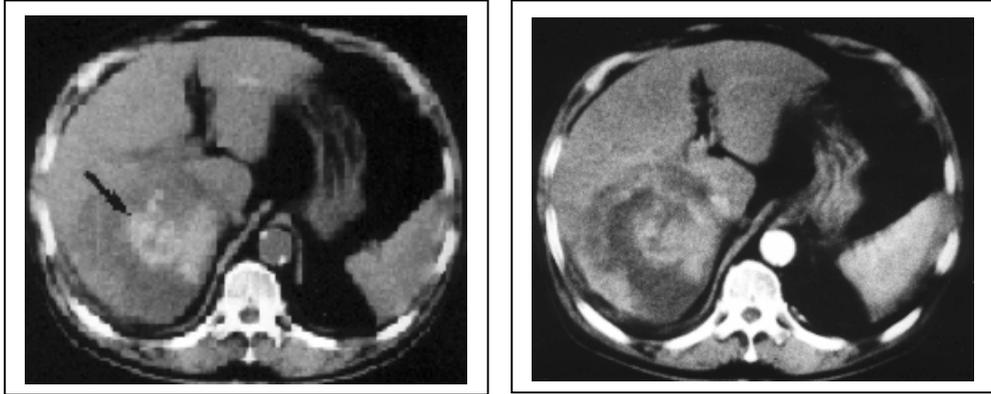


Fig.2. Left: CT scan shows a 10x10x10 cm low attenuation density mass over right lobe with high attenuation density area in the center representing hemorrhage (arrow). Right: Peripheral enhancements of the tumor on post-contrast scan.

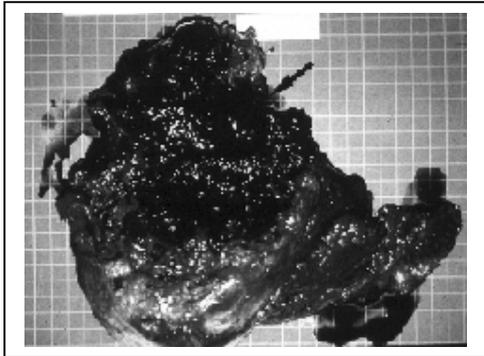


Fig.3. Gross specimen: an ill-defined yellowish brown tumor mass, measuring 16x10x10 cm. Foci of hemorrhage and necrosis are present. (arrow)

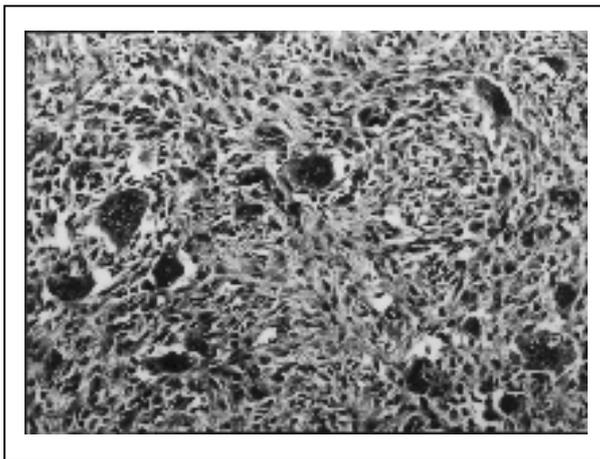


Fig.4. Microscopically, the tumor shows cellular proliferation of fibroblast-like and histiocyte-like cells admixed with osteoclast-like giant cells.(H&E ,x150)