

**Primary Hepatic Leiomyosarcoma:
A Case Report and Review of The Literatures**

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Abstract

Primary hepatic leiomyosarcoma (PHL) is exceedingly rare, and only few cases have been sporadically reported in the literature. Herein, we present a case of PHL in a 50-year-old male who had epigastric pain and hepatomegaly. This giant PHL had prominent cystic necrosis on ultrasonography. Computed tomography revealed no other tumor in abdomen. Surgical resection was performed and no mass in abdomen was reviewed. He died of recurrent tumor in the left lobe of liver 21 months after operation. It may be the first case report in Taiwan. PHL occurred more commonly in the right lobe. The most common symptom and sign were right upper quadrant pain and hepatomegaly. The employed therapy included surgery, systemic chemotherapy, combination of surgery and chemotherapy, and conservation treatment. Clinical diagnosis of this tumor is difficult. While in case of a giant liver tumor with an image providing the characteristic figure of extensive necrosis, PHL should be considered.

Keywords: primary hepatic leiomyosarcoma

Introduction

Metastatic leiomyosarcomas in liver are not common. It usually comes from gastrointestinal tract, female genital system and lung (1). While, primary hepatic leiomyosarcoma (PHL) is a very rare tumor of liver. To our knowledge, there are only 67 case reports in the world literature, and no report was found in Taiwanese literature. Herein, we present a patient who had a giant PHL and may be the first report in Taiwan.

Case report

A 50-year-old male had suffered from sharp and intermittent epigastric pain for one month. He had no history of liver disease and alcohol abuse. Physical examination revealed a marked hepatomegaly extending seven finger-breadths below the right costal margin. On admission, laboratory analysis revealed aspartate aminotransferase 32 IU/L (0-34 IU/L), alanine aminotransferase 16 IU/L (0-40 IU/L), alkaline phosphatase 161 IU/L (28-94 IU/L), γ -glutamyl transpeptidase 73 IU/L (0-26 IU/L), total bilirubin 0.8 mg% (0-1.3 mg%), and hemoglobin 9.5 gm% (13.5-17.5 gm%). Other laboratory values, including albumin, prothrombin time, white blood cell count, platelet, α -fetoprotein, and carcinoembryonic antigen, were normal. Antibody to hepatitis C virus was positive and hepatitis B surface antigen was negative. Abdominal computed tomography (CT) done outside our hospital showed a huge multi-loculated, heterogeneously hypodense tumor with well demarcated margin occupying the whole right lobe of the liver (Fig. 1A). Abdominal ultrasonography revealed a heterogeneous mixed-echoic mass with cystic components, at least 14cm in diameter, in right lobe (Fig.2). Selective angiography of the celiac trunk and superior mesenteric artery showed a huge hypovascular tumor in the right lobe of the liver (Fig. 1B). Chest X-ray, small bowel series and lower gastrointestinal series all revealed negative findings.

Surgical exploration was undertaken. A huge tumor, measured 20cm × 18cm × 12cm, occupying almost the whole right lobe of the liver with compression to middle hepatic vein was removed by right hepatic trisegmentectomy with cholecystectomy. Careful inspection of the abdominal and pelvic contents did not find any other masses or lesions. Grossly, the tumor was lobulated, well encapsulated, and prominent in fibrotic bands and cavities (Fig.3). Pathological examination revealed this mass to be a leiomyosarcoma that is composed of dense spindle cells with mitosis (Fig.4A). Immunohistochemical study showed a positive desmin stain in the cytoplasm of tumor cells (Fig.4B). Recurrent tumor in left liver was found 12 months later. An abdominal CT showed no other tumor outside the liver. However, he died of recurrent disease 21 months after operation.

Discussion

Hepatic tumor of mesenchymal origin is rare. It accounts for only 1.2% of all primary hepatic tumors (2). The most common type of primary malignant mesenchymal tumor is angiosarcoma, representing 36%, followed by leiomyosarcoma (12%), fibrosarcoma (7%), and unspecified sarcoma (44%) (3). Most of hepatic leiomyosarcoma are metastatic from other sites of leiomyosarcoma including gastrointestinal tract, uterus, retroperitoneum and lung (1,4). So, exclusion of metastatic leiomyosarcoma in the liver is an essential event in diagnosing primary lesion.

Primary hepatic leiomyosarcoma may arise from intrahepatic vascular structures (5), bile ducts (6) or ligamentum teres (7). The tumors arising from the hepatic veins may develop Budd-Chiari syndrome and have worse prognosis than those from other origins (4). Eight cases arising from the ligamentum teres of the liver were reported. All of them were resectable and had the best prognosis in this group (7). The tumors originating from the bile ducts could cause cholestatic liver function test (6). Because our case had a giant tumor size,

the origin of this tumor could not be traced. Our case located in the right lobe of the liver. Among 67 cases reported, 63 patients could identify the location of tumor. 31 cases occurred in the right lobe, 17 cases were in the left lobe, 13 cases were in bilateral lobes, and two cases were in medial lobe. Metastasis occurred in 24 patients. The most common metastatic site was lung and/or pleura, and other metastatic sites were diaphragm, adrenal gland, kidney, gut, bone, peritoneum, skin, retroperitoneum, and lymph node (4,8).

For 67 cases in the world literature, patient's ages ranged from 9 to 86 years, with a mean age of 54 years (55.9 in male, 48.7 in female). The male to female ratio was 33 to 34. The most common symptom was right upper quadrant pain. The most common sign was hepatomegaly (4,8). Other complaints were weight loss, anorexia, epigastric fullness, jaundice, distended vein, and ascites. None of the 67 patients had a history of hepatitis or cirrhosis. One patient had been exposed to thorotrast 50 years earlier (9).

Liver function test were variable, therefore, not useful in diagnosis. Carcinoembryonic antigen and α -fetoprotein were normal. Diagnostic imagings readily showed abnormal results but were seldom diagnostic. Different ultrasonic patterns were observed in tumors with different sizes. Generally, a homogeneous hypoechoic pattern was found in tumors less than 10cm in diameter (1,6,7); and a hypoechoic or mixed-echoic pattern with necrotic zones were showed in tumors more than 15cm in diameter (3,5,10,11). The larger tumors have a great tendency to outgrow their blood supply and easily cause central necrosis with cavitation and hemorrhage (12). Our case had a giant tumor about 20cm \times 18cm \times 12cm and showed massive necrosis with cavitation and hemorrhage. The ultrasonogram of our case presented a figure as the cut-surface of tumor tissue. (Fig.1 and 3). Soyer et al. described the tumors on CT scan showed a large, well-delineated hypodense mass, frequently with a predominantly peripheral enhancement, and occasionally displaying central necrosis or cystic appearance (13). Angiography showed a vascular tumor in five patients and a relatively avascular mass in

two others, including our patient. The magnetic resonance images displayed homogeneous hypointensity on T1-weighted images and inhomogeneous hyperintensity on T2-weighted images (5,10).

Grossly, the tumor size ranges from 7 to 36cm in diameter. The cut surface is variable but usually pinkish-white with yellow areas of necrosis or dark red hemorrhagic foci. Histologically, the tumor is composed of intersecting bundles of elongated, spindle-shaped cells. Nuclei are hyperchromatic and elongated (2). Immunohistochemistry stains are positive for desmin, vimentin and smooth muscle actin, and negative for keratin, S-100 protein, and neuron-specific enolase (2, 14)

Therapy had been performed in 55 patients including surgical excision (29 patients), systemic chemotherapy (10 patients), combination of surgery and chemotherapy (5 patients), and conservative treatment only (11 patients). It is difficult to make comment about the treatment because all of the reported cases have variable follow-up and therapy. However, surgical excision is a better choice which offers a hope for long-term survival (1,3,7,8,15,16). Chemotherapy is the alternative treatment for inoperable patients and the precise role has to be clarified (4,17). The mean survival of untreated intrahepatic leiomyosarcoma is about 10 months. With a combination of surgery and chemotherapy, the mean survival is 3.3 years (8).

Primary hepatic leiomyosarcoma is rare. Its prognosis is poor. The clinical diagnosis of this tumor is difficult, especially in case of small tumor. However, in case of a giant hepatic tumor which presents extensive necrosis or cystic change in images, the diagnosis of primary leiomyosarcoma should be taken into consideration.

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Figure legends

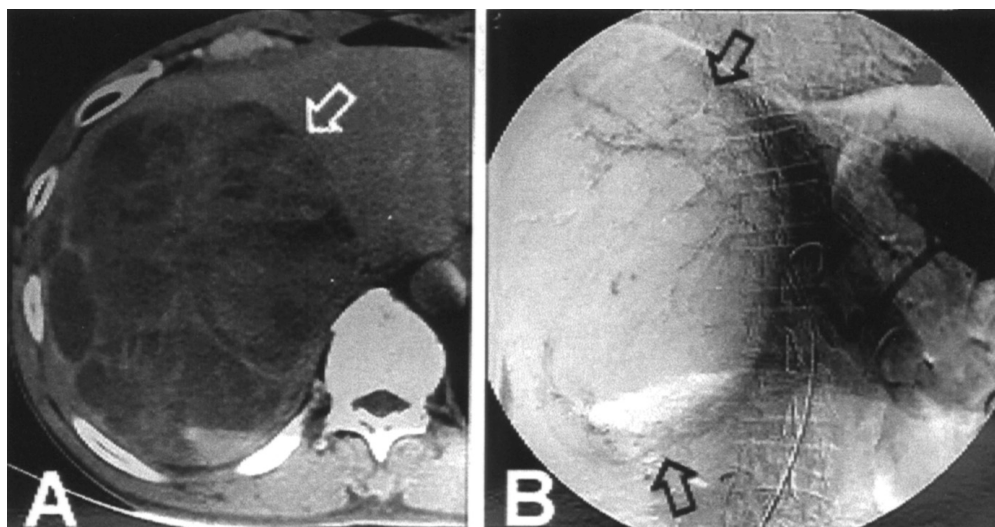


Fig. 1. (A) Contrast-enhanced CT scan demonstrated a well-demarcated margin, multi-lobulated, heterogeneous hypodense mass with peripheral enhancement in right lobe of liver (arrow). (B) Liver angiography shows a huge hypovascular tumor in right lobe of liver (arrows).

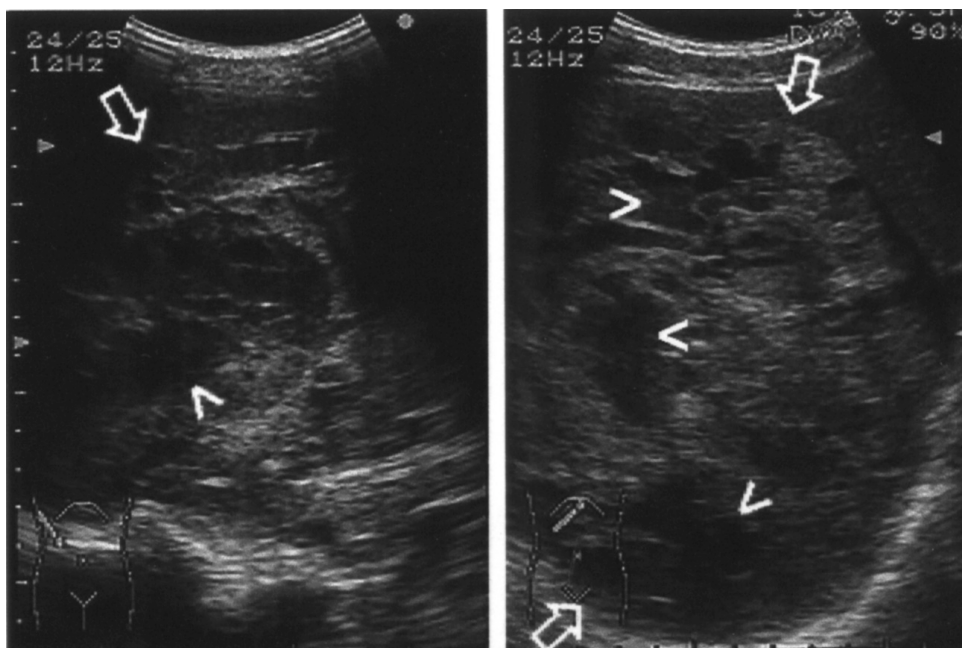


Fig. 2. Right: Abdominal ultrasonography from right subcostal approach revealed a heterogeneous mixed-echoic mass (arrows) with many cystic components (arrowheads) in right lobe. The diameter of mass is at least 14 cm. The sonographic figure is correspondent to the cut-surface of tumor tissue in figure 3. Left: From right intercostal approach, sonography revealed tumor in tumor with cavitation (arrowhead).

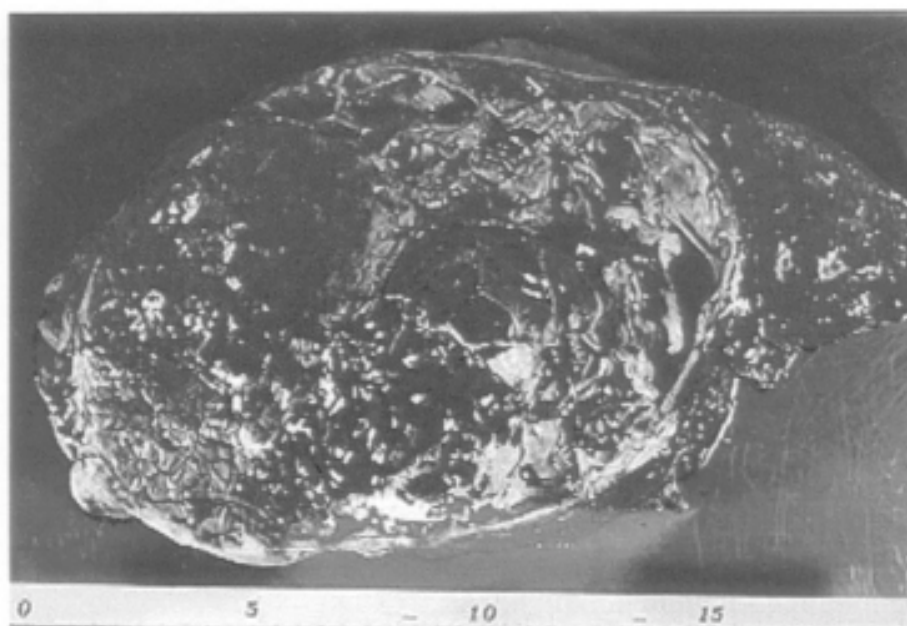


Fig. 3. Surgical specimen shows an encapsulated giant tumor with extensive necrosis, hemorrhage and cavitation separated by dense fibrotic bands.

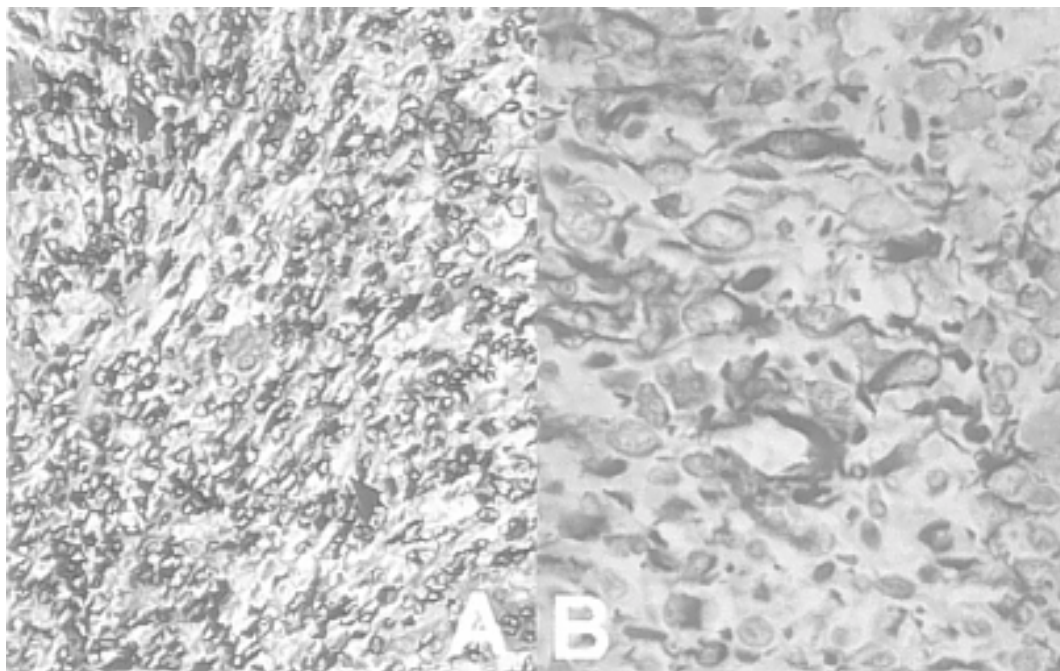


Fig. 4. (A) Hematoxylin-eosin staining of the tumor (original magnification $\times 200$) demonstrates interlacing bundles of spindle-shaped cells with pleomorphic and hyperchromatic nuclei, and mitotic figures (arrowhead). (B) Immunohistochemical study with desmin stain showed positive staining in the cytoplasm of tumor cells (original magnification $\times 400$)