Solitary fibrous tumors (SFTs), also known as localized fibrous tumors, are a rare mesenchymal neoplasm. The classic presentation of SFTs is that of a pleura-based tumor, although they have been reported at various sites. SFTs occur mainly in adults and are slightly predominant in women [1-3]. Hypoglycemia has been reported in patients with SFT. We report on SFT in the liver of a patient who presented with a huge hepatic tumor and severe hypoglycemia. We found that the image features and associated hypoglycemia were useful in making a correct diagnosis of this rare hepatic tumor.

CASE REPORT

A 78-year-old male patient had suffered from anorexia and body weight loss for 6 months. He was transferred to our hospital in November 1999 due to a liver mass revealed by ultrasonography in another hospital. On physical examination, a large mass was palpated in the right upper quadrant of the abdomen. Severe hypoglycemia of 34 mg/dL was found on laboratory examination. Abdominal ultrasonography and computed tomography (CT) showed a huge, well-demarcated mass in the liver. The patient underwent exploratory laparotomy and S4, S7, and S8 segmentectomy. The blood sugar rapidly returned to normal level. The postoperative period was uneventful, and there has been no evidence of tumor recurrence during 18 months of follow up.

Marked hepatomegaly with elevation of the right hemidiaphragm was identified on chest roentgenography and plain abdominal films. Abdominal ultrasonography showed a huge intrahepatic mass larger than 15 cm in diameter with heterogeneous echogenicity (Fig. 1). On unenhanced CT scans, the intrahepatic mass was...
well demarcated and measured about 20 cm in diameter. It appeared as low attenuation mixed with isodense areas. No calcifications or areas of hemorrhage were identified (Fig. 2a). Contrast enhancement showed displacements of the hepatic arteries by this tumor mass and scanty tumor vessels in the arterial phase (Fig. 2b). Images in the portal venous phase showed multiple heterogeneous contrast-enhanced areas (Fig. 2c).

The pathologic gross specimen showed the tumor to be well circumscribed, firm, lobulated, gray white with a whirling fasciculated pattern, and measuring 21.0 x 20.0 x 18.0 cm (Fig. 3). Focal myxoid, cystic change and necrosis were present. There was no area of extracapsular extension in this well-circumscribed encapsulated tumor.

Microscopically, the tumor was composed of a mixture of spindle-shaped fibroblast-like cells and collagenous stroma. The cells were arranged in a “patternless pattern”. Foci of cystic degeneration, myxoid change, and necrosis were identified. Cellular atypia was mild, and rare mitotic figures were identified. Immunohistochemical staining for CD34 showed

Figure 1. Abdominal ultrasonogram showing a huge intrahepatic mass larger than 15 cm with heterogeneous echogenicity.

Figure 2. CT of a solitary fibrous tumor. a. Unenhanced image showing low attenuation mixed with isodense areas. No calcification or hemorrhage was identified. Focal areas with a myxoid matrix in the tumor are identified (white arrows). b. Arterial phase image showing displacement of the hepatic arteries by this huge tumor and scanty tumor vessels. The focal myxoid matrix of the tumor showed no significant enhancement (white arrows). c. Portal venous phase images showing heterogeneous enhancement. Some areas with marked enhancement in this phase correspond to fibroblast-rich tissues (arrowheads). The myxoid matrix of the tumor still shows no significant enhancement (white arrows). Some areas with slight enhancement in the arterial and portal venous phases are the collagenous stroma (black arrow).
DISCUSSION

A solitary fibrous tumor (SFT) or localized fibrous tumor, first described in 1931 [4], is a pleura-based neoplasm that seems to originate from the submesothelial connective tissue [5]. SFTs have also been described in extrathoracic locations such as the upper and lower respiratory tract, orbit, salivary glands, liver, breast, soft tissues, peritoneal cavity, retroperitoneum, thyroid, meninges, heart, etc [3]. SFTs occur in a broad age group between the second and seventh decades and are slight predominant in women [2, 3]. The etiology and pathogenesis of SFTs are still unknown. SFTs are usually benign neoplasms, but atypical and malignant SFTs have been reported in some series [5, 6].

Few cases of SFT in the liver have been reported in the English-language literature. The clinical features of SFT in the liver include a palpable mass, abdominal pain, nausea, body weight loss, symptoms of cholecystitis, hypoglycemia, and incidentally detected abnormal findings on liver function tests. The incidence of hypoglycemia in patients with SFT has been reported to be as high as 25% [5]. Insulin-like growth factor (IGF-II) is regarded as the cause of SFT-associated hypoglycemia [7, 8]. This phenomenon has been referred to as non-islet cell tumor hypoglycemia. The most effective therapeutic approach for SFT and SFT-associated hypoglycemia is to resect or debulk the tumor [9].

On gross pathologic findings, the tumor is well-circumscribed and firm, with a gray-to-white appearance of various sizes from 2 to more than 20 cm in greatest dimension. Some authors regard SFTs as any tumor located adjacent to or attached by a pedicle to the liver capsule without infiltration into the liver parenchyma [1, 10]. However, it is difficult to identify the pedicle when the SFT is huge. Histologically, the tumor is composed of dense hyalinized areas with collagen interspersed with spindle cells resembling fibroblasts; this is the so-called patternless pattern [1, 11]. The absence of cellular atypia, mitoses, and/or necrosis is a feature of benign SFTs. There are marked cellular atypia and mitotic figures varying from 2 to 4 mitoses per 10 high power field in 2 of 9 cases of SFT of the liver in a series of Moran et al. [1]. Immunohistochemically, SFTs show a strong positive reaction against antibodies for CD-34 and vimentin [1]. CD-34 is highly specific for SFT.

There are no typical radiological characteristics to differentiate SFTs from other hepatic tumors. Radiological findings of slight enhancement in arterial phase images and relatively obvious heterogeneous enhancement in portal venous phase images on post-contrast CT scan suggest the abundance of fibrous tissue in SFTs. Valls et al. regarded SFTs of the liver as hyperenhancing focal liver lesions, especially in the portal vein and equilibrium phases of post-contrast CT scans [12]. Some areas of the portal venous phase of the patient’s CT scan showed marked enhancement compared with those in the arterial phase (Fig. 2b and c). Those areas with marked enhancement in the portal venous phase CT scan corresponded to fibroblast-rich tissues determined pathologically. However, some areas had slight post-contrast enhancement either in the arterial or portal venous phase images. These areas were collagenous stroma as correlated with pathological features (Figs. 2, 3). A focal myxoid matrix could be seen in the SFT. The myxoid matrix showed low attenuation on the pre-contrast CT scan and no significant post-contrast enhancement in the arterial and portal venous phase images (Fig. 2a, b, and c). The attenuation values of the myxoid matrix ranged from 25 to 45 HU. Calcification or ossification is rare in SFTs.
Similarly, no calcified density was identified by CT scan in our patient’s SFT. The common differential diagnoses include atypical appearance of hepatocellular carcinoma, hepatic adenoma, giant hemangioma, metastatic tumor, inflammatory pseudotumor, and other rare mesenchymal tumors such as angiosarcoma, fibrosarcoma, and other sarcoma groups, especially in cases with prominent fibrous tissues. A preoperative diagnosis is made with difficulty and is usually erroneous. The clinical presentation of associated hypoglycemia may remind us of the possibility of SFT. However, a definitive diagnosis of SFT should be made on the basis of histopathologic and immunohistochemical findings [6, 10, 13, 14].

In summary, SFT should be included in the differential diagnoses of a solitary huge hepatic tumor with a well-demarcated margin, slight post-contrast enhancement in the arterial phase, and relatively obvious uneven enhancement in the portal venous phase images of CT scans. The clinical presentation of associated hypoglycemia provides an important clue for making a correct diagnosis of a solitary fibrous tumor preoperatively. The strong immunoreactivity for CD34 is specific and a hallmark of SFT.

REFERENCES

肝臟單一纖維瘤：一病例報告

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肝臟單一纖維瘤是一種罕見的腫瘤。我們報告一例合併嚴重低血糖的肝臟單一纖維瘤的病例。超音波檢查可以看到具有不均勻回音和邊界明顯的巨大腫瘤。在靜脈注射顯影劑後的電腦斷層影像上，腫瘤在動脈相時期只有些微的顯影，而在肝門脈相時期卻有明顯但不均勻的顯影。根據肝臟內的大型腫瘤、影像檢查的表現和臨床上表現有低血糖，肝臟單一纖維瘤應該要列入鑑別診斷之中。

關鍵詞：單一纖維瘤，局部性纖維瘤，肝臟腫瘤，低血糖，CD34