Case Report
Hepatic fibrosarcoma in a middle-aged man

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Abstract: Hepatic fibrosarcoma (HF) is a rare sarcoma with a high malignancy and a poor prognosis. Moreover, it is hard to diagnose before completing a pathological examination, for HF has almost no features of clinical or imaging manifestations. Here we report a case of HF in a 42-year-old male who complained of pain in the right upper abdomen. Computed tomography (CT) and ultrasonography confirmed a large mass was occupying the right lobe of his liver. The patient was finally diagnosed with HF based on the morphology and immunohistochemistry of the tumor after resection. This case indicates that a diagnosis of HF should be considered, especially when the results of imaging examinations and tumor markers do not support the common hepatic diseases.

Keywords: Liver, hepatic fibrosarcoma, diagnosis

Introduction

Hepatic fibrosarcoma (HF) is a kind of rare malignant tumor which is derived from hepatic fibroblasts. Usually, fibrosarcoma occurs in the limbs, the head and neck, and the trunk. Rarely, it originates from the visceras, such as the kidneys, the ovaries, and the breasts, but the most common location is the lungs [1-4]. According to the previous reports, the HF lesions are usually located in the right lobe of the liver [5]. HF patients generally are male and range from 27 to 87 years old, with no features found at the tumor serological examination or in the imaging examinations, making it easily confused with other liver diseases [6, 7]. Therefore, a diagnosis of HF largely depends on the pathological examination after an operation [8]. Here we mainly aim to present a case of HF, a middle-aged man with a giant mass occupying the right lobe of his liver, and we review the pathological and imaging characteristics of HF.

Case report

A 42-year-old male, with pain in the right upper portion of his abdomen, was found to have a large mass in the right lobe of his liver for more than one month, by CT scan. A series of examinations were conducted after he was hospital-ized. Through the physical examination, mild pain was reflected in his hepatic region, but no abnormal symptoms were found elsewhere. The results of laboratory examinations showed the indexes were within normal limits, including tumor markers such as abnormal prothrombin of 14.17 mAU/ml, alpha fetoprotein of 1.44 ng/ml, and carcinoembryonic antigen of 2.29 ng/ml. An abdominal CT scan revealed that a quasi-circular, low density lesion was found inside the enlarged right lobe of his liver, and it stretched to the liver surface. The volume of low-density area was measured at approximately 17.3 × 12.5 × 16.8 cm, and the patchy necrosis in the lesion was distinctly in the center of the liver (Figure 1A). Further an enhanced CT scan with dynamic contrast enhancement showed that when the right hepatic artery was enhanced at the arterial phase, the lesion showed an inhomogeneous enhancement and was also enhanced peripherally, while the reinforcement level was decreased at the portal phase and phase equilibrium. The necrosis area of the lesion was not enhanced, and there was no abnormal density or enhancement in any other area of the liver (Figure 1B). Also, no signs of filling defect were detected in the intrahepatic vessels; the intrahepatic bile duct was not dilated, so the gallbladder only had a slight-
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A slight hypoechoic mass in the right lobe of the liver measured 17.0 × 13.5 cm and had an irregular shape and a clear border (Figure 1C). Color Doppler flow imaging (CDFI) showed that few blood flow signals were in the lesion, and the echoes in the other areas of the liver were rough and inhomogeneous; the intrahepatic vessels were compressed, and the intrahepatic bile duct was not dilated. Part of the focal lesion was hyper-enhanced in inhomogeneity at the arterial phase and low-enhanced at the portal and equilibrium phases, but the other lesion was not enhanced in any of the three phases (Figure 1D). Further, no abnormal enhancement was found in the other liver area at the equilibrium phase. Ultimately, the patient had a complex hepatectomy in his right hepatic resection as well as a cholecystectomy. The histopathology after the operation showed that the size of the large mass was 22.5 × 19.5 × 15 cm in the right lobe of the liver, the section of which was yellow-white and multinodular. A microscopic examination found that the tumor was composed of spindle cells and had a large area of necrosis and fibrosis (Figure 2). The Glisson’s capsule was invaded but not completely broken through. An immunohistochemistry analysis showed the tumor was positive for vimentin and 60% Ki-67, but negative for CK, GPC3, CD34, Dog-1, CD117, S-100, EMA, BCL-2, SMA, desmin, actin, CD68, and CD99 (Figure 3). Based on the above examination, the patient was eventually diagnosed with HF.

Discussion

Hepatic malignancy is one of the most important causes of morbidity and mortality in the Chinese population [9]. There are several path-
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Figure 3. The results of immunohistochemistry showed the lesion was positive for vimentin and 60% Ki-67, but negative for CK, GPC3, CD34, Dog-1, CD117, S-100, EMA, BCL-2, SMA, desmin, actin, CD68, and CD99 (200 × magnification).

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The clinical manifestations seen in HF patients usually depend on the location and the size of the lesion. Some have simultaneous liver carcinoma and liver dysfunction. Most of the HF patients only have upper abdominal pain with no other symptoms, and cirrhosis is not present in every patient. In this case, the condition was similar to the previous one. The patient was a middle-aged man, only complaining of an ache in the area of the liver, and the tumor markers AFP and CEA were within normal limits. To sum up, the clinical manifestations of this patient were non-specific, so the HF was difficult to diagnose.

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a hypervascular mass and arteriovenous shunting under enhanced scanning; ultrasonography indicated that the lesion had clear borders and irregular morphology; the sizes of the lesions varied depending on whether we used the hypoecho or the mixed echoic pattern [10, 12, 13]. Liver scanning with 99mTc showed a large defect of uptake in the right lobe of the liver [14]. The CT scan and ultrasonography in the present case accordingly showed a large mass occupying the liver with an inhomogeneous enhancement inside. The border of the lesion was enhanced and clear, but the CDFI showed few blood flow signals in the lesion. So, it seemed the image of the lesion could not provide a definite diagnosis. Meanwhile, by reason of its low incidence and nonspecific presentation, it was hard to differentiate it from primary hepatic carcinoma or other non-tumor benign disease. However, the diagnosis of HF should be taken into account for the liver mass, especially when the indicators do not support a common hepatic tumor.

The definite diagnosis of HF depends on a pathological examination at present. The pathological and histological characteristics of fibrosarcoma include tumor cells that are spindle-shaped and round or oval nuclei containing one or more nucleoli with atypical division; the cytoplasm is scant, and the cell boundary is not obvious. Often, poorly differentiated tumors make more profuse collagenic fibers. The arrangement of the tumor cells can be in a herringbone or fishbone pattern. When the cells are highly malignant, the liver capsule can be damaged, resulting in an invasion of the adjacent liver tissue. Immunohistochemistry is essential for the differential diagnosis of HF. The tumor is positive for vimentin, and negative for desmin, actin, Syn and SMA could be diagnosed as HF. In the present case, the postoperative pathological portrait of the patient corresponds to the above description. The lesion was composed of spindle cells, with necrosis and fibrosis; the capsule of the liver was invaded but not broken through completely. Immunohistochemistry showed that the tumor was positive for vimentin and 60% Ki-67. Based on pathological information of this patient, HF was the definite diagnosis.

Currently, surgery is considered the most effective treatment for HF, and for patients with infiltration or metastasis, postoperative chemotherapy is recommended. It has been reported that the prognosis of HF might be associated with tumor size and R0 resection [15].

Conclusion

In summary, hepatic fibrosarcoma is extremely rare, with a high malignancy and a poor prognosis. The clinical manifestations are not specific, and the patients often complain of pain in the abdomen and have hepatomegaly. Imageology usually indicates a large mass in the liver, often with hemorrhaging, necrosis and cystic degeneration. A liver puncture may effectively help the diagnosis before the operation. Even though the definite diagnosis of HF is clinically dependent on a pathological examination, the diagnosis of HF should be considered when the serum tumor markers, like AFP and CEA, are normal but with a focal liver lesion.

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Disclosure of conflict of interest

None.

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