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Low-grade myofibroblastic sarcoma of the liver misdiagnosed as a cystadenoma: A case report

Jie Li, Xin-Yue Huang, Bo Zhang

Abstract

BACKGROUND

Low-grade myofibroblastic sarcoma (LGMS) is a rare malignant tumor. It has no specific clinical manifestations and commonly occurs in the head and neck, extremities and other body parts, with the liver not as its predisposing site.

CASE SUMMARY

This is a case report of a 58-year-old man with right upper abdominal pain for 11 d. Contrast-enhanced computed tomography (CECT), CE magnetic resonance imaging and CE ultrasound (CEUS) all showed a cystic-solid mass in the right liver. As the initial clinical diagnosis was a hepatic cystadenoma, a surgical resection was performed, and the postoperative pathology indicated a hepatic LGMS. The 3-mo follow-up showed a favorable recovery of the patient. However, at the -month 7 of the follow-up, two-dimensional US and CECT showed a suspected metastatic lesion in the right-middle abdomen of the patient.

CONCLUSION

Hepatic myofibroblastic sarcoma is easily misdiagnosed as it has no specific clinical and imaging manifestations, and surgical resection is the preferred treatment. Pathological examination is the gold standard for the diagnosis of hepatic myofibroblastic sarcoma. More reported cases will contribute to the understanding of the disease and improve the accuracy of diagnosis.

Key Words: Myofibroblastic sarcoma; Liver; Cystic-solid mass; Imaging; Diagnosis; Case report

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Core Tip: Myofibroblastic sarcoma is a rare malignant spindle-shaped cell tumor derived from mesenchymal tissue, and those occurring in the liver are particularly rare to see. Pathological examination is the gold standard for the diagnosis of hepatic myofibroblastic sarcoma. As the discrepant biological characteristics of different lesions and discrepant protocols for the surgical treatment, we should pay attention to more information conducive to differential diagnosis in order to improve the preoperative diagnosis, and to choose an appropriate surgical approach.

INTRODUCTION

Myofibroblastic sarcoma is a rare mesenchymal spindle-shaped cell tumor, first discovered by Mentzel *et al*[1] in 1998. According to the differential degree of myofibroblasts, it can be divided into low-, intermediate- and high-grade. The first two are collectively called low-grade myofibroblastic sarcoma (LGMS)[2]. Myofibroblastic sarcoma is common in the head and neck, limbs and trunk, but those in the liver are extremely rare. So far, there are only four English literature reports available. Here, we present a case of LGMS of the liver.

CASE PRESENTATION

Chief complaints

Right upper abdominal pain for 11 d.

History of present illness

The patient, a 58-year-old Chinese male, presented with right upper abdominal pain without obvious predisposing factors 11 d prior to the visit. The symptom was not associated with eating and body position. He experienced no nausea or vomiting, no chills or fever, and no significant weight loss.

History of past illness

Right inguinal hernia surgery had been performed at another hospital 3 years prior to the present visit.

Personal and family history

No history of hepatitis and family or genetic history was claimed.

Physical examination

Physical examination showed a flat and soft abdomen without tenderness and rebound pain, and no palpable mass was found. The rest of the physical examination showed no abnormalities.

Laboratory examinations

Cholesterol (5.3 mmol/L) and low density lipoprotein (3.78 mmol/L) were slightly elevated. Blood routine, liver function, renal function and prothrombin were normal. Complete quantitative detection of hepatitis B was negative. Alpha-fetoprotein, carcinoembryonic antigen and carbohydrate antigen were normal.

Imaging examinations

Abdominal contrast-enhanced computed tomography (CECT) and hepatic vascular imaging showed that there was a cystic-solid hypodense mass with a size of approximate 99 mm × 92 mm in the right liver. Septa, which were slightly enhanced along with the solid parenchyma, were visible in the mass. The right hepatic vein was displaced due to the compression of the mass supplied by the right hepatic artery

(Figure 1). Abdominal CE-magnetic resonance imaging (CE-MRI) + diffusion weighted imaging (DWI) showed: (1) There was a cystic-solid mass with low signal on T1WI and high signal on T2WI in the right liver, about 101 mm × 98 mm in size; (2) there were multiple uneven septa and fluid-fluid levels in the lesion; (3) DWI and ADC showed high signal and high-b-value, respectively; and (4) after enhancement, solid components and internal septa of the tumor were significantly enhanced accompanied by unenhanced cystic components (Figure 2).

Ultrasound (US) examination showed: A 123 mm × 98 mm mixed-echo mass with regular shape and clear boundary was detected in the right liver. The anechoic dark area was the main portion of the mass. The parenchymatous septa with uneven thickness were visible in the mass in which no obvious parasitic body sonogram was found. Color Doppler flow imaging showed punctate blood flow signals in the mass. CEUS showed that the peripheral and internal septa of the tumor were hyper-enhanced in the arterial phase, iso-enhanced in the portal phase and delayed phase, and unenhanced in the anechoic area in these three phases (Figure 3). Based on the above-mentioned examination, the mass was preliminarily identified as hepatic cystadenoma.

FINAL DIAGNOSIS

The postoperative pathology diagnosis of the presented case is LGMS of right liver (Figure 4).

TREATMENT

The patient underwent laparoscopic right hepatectomy under general anesthesia after preoperative investigations were completed.

OUTCOME AND FOLLOW-UP

The patient recovered well after the operation with the surgical incision healed uneventfully. No apparent event was observed at the postoperative 3-mo follow-up. However, at the 7-mo follow-up, two-dimensional ultrasound and CECT showed a 100

mm × 40 mm cystic-solid lesion occurring in the right middle abdomen of the patient, which showed the similar property as a liver lesion. Meantime, the mass wall was enhanced after enhancement (Figure 5). The patient refused further surgery although the clinician initially believed that metastasis probably occurred.

DISCUSSION

Myofibroblastic sarcoma is a rare histological type of hepatic sarcoma. Given its rareness, the etiology is not clear temporarily, and most of the relevant reports are case reports or case series. Currently, only seven relevant case reports[3-9] were obtained after a comprehensive literature search was conducted by us in CNKI, PubMed and Web of Science databases, involving 7 patients (4 men, 3 women; aged 25-38 years, n = 5; > 60 years, n = 2). As described in WHO classification, male were slightly more predisposed for this disease than female, thus not surprisingly the present case is male. Most of the myofibroblastic sarcomas were identified due to the swelling lesion detected or during physical examination. In the literature reports, 5 of the 7 patients presented with abdominal distension and pain, and another 2 patients were asymptomatic. As for the present patient, the complaint was right upper abdominal pain. For the patient reported herein, no history of hepatitis was elicited, although 2 of the 7 patients reported previously had a history of hepatitis B. Serological examination, such as blood routine, liver function, renal function, and alpha-fetoprotein and other tumor markers such as carcinoembryonic antigen, carbohydrate antigen 199 as well, all of which were in the normal range, were available for 6 of the 7 patients reported previously and for the patient reported herein. The remaining patient reported previously had impaired liver function. The lesions reported were mostly located in the right liver, of which cystic-solid tumor was found in 5 cases, and no relevant data were available in another 2 cases. For the present case, a cystic-solid tumor located in the right liver was also observed. Among all of the 7 cases reported previously, 5 were misdiagnosed as liver cancer or hydatid disease as their preliminary clinical diagnosis, while the present case was misdiagnosed as hepatic cystadenoma. Moreover, surgical resection was

performed in 6 of the 7 patients, and liver lesions were found and biopsy was performed at 3 mo after operation for the remaining patient initially diagnosed as retroperitoneal inflammatory myofibroblastic tumor. The pathological diagnosis for the patient was LGMS infiltrated in the liver. Currently, the surgical resection of myofibroblastic sarcoma is the preferred treatment, although primary treatment, prognosis and follow-up treatment are not clear. More data are needed to obtain reliable conclusions about the role of follow-up treatment such as radiotherapy and chemotherapy.

No specific clinical signs and symptoms were established currently due to the low incidence of hepatic myofibroblastic sarcoma, which was generally manifested as abdominal distension or discomfort and/or abnormal liver function, sometimes accompanied by sweating and weight loss. Although there was no clear report on tumor recurrence and metastasis, most of hepatic myofibroblastic sarcomas were observed as cystic-solid masses in the literature we reviewed, showing infiltrative and destructive growth.

At present, hepatic myofibroblastic sarcoma is mainly diagnosed by histopathology and immunohistochemistry. Under light microscopy, the tumor tissue of the present case was mainly composed of spindle-shaped cells with a diffuse infiltrative distribution. The cytoplasm was eosinophilic and the nucleus was polymorphic. Abnormal mitotic figures were present. The histopathological findings of this case are consistent with those cases previously reported above. In the immunohistochemical study of the cases reported previously, an expressed vimentin, positive smooth muscle actin (SMA) and Desmin, and partially positive Actin, is commonly observed, although no hepatocyte markers, epithelial markers, S100, CD117, CD34, myogenin, CK-Pan and h-caldesmon were not found in the tissue[10]. Additionally, in the immunohistochemical study of the case reported herein, a positive vimentin, SMA and desmin, and negative HePar-1, Glypican-3, CK-Pan, myogenin, Ard-1, MYOD1, SOX10, and CD31, was found, with an approximately 10% of a Ki-67 index. As can be seen, the present case is mostly similar to the cases reported previously for this profile.

To date, due to lack specific manifestations in imaging examinations, hepatic myofibroblastic sarcoma is easily misdiagnosed as liver cancer, hepatic hydatid disease, cystadenoma and other space-occupying lesions, and it is difficult to make a predisposed diagnosis in clinical practice as well. The liver myofibroblastic sarcoma reported herein is mainly observed as a cystic-solid mass with well-defined borders and regular morphology. The cystic components within the mass were separated by septa of varying thickness, with a partially visible fluid-fluid level. After enhancement, the solid portion and septa of the mass were enhanced to different degrees, with unconspicuous regression for the degree and velocity.

In addition, the CEUS of this case showed that the peripheral and internal septa of the tumor were hyper-enhanced in the arterial phase, iso-enhanced in the portal phase and delayed phase, while in 2 of the 7 cases reported in the literature, the portal phase and delayed phase were hypo-enhanced in both. Hence, the CEUS features of liver myofibroblastic sarcoma require more case data to accumulate.

Liver myofibroblastic sarcoma needs to be differentiated from liver cancer (particularly with liquefaction and necrosis), hepatic hydatid disease, and cystadenoma. When liver cancer is accompanied by liquefaction and necrosis, three-phase non-enhanced areas in the lesions may also be found, but most of which are patchy or irregular. Moreover, the substantial portion of the mass shows high enhancement in the arterial phase, and low enhancement in the portal and delayed phases. The process behaves in a fast-in and fast-out mode. In contrast, the non-enhancing area of hepatic myofibroblastic sarcoma lesions was separated by the internal septa of the mass, and the parenchyma and internal septa of the mass were not significantly regressed after enhancement. The patient with liver hydatid disease generally have a history of living in pastoral areas or exposure to pathogens. The typical imaging manifestations are mainly cystic masses, showing the mother and daughter sporocysts, with varying degrees of eggshell-like cyst wall calcification, intracystic streamer sign, lily sign, and cyst-within-cyst. Liver cystadenoma are also cystic-solid masses with predominantly cystic components, along with mural nodules (papillary-solid components protruding

into the cystic cavity on the cyst wall) as specific signs. Mural nodules show nodular enhancement in the portal phase and can continue to enhance in the delayed phase. Imaging examination can clearly show the size, location, border, number, blood supply, and internal and surrounding adjacent tissues of the tumor. However, there is currently no characteristic diagnostic basis for the imaging manifestations of hepatic Myofibroblastic Sarcoma.

The presented case was misdiagnosed for several reasons. First, the patient seldom had specific clinical history, presentation and abnormal laboratory indexes. Second, the preoperative imaging images in this case were similar to the cystadenoma. The internal separation of the mass was relatively uniform, the blood flow of the mass was not abundant, and there was no washout after enhancement. Therefore, imaging diagnosis was limited to benign lesions, and malignant behavior was not considered.

To sum up, myofibroblastic sarcoma of the liver has no characteristic manifestations in clinical performance, serological indicators and imaging findings. Therefore, it has an unmet demand to accumulate a large number of cases with complete data to provide meaningful research data for the diagnosis and treatment strategies of myofibroblastic sarcoma of the liver.

CONCLUSION

In word, due to the rarity of liver myofibroblastic sarcoma, more clinical data and in-depth research are needed to further analyze its pathological mechanism, biomarkers, and imaging manifestations. Improving the understanding of the imaging manifestations of liver myofibroblastic sarcoma will help improve the preoperative diagnosis rate and assist clinicians in performing precise surgical resection and follow-up treatment.

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Figure Legends

Figure 1 Title. A: The right hepatic cystic-solid mass is supplied by the right hepatic artery, and the twisting of the supplying artery can be seen; B and C: The portal venous phase and the delayed phase, respectively; the solid portion and the septum are enhanced, and the intracapsular septum is clearly displayed.

Figure 2 Title. A and B: The lesions were low signal on magnetic resonance imaging (MRI) T1WI, and high signal on MRI T2WI, with multiple septa of uneven thickness and liquid-liquid levels; C: After enhancement, the solid components and septa of the mass were significantly enhanced, but the cystic components were not enhanced.

Figure 3 Title.

A:

B:

C:

D:

In the contrast-enhanced ultrasound, the peripheral parenchyma and internal septa of the mixed-echo mass showed hyper-enhancement in the arterial phase, and iso-enhancement in the portal phase and delayed phase.

Figure 4 Title. A: A cystic-solid mass was seen with the naked eye, with soft texture and inconspicuous tumor capsule, its cut surface is polycystic, with yellowish jelly-like substances inside cysts, and the cyst wall thickness is 1 mm-12 mm, no cirrhotic changes were observed in the peripheral liver tissue (arrow); B and C: Spindle-shaped cells are

arranged in fascicles under light microscope, eosinophilic cytoplasm with atypia and mitotic figures can be seen.

Figure 5 Title. A and B: The right middle abdomen of the patient in contrast-enhanced computed tomography showed a cystic-solid lesion, with the similar property as a liver lesion. After enhancement, the mass wall was enhanced.

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