Clinical Case Report

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Primary hepatic malignant fibrous histiocytoma combined with invasion of inferior vena cava

A case report and literature review

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Abstract

Rationale: Malignant fibrous histiocytoma (MFH), primary presented in liver, was very rare and displayed a poor prognosis because of high aggression. As a few of cases had been reported merely, we shared the case of primary hepatic MFH combined with invasion of inferior vena cava (IVC).

Patients concerns: A 69-year-old women presented with abdominal pain.

Diagnoses: Abdominal computed tomography and magnetic resonance imaging indicated a soft mass about 5.4 × 4.2 cm in the caudate lobe, accompanied with IVC invaded.

Interventions: After the multidisciplinary consultation, laparotomy was performed, followed by chemotherapy and radiotherapy. Primary hepatic MFH was demonstrated pathologically. Till now, the patient was alive for >22 months after surgery and no evidence of recurrence or distant metastasis was suspected.

Outcomes: We discussed the integrated procedure of diagnosis and treatment, combined with data from literature review.

Lessons: To our knowledge, the primary hepatic MFH combined with invasion of IVC was hardly reported. Despite the poor prognosis, the comprehensive treatment integrating the surgery, chemotherapy, and radiotherapy showed the satisfactory disease-free and overall survival. However, further investigations are definitely warranted.

Abbreviations: CT = computed tomography, IVC = inferior vena cava, MFH = malignant fibrous histiocytoma.

Keywords: case report, inferior vena cava, malignant fibrous histiocytoma, pathology

1. Introduction

The malignant fibrous histiocytoma (MFH), an ordinary soft tissue sarcoma, was first described by O'Brien and Stout in 1964,[1,2] which presented in extremities frequently, less commonly in posterior peritoneum.[3,4] Till now, only a handful of these case reports could be recorded through the relative literature.[3–7] Furthermore, no successful comprehensive treatment of primary hepatic MFH combined invasion of inferior vena cava (IVC) was reported, except the case of primary hepatic MFH combined invasion of IVC but dead of pulmonary embolism, was published by Schweyer et al.[8]

Therefore, we shared a case of comprehensive treatment for primary hepatic MFH with invasion of IVC, with a terrific disease-free and overall survival.

2. Case presentation

A 69-year-old women was admitted to our hospital as having recurrent upper abdominal pain for about half a year, aggravated for a week. The patient denied fever, cough, vomiting, or diarrhea in recent period. In terms of previous history, the hypertension was stabilized at 140 mmHg more or less by daily taking oral medicines for 15 years. Besides, well-controlled Hepatitis B for >10 years with regular antiviral therapy was acknowledged. However, the level of blood glucose was indistinct since the diabetes was diagnosed last year.

On physical examination, general condition of patient was well-preserved. The vital signs were stable, and the lung auscultation revealed no rales. Except a slight tenderness in epigastrium and bilateral lower limbs edema, there was no other significant finding.

Laboratory tests showed the white blood cell count of 8.1×10^9 cells/L, the hemoglobin of 111 g/L, and the platelets count of 145×10^9 cells/L. Liver function indicated mildly elevated alanine aminotransferase (58 U/L) and gamma glutamyl transpeptidase (93 U/L), whereas a degressive albumin of 34.8 g/L. Viral serology revealed that HBsAg was positive, corresponding to the history of

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Informed Consent: The data of case were evaluated and approved by the Ethical Committees for Human Subjects at Sir Run Run Shaw Hospital affiliated to medical college of Zhejiang University. Informed consent, paper-based form, was agreed by the patient and her families.

The authors report no conflicts of interest.

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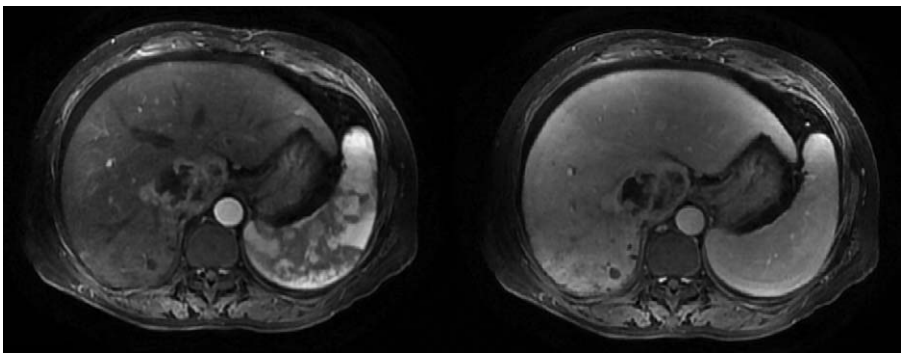

Figure 1. Liver enhanced magnetic resonance imaging showed a large lesion with low signal in the second hilum on T2-weighted images. The left is arterial phase, circular irregular intensification in the fringe of the mass was observed and the largest size of the tumor was 5.8 × 4.8 cm in dimension. The right is venous phase, the mass presented rapidly attenuation of signal and "Space Occupying Effect" in the inferior vena cava.

hepatitis B infection. Tumor makers including carcinoembryonic antigen, carbohydrate antigen 19–9, and alpha fetal protein were negative. Arterial blood gases showed no signs of anoxia or acidosis.

Furthermore, abdominal ultrasound indicated a mass adjacent to the second porta hepatis of the liver. An enhanced computed tomography (CT) revealed a hypodense mass (5.4 × 4.2 cm, CT values from 5 to 35 HU) in the caudate lobe, accompanied with IVC invaded, and cholecystolithiasis. Enhanced magnetic resonance imaging demonstrated similar results that soft mass presented rapid intensification and attenuation, "Space Occupying Effect", and cancerous embolism of IVC formed (Fig. 1). However, the heart was tumor-free on echocardiography. Moreover, chest x-ray and CT pulmonary angiography showed no positive signs, although the electrocardiogram revealed sinus rhythm and atrioventricular conduction delay.

After multidisciplinary consultation, the patient diagnosed as having hepatocellular carcinoma initially underwent an laparotomy by surgical team (see supplement firgue 1, http://links.lww.com/MD/B732). In operation, cancerous thrombus was monitored by the esophagus cardiac ultrasound. Meanwhile, we confirmed the tumor was restricted in the left caudate lobe, closed with the IVC but with tumor-free of atrium dextrum. Therefore, the transabdominothoracic left caudate lobe resection, holecystectomy, and embolectomy of IVC were preformed ultimately. First of all, dissociating the entire liver followed by exposing the caudate lobe as possible as we could. Then cardiac surgeon exposured the hepatic superior IVC with thoracotomy procedure. Eventually, transiently blocking the IVC, removing the tumor and cancerous thrombus integrally, and suturing the IVC rapidly. Afterwards, the patent recovered smoothly, and discharged at 30th day after the operation.

Grossly, the specimen measuring 7.2 × 4.0 × 2.5 cm presented a gray-to-white fleshy mass about 4.0 × 2.0 × 2.3 cm filled with necrotic debris and blood clots (Fig. 2). Pathologically, the primary hepatic MFHwas confirmed. Microscopically, the tumor

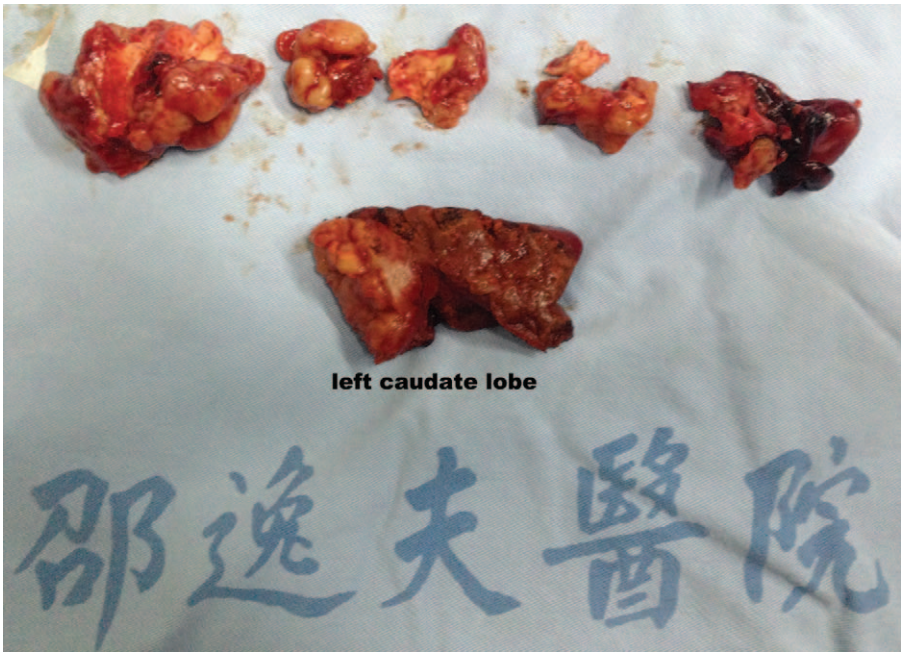

Figure 2. Surgical specimens contained left caudate lobe and cancer embolus of inferior vena cava.

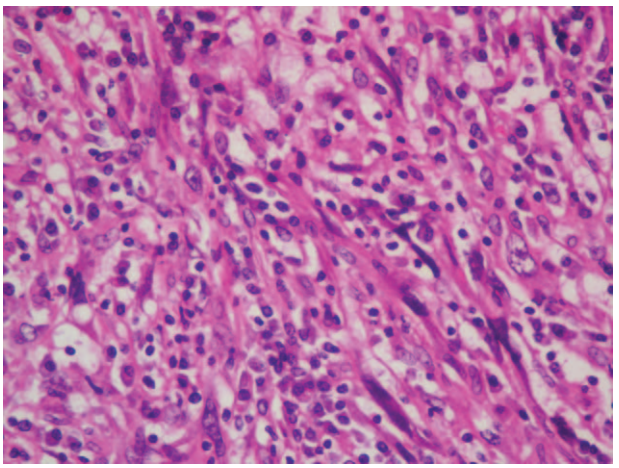

Figure 3. Pathological examination showing storiform-pleomorphic spindle cells (hematoxylin and eosin original magnification ×400).

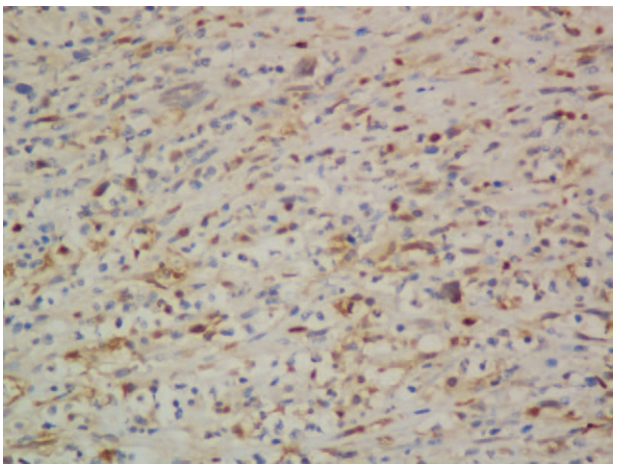

Figure 5. Immunohistological staining for α1-antichymotrypsin demonstrating positive reaction in the tumor cells (original magnification ×400).

was consisted of spindle cells arranged in a storiform pattern and contained various amounts of polymorphic cells. In addition, incomplete fibrous intervals were observed mixed with necrotic and hemorrhagic area around the tumor (Fig. 3). Immunohistochemically, CD-68 (Fig. 4) and α1-antichymotrypsin (Fig. 5) were positive, whereas CK, calretinin, CD-117, CD-99, CD-34, SMA, S-100, and Desmin were negative.

A month later, the patient started to receive the chemotherapy as an adjuvant therapy in local hospital. The formula was the ifosfamide 3.0 g from 1st day to the 5th day plus liposomal doxorubicin 60 mg in the 1st day initially. Subsequently, the patient received a lower dose of following 5 periods of chemotherapy (ifosfamide 3.0 g from 1st day to the 4th day plus liposomal doxorubicin 40 mg in the 1st day) because of the severe bone marrow inhibition reaction with grade 4, but rapidly recovered with treatment of granulocyte-macrophage colony-stimulating factors. Additionally, a targeted radiotherapy (Experientially, 10MV–X SAD100 DT200cGy/1F 1st, DT600cGy/3F 5th, DT1600cGy/8F 12th, DT2600cGy/13F 19th, DT3600cGy/18F 26th, DT4600cGy/23F 34th,) around the invaded IVC was performed prophylactically without adverse effect.

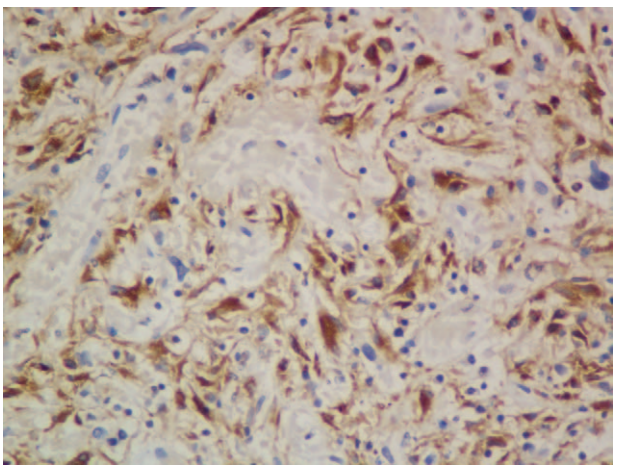

Figure 4. Immunohistological staining for CD68 demonstrating positive reaction in the tumor cells (original magnification ×400).

Follow-up was carried on every 3 months. The physical examination, serum tumor markers, and abdominal enhanced CT were performed routinely. Till the November 2016 (see supplement Figure 2, http://links.lww.com/MD/B732), the patient was still alive over 22 months, even the recent abdominal enhanced CT showed no evidence of recurrence or metastasis (Fig. 6).

2.1. Literature review

A search of the database Pubmed, according to the "Histiocytoma, Malignant Fibrous"[Mesh], and ("liver" or "hepatic" or "hepato"), was performed. While unrelated to hepatic HMF, or published not in English, were excluded. Full-text articles, including case reports, literature review, letters, editorials, as well as opinion articles, were assessed for eligibility. Reference lists of relevant articles were reviewed and duplicates or information uncomplete cases were removed.

Totally, 40 related literatures were searched, while published not in English (6), abstract only (3), unrelated to hepatic MFH (20), or information (overall survival time) uncomplete cases (1) were excluded. Ten literatures were eligible,[3,6,9–16] which contained 41 cases of hepatic MFH with overall survival (Table 1). Among the 41 cases confirmed pathologically, there were 23 males and 18 females. Generally, the 1- ,3- ,and 5-year overall survival rates were 51.9%, 25.6%, and 16.2%, respectively

3. Discussions

Theoretically, the diagnosis of MFH depends on an accurate differential diagnosis from other sarcomas, which expresses specific surface molecules such as vimentin, CD-68, and α1-antichymotrypsin.[4,9–10] However, the histopathologic concept of MFH including storiform-pleomorphic, myxoid, inflammatory, giant cell, and angiomatoid variants had been eliminated; concomitantly World Health Organization denominated most of the MFH as undifferentiated pleomorphic sarcomas in 2002.[17]

The typical clinical manifestation of primary hepatic MFH contained abdominal pain, jaundice, fever, malnutrition, or asymptomatic.[4,6,11,18,19] Literature review illustrated that hepatic MFH was lack of classical tumor makers and imaging

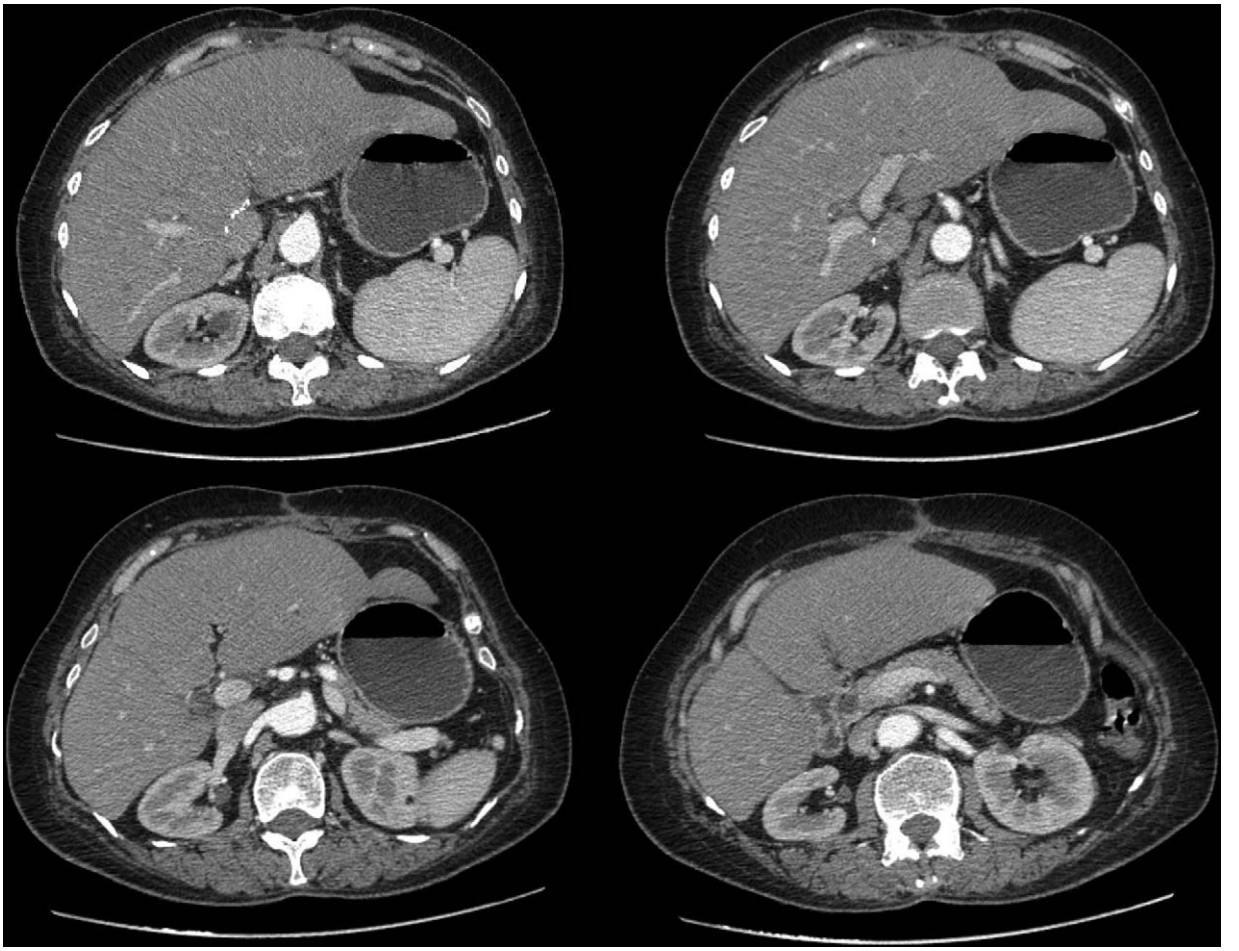

Figure 6. In November 2016, the patient received enhanced computed tomography scan, which showed no evidence of recurrence or metastasis.

Table 1

Literature review of hepatic malignant fibrous histiocytoma.

Published date/author	Article types	No. of cases	Age/sex	Tumor location	Overall survival
1985/Alberti-Flor et al	Case report	1	59/M	Left and right lobe	14 days
1985/Conran et al	Case report	1	61/M	Left and Right lobe	18 days
1986/Fukayama et al	Case report	1	38/F	left lobe	4 y/alive
1987/Arends et al	Case report	1	78/F	Left and right lobe	6 days
1988/Bruneton et al	Case report	2	52/F	Right lobe	2 y/alive
			34/M	Right lobe	6 mo
1988/Honda et al	Case report	1	71/F	Right lobe	4 mo
1988/Katsuda et al	Case report	1	61/M	Right lobe	6 mo
1991/Hamasaki et al	Case report	1	35/M	Left lobe	34 mo
1992/Akifuji et al	Case report	1	79/M	Left lobe	5 mo
1992/Zornig et al	Case report	1	36/F	Liver	63 mo/alive
1992/McGrady et al	Case report	1	53/F	Left lobe	9 y/alive
1993/Reed et al	Case report	1	52/M	Right lobe	2 mo
1994/Pinson et al	Case report	1	41/M	Left and right lobe	10 mo
1998/Fujita et al	Case report	1	70/M	Left and right lobe	3 mo
1998/Ferrozzi et al	Case report	1	62/F	Right lobe	3 y/alive
2002/Chou et al	Case report	1	72/M	Right lobe	6 mo/alive
2005/Anagnostopoulos et al	Case report	1	87/F	Right lobe	6 mo
2006/Ding et al	Case report	1	50/M	Left and right lobe	2 mo
2006/Su et al	Case report	1	38/F	Liver, heart, and right Humerus	3 mo
2007/Ye et al	Case report and literature review	1	50/M	Left lobe	4 mo
2007/Chen et al	Case report	1	70/M	Right lobe	1 mo
			77/F	Right lobe	1 y
			54/M	Left lobe	4 y
2008/Li et al	Case series and literature review	6	34/F	Liver and lung	4 mo
			80/F	Right lobe	1 y
			46/F	Right lobe	3 mo/alive
			70/M	Right lobe	4 y
2009/Sugitani et al	Case report	2	45/F	Left lobe	34 mo
			70/F	Right lobe	8 mo
2009/Kim et al	Case report	1	60/M	Right lobe	41 mo/alive
2010/Caldeira et al	Case report	1	63/M	Right lobe	45 days
2010/Julien et al	Case report	1	45/M	Pancreas, liver, and lung	3 y/alive
2012/Yao et al	Case report a nd literature review	1	56/M	Right lobe	6 mo/alive
2012/Yan et al	Case series	7	44/(4M/3F)	Liver	15 mo

Age is presented as mean in case series; data are presented as survival time and only alive are shown in overall survival. d = days, mo = months, y = years.

presentation clinically.[19,20] In terms of treatment, surgical resection with negative margin remained the optimal choice,[3,8] although the median survival without distant metastasis was 8.5 months.[21] While, either chemotherapy or radiotherapy, as an adjuvant therapy, was benefit of local recurrence but not prolong overall survival, or was preferred for the patient with distant metastasis.[8,22–25]

In the present case, the chief complaints were abdominal pain and chest distress. Radiology work-ups merely manifested a soft mass near to the second hilum and invasion of IVC, but were unable to identify the MFH from hepatocellular carcinoma or other malignancies, corresponding with literature published.[3,11]

According to the immunohistochemical results, sarcomatoid carcinoma (CK-negative), malignant mesothelioma (Calretinin-negative), gastrointestinal stromal tumor (CD-117-, CD-99-negative), angiosarcoma (CD-34-negative), leiomyosarcoma (Desmin-negative), malignant peripheral nerve sheath tumor and melanoma (S-100-negative), and rhabdomyosarcoma (SMA-negative) were ruled out. Meanwhile, CD-68, KP-1, and α1-antichymotrypsin were positive, conforming to the feature of MFH.[4,9,10] Thus, MFH with margin negative was confirmed pathologically.

Besides the radical resection, chemotherapy and targeted radiotherapy, as adjuvant treatments, were carried out. Till now, the patient was alive for >22 months and no evidence of recurrence or distant metastasis was suspected clinically.

4. Conclusions

In conclusion, primary hepatic MFH possessed a high aggressive behavior and poor prognosis. However, the comprehensive treatment integrating the surgery, chemotherapy, and radiotherapy displayed a terrific short-term result, and presented the potential to improve the disease-free and overall survival. Further investigations are warranted, definitely.

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