

Case report**Malignant fibrous histiocytoma of the spleen accompanying with hepatic cyst:  
report of a case and review of the literature**

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**Abstract**

Primary malignant fibrous histiocytoma (MFH) of the spleen is extremely rare. Since the first description of primary splenic MFH reported by Govoni *et al* in 1982, to the best of our knowledge, only 13 cases of MFH of the spleen have been reported in the English language literature in Medline. We herein report a rare case of primary splenic MFH accompanying with hepatic cyst in a 48-year-old Chinese female who treated successfully by laparoscopic splenectomy and fenestration, which has not yet been reported in the literature. Compared with the 13 previously cases of MFH of the spleen, our case is the first case accompanied with other disease, and also the first case treated successfully by laparoscopic splenectomy. A literature review of MFH of the spleen previously reported in the English language literature in Medline is also provided.

**KEYWORDS:** Malignant Fibrous Histiocytoma, Hepatic Cyst, Laparoscopic Splenectomy.

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**M**alignant fibrous histiocytoma (MFH) is a soft tissue sarcoma mainly occurring in the soft tissues, especially in the extremities and trunk, and other less common sites include the retroperitoneum, and the head and neck.<sup>1-3</sup> It is an aggressive malignancy with high potential of local recurrence and distant metastases, and surgery with radical resection of the primary tumor with histological negative margins is the treatment of choice even with recurrence or metastasis.<sup>1, 4, 5</sup> MFH of the spleen is extremely rare, although the incidence of splenic MFH remains undetermined, only 13 cases have been reported in the English language literature in Medline.<sup>6-17</sup> Because of its rarity, the clinical characteristics and biological potentials of MFH of the spleen are yet to be determined.

We herein report a 48-year-old Chinese female with MFH of the spleen accompanying with hepatic cyst, which has not yet been reported in the literature. To the best of our

knowledge, this case is the first case of MFH of the spleen accompanied with other disease, and also the first case treated successfully by laparoscopic splenectomy. A literature review of MFH of the spleen previously reported in the English language literature in Medline is also provided.

**Case Report**

A 48-year-old Chinese female was admitted to Department of Oncological Surgery (Zhejiang Cancer Hospital, Hangzhou) with left upper abdominal pain, accompanying with belching and weight loss of 2.5 kg over the previous 3 months. Prior to this presentation, she had been healthy, without any previous medical illness. On physical examination, the left upper quadrant was slight tender but no abdominal mass was detected by abdominal palpation. Laboratory data were all normal. Computed tomography (CT) demonstrated a low-density mass with a CT value of 1 Hounsfield Unit

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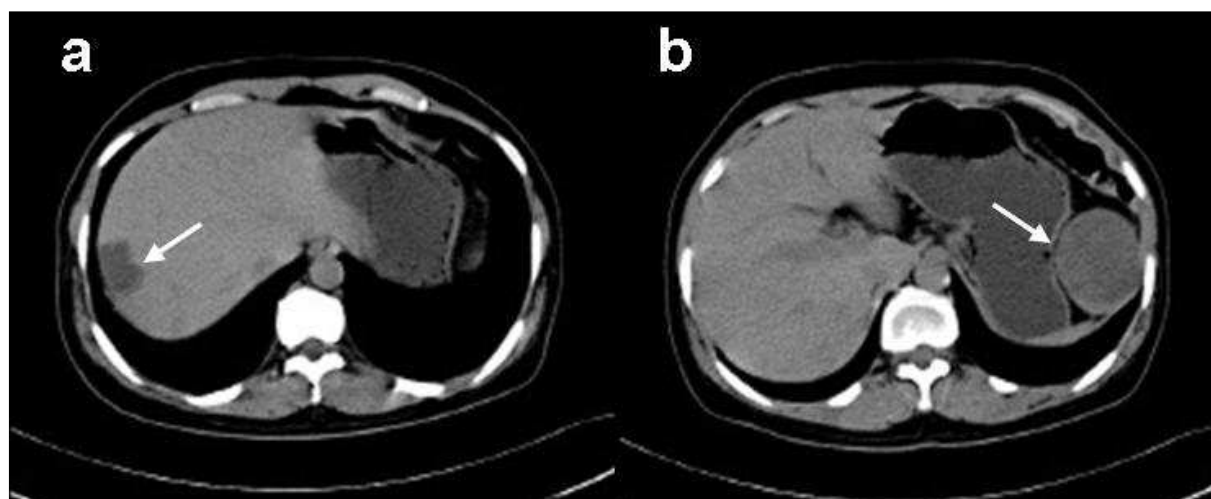
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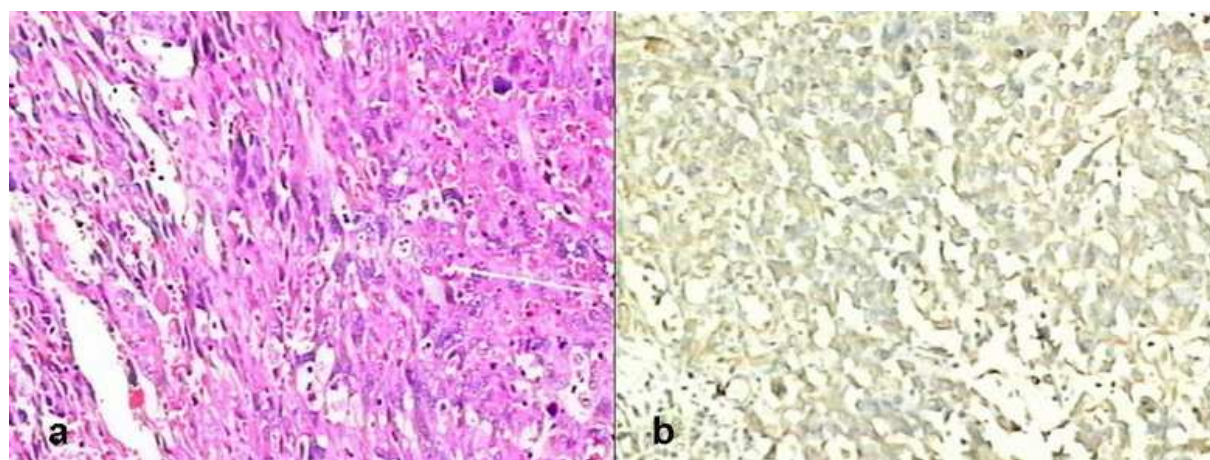
(HU), measuring about  $3.2 \times 2.2$  cm in the right lobe of liver (Figure 1A), and a solid low-density mass with a CT value of 8HU, measuring about  $5.2 \text{ cm} \times 4.6 \text{ cm}$  in the upper pole of the spleen (Figure 1B).

Based on the physical examination and CT, the preoperative diagnosis of the hepatic cyst was definite but the preoperative diagnosis of the splenic tumor was difficult. Then a laparoscopic splenectomy was performed under a presumed diagnosis of a malignancy of the spleen. Intraoperatively, a solid mass approximately 5 cm in diameter was found in the upper pole of the spleen. Intra-abdominal

lymph nodes and the adjacent organs looked normal. There were not any signs of metastases or any visible neoplastic tissue remnant at the operation site. Laparoscopic fenestration of the hepatic cyst was also performed. The resected spleen measured  $11 \times 10 \times 8$  cm, weighed 740 g. Histopathology showed that tumor cells were pleomorphic and consisted of fibroblasts, histiocytes, and myofibroblast-like cells arranged in a storiform pattern (Figure 2A). Immunohistochemistry showed these tumor cells were positive for vimentin (Vim),  $\alpha$ -smooth muscle actin ( $\alpha$ -ACT) and CD68 (Figure 2B). Based on these results of histopathology



**Figure 1.** CT demonstrated a low-density mass measuring about  $3.2 \text{ cm} \times 2.2 \text{ cm}$  in the right lobe of liver (A), and a solid low-density mass measuring about  $5.2 \text{ cm} \times 4.6 \text{ cm}$  in the upper pole of the spleen (B).



**Figure 2.** Histopathology showed tumor cells were pleomorphic and consisted of fibroblasts, histiocytes, and myofibroblast-like cells arranged in a storiform pattern (A). Immunohistochemistry showed tumor cells were positive for CD68 (B). (HE: 200 $\times$ )

and immunohistochemistry, the final diagnosis of MFH of the spleen was conformed. The post-operative course of this patient was uneventful. The patient was discharged on the 7th day after the initial operation and was not given any additional treatment. She was followed up for 13 months without recurrence or metastases after the initial surgery at the time of reporting.

## Discussion

MFH is the most common soft tissue sarcoma mainly occurring in the extremities (70%), followed by the abdominal or retroperitoneum (16%).<sup>1</sup> The origin of MFH is still controversial. Primary MFH of the spleen is extremely rare, although the incidence of MFH of the spleen remains undetermined. Since the first description of primary splenic MFH reported by Govoni *et al.*<sup>6</sup> in 1982, only 13 cases have been reported in the English language literature in Medline.<sup>6-17</sup> Compared with the 13 previously reported cases of MFH of the spleen, our case was the first case of MFH of the spleen accompanied with other disease, and also the first case treated successfully by laparoscopic splenectomy.

The spleen, for poorly documented reasons, generally shows relative resistance to malignant tumors. Accordingly, the preoperative diagnosis of MFH of the spleen is difficult because the cases are extremely rare, the clinical symptoms are nonspecific, and the imaging patterns are not suggestive. The final diagnosis is confirmed by the result of the histopathology and immunohistochemistry. These tumor cells consist of histiocyte-like and fibroblast-like cells with other multinucleated giant cells.<sup>1,13,18</sup> There are positive stainings for lysozyme, Vim, CD68, and  $\alpha$ -1 antitrypsin.<sup>13</sup> In our case, we could not get a definite preoperative diagnosis of the splenic tumor according to the physical examination and CT scanning. A final diagnosis of MFH of the spleen was confirmed by the histopathology and immunohistochemistry.

In review of the English language literature in Medline, we found 13 previously reported cases of the MFH of the spleen (Table 1). Among the 14 cases of MFH of the spleen, in-

cluding our case, six cases were females and eight cases were males with the age from 35 to 82 years old, mainly in the fourth or fifth decade of life (9/14). The presenting symptoms were usually nonspecific, but the clinical characteristics such as abdominal pain, fever, weight loss, and splenomegaly were prominent features in about 70% of the patients.<sup>17</sup> About 35.7% (5/14) of the cases with splenic MFH presented metastases at the initial surgery including liver (3 cases), stomach and pancreas (1 case), and omentum (1 case); and the liver was the most common metastatic site. MFH has been categorized into five types, based on the histopathologic subtype, including storiform-pleomorphic, myxoid, inflammatory, giant cell and angiomatoid variants.<sup>1,6-9,13</sup> However, only the subtype of storiform-pleomorphic and inflammatory variants have been reported in the MFH of the spleen in the previously reported literature. In review of the 14 cases, although 4 cases were not mentioned, eight cases were storiform-pleomorphic types including our case and two cases were inflammatory types. All of the 14 patients underwent the surgery of splenectomy, while our case was the first case treated successfully by laparoscopic splenectomy. Four cases were also treated by adjuvant chemotherapy and/or radiotherapy. The survival time was mentioned in 13 cases. Six patients died of local recurrence with multiple metastasis, 3, 6, 7, 8, 15, and 19 months postoperatively. One patient was dead after splenectomy with hepatic involvement and the other was alive at 18 months with multiple hepatic metastases at the time of reporting. Five cases, including our case, survived for 3, 13, 17, 18, and 46 months, without local recurrence and distant metastasis at the time of reporting.

## Conclusion

The incidence of splenic MFH remains undetermined due to limited data on a rare malignancy. The clinical symptoms of primary splenic MFH are usually nonspecific, but the characteristics such as abdominal pain, fever, weight loss, and splenomegaly are prominent

**Table 1.** Reported cases of malignant fibrous histiocytoma of the spleen in the English language literature in Medline

References	Published year	No.	Sex/ Age	Splenic weight (g)	Clinical characteristics	Metastatic site	Treatment	Histological subtype	Survival (m)
Govoni <sup>6</sup>	1982	1	F/51	1400	Abdominal pain, weight loss, splenomegaly	No	Splenectomy	Storiform variant	7 (Alive without metastasis)
Wick <sup>7</sup>	1982	2	M/48	1184	Fever, weight loss, fatigue, splenomegaly	Hepatic metastasis	Splenectomy Chemotherapy	Inflammatory variant	18 (Alive with multiple hepatic metastasis)
			M/54	1235	Abdominal pain, splenomegaly	No	Splenectomy	No data	3 (Alive without metastasis)
Bruntton <sup>8</sup>	1988	1	M/54	No data	Abdominal pain, melena, gastrointestinal symptoms	Gastric, pancreatic involvement	Splenectomy	No data	3 (Died with extensive metastasis)
Sieber <sup>9</sup>	1990	1	M/41	1850	Fatigue, weakness, rib and back pain, night sweats, exertional dyspnea, weight loss, splenomegaly	Omentum, intra-abdominal metastasis	Splenectomy Omentectomy Radiotherapy Chemotherapy	Storiform variant	6 (Died with extensive intra-abdominal metastasis)
					Abdominal pain, weight loss, splenomegaly	Hepatic involvement	Splenectomy	Storiform variant	Died after splenectomy
Lieu <sup>10</sup>	1993	1	M/71	1870	Abdominal pain, weight loss, splenomegaly	Hepatic involvement	Splenectomy	Storiform variant	Died after splenectomy
Bonilla <sup>11</sup>	1994	1	F/42	No data	Itching, asthenia, fever, abdominal pain, weight loss	No	Splenectomy Chemotherapy Radiotherapy	No data	8 (Died with multiple peritoneal metastasis)
					Diarrhea, nausea, vomiting, anorexia, night sweats, lethargy, weight loss	No	Splenectomy	Storiform variant	19 (Died with multiple metastasis)
Colovic <sup>13</sup>	2001	1	F/45	700	Abdominal pain, night sweats, fever, weight loss	Hepatic involvement	Splenectomy chemotherapy	Storiform variant	15 (Died with multiple hepatic metastasis)
Ozaras <sup>14</sup>	2003	1	F/51	No data	Fever, weight loss, abdominal pain, splenomegaly	No	Splenectomy	Storiform variant	No data
Katsura <sup>15</sup>	2006	1	M/82	No data	Abdominal discomfort, loss of appetite, weight loss, fever	No	Splenectomy	Inflammatory variant	18 (Alive without metastasis)
Mantas <sup>16</sup>	2010	1	F/66	1300	Abdominal discomfort, splenomegaly	No	Splenectomy	No data	46 (Alive without metastasis)
He <sup>17</sup>	2011	1	M/35	984	Abdominal pain, anemia	No	Splenectomy	Storiform variant	7 (Died with generalized metastasis)
Current case		1	F/48	740	Abdominal pain, belching, weight loss	No	Laparoscopic splenectomy	Storiform variant	13 (Alive without metastasis)

features. Splenectomy including laparoscopic splenectomy with histological negative margins is the treatment of choice. Chemotherapy

and radiotherapy are other therapeutic modalities for patients with metastasis, but the therapeutic effect is poor.

### Conflict of Interests

Authors have no conflict of interests.

### Authors' Contributions

JFF carried out the design and coordinated this study, HL and JL collected and analyzed the clinicopathologic data. All writers have contributed in writing the manuscript. All authors have read and approved the content of the manuscript.

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