Oncology and Translational Medicine
DOI 10.1007/s10330-015-0075-y

December 2015, Vol. 1, No. 6, P261–P263

CASE REPORT

A case of primary malignant fibrous histiocytoma of the pancreas with liver metastasis*

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Abstract

Malignant fibrous histiocytoma (MFH) is one of most common types of soft-tissue sarcoma. However, it accounts for less than 1% of all human cancer types. In addition, primary MFH of the pancreas is very rare. Herein, we report of a 37-year-old man who presented with a pancreatic mass and did not show any specific symptoms. Pre-contrast computed tomography (CT) showed a heterogeneous, liquid-filled necrotic mass in the uncinate lobe of the pancreas. Contrast-enhanced CT showed a mild enhancement of solid components and the pancreaticoduodenal artery across the mass. The patient underwent a pancreaticoduodenectomy, ethanol ablation of the liver lesions, and chemotherapy with 4 cycles of ifosfamide and doxorubicin. The tumor recurred in the liver, but not in the pancreas, after 8 months.

Received: 2 March 2015
Revised: 24 March 2015
Accepted: 25 April 2015

Key words: malignant fibrous histiocytoma (MFH); pancreatic neoplasm; metastatic liver cancer

Malignant fibrous histiocytoma (MFH) is a mesenchymal tumor mainly composed of fibroblasts and histiocytes [1]. MFH is extremely uncommon, and occurs most often in the extremities, trunk, and retroperitoneum [2], with involvement of the pancreas being extremely rare. Only 20 cases of pancreatic MFH have been reported in the literature [3]. Owing to the lack of data, there are no guidelines for the treatment of MFH of the pancreas. It was reported that, of 16 patients with primary pancreatic MFH, approximately 50% of patients died within 1 year. Herein, we report of a patient with primary MFH of the pancreas and liver metastasis who is still alive 13 months after diagnosis, even though the lesions in the liver increased in size after therapy.

Case report

A 37-year-old man was referred to our hospital because of abnormal findings in the pancreas on computed tomography (CT) during a routine medical check-up. He was asymptomatic apart from a 10 kg decrease in body weight over the past year. The patient had no history of trauma or pancreatitis. Physical examination upon admission was unremarkable. No jaundice was found, and no lymph nodes were palpable. Serum levels of alanine aminotransferase and aspartate aminotransferase were 191 U/L (normal range: 0–40 U/L) and 216 U/L (normal range: 0–40 U/L), respectively, while the white blood cell count was 10.40×10^9/L (normal range: $4–10 \times 10^9$/L) and the neutrophil number was 89.9%. Serology results for hepatitis B and C were negative. Serum levels of carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA19-9), carbohydrate antigen 125 (CA125), and alpha-fetoprotein (AFP) were negative. Pre-contrast abdominal CT showed a heterogeneous, liquid-filled necrotic mass in the uncinate lobe of the pancreas. Contrast-enhanced CT showed mild enhancement of solid components and of the pancreaticoduodenal artery across the tumor mass. There were two hypodense lesions in the right lobe of the liver on the pre-contrast CT scan, and the lesions showed a mild enhancement on the contrast CT scan (Fig. 1). Imaging examinations did not show any abnormal findings in the lungs, spleen, or kidneys. Surgical exploration revealed a tumor in the uncinate lobe of the pancreas, which adhered to the horizontal part of the duodenum. The patient underwent pancreaticoduodenectomy and ethanol ablation of the liver lesions, and received 4 cycles of ifosfamide and doxorubicin chemotherapy. Macroscop-

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* Supported by a grant from the Research Fund for Young professors of Zhengzhou University (No. 1421412090).

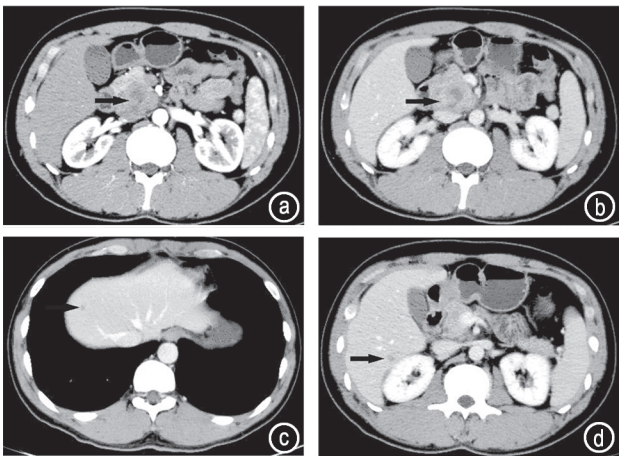

Fig. 1 CT findings before therapy. (a–b) Abdominal CT showed a heterogeneous, liquefacient necrotic mass in the uncinate lobe of the pancreas on contrast CT. The lesion showed a mild enhancement of solid components and pancreaticoduodenal artery across the mass on the contrast CT; (c–d) There were two low density lesions in the right lobe of the liver and the lesion showed a mild enhancement on the contrast CT

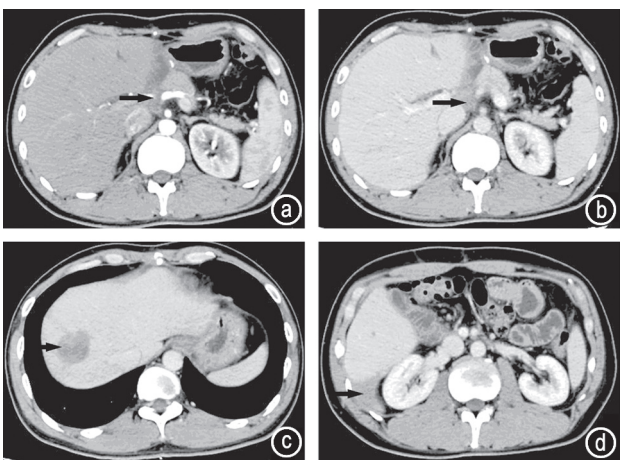

Fig. 2 CT findings after therapy for 8 months. (a–b) Abdominal CT showed no recurrence in the uncinate lobe of the pancreas after resection on contrast CT; (c–d) CT showed two low-density masses in the right lobe of the liver

ically, the tumor specimen measured 4 cm × 3 cm × 3 cm (Fig. 2). The cut surface of the hard tumor was grayish-white. Microscopically, the tumor cells were negative for CA19-9, Melan A, CK8, CEA, CD21, CD3, CD20, ALK, and SM. Tumor cells stained positive for Syn, CgA, CD56, NSE, S-100, CD68, and CD163. The Ki-67 labeling index was 50% (Fig. 3).

Discussion

MFH, also known as undifferentiated pleomorphic sarcoma, is the most frequent type of soft-tissue sarcoma and accounts for less than 1% of all human cancer types.

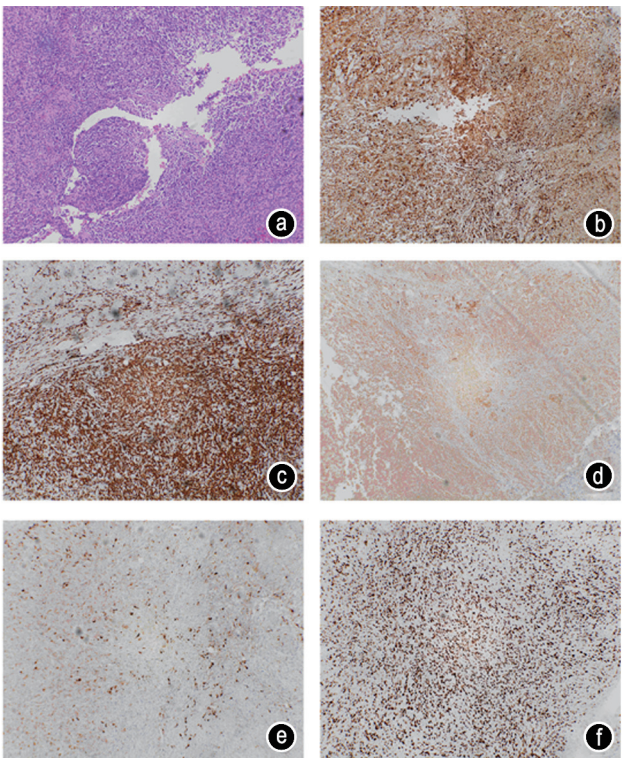

Fig. 3 (a) HE (× 100); (b–f) Tumor cells exhibiting immunoreactivity of CD68 (× 100), CD163 (× 100), CD56 (× 100), S-100 (× 100), and Ki-67 (50%), respectively

Men aged 50–70 years are more vulnerable than women of the same age. MFH often occurs at the extremities, as well as in the trunk and the retroperitoneum, [2] although it could occur almost anywhere in the body because of its mesenchymal origin [4]. Primary MFH of the pancreas is an extremely rare type of sarcoma. To date, only 20 cases of primary MFH of the pancreas have been reported in the literature [3]. MFH typically exhibits a broad diversity in its histopathological appearance, and it has been classified into five subtypes [5]: undifferentiated high-grade pleomorphic sarcoma, myxofibrosarcoma, undifferentiated pleomorphic sarcoma with giant cells, undifferentiated pleomorphic sarcoma with prominent inflammation, and angiomatoid fibrous histiocytoma. Primary MFH is known to have a high potential for metastasis or recurrence, even if the resection margin is tumor-free [6]. The lungs are the most common site of metastasis that mainly spread through the circulatory and lymphatic systems, with the liver being the second most common metastasis site. In our case, the patient displayed liver metastasis. MFH is difficult to diagnose because of the lack of specific symptoms and imaging findings, [7] and the lesions in the pancreas are quite similar to pancreatic cancer. The most common presenting symptoms of pancreatic MFH [3] are abdominal pain or discomfort, jaundice, vomiting, epigastralgia, reflux esophagitis, and anorexia. Only

a few reports discussed the radiological findings of MFH of the pancreas. The CT findings that best help in the diagnosis of MFH are as follows: a large, heterogeneous, hypodense, or multi-nodular mass on pre-contrast CT, which could become a non-homogeneously enhanced mass with numerous areas of necrosis. MRI, which is more sensitive and specific for primary MFH than CT, shows that primary MFH has high-signal intensity on T2-weighted images and non-homogeneous iso-signal intensity compared to its surroundings [7]. To date, the best approach to the management of pancreatic MFH is still controversial. As previously reported, MFH is composed histologically of fibroblasts and histiocytes. Surgical excision using the Whipple operation, distal resection or, if the tumor is multi-centric, total pancreatectomy, remain the primary treatment for pancreatic MFH [3]. Both adjuvant radiotherapy and chemotherapy, either alone or in combination, may be additional approaches for those patients with an insufficient surgical margin, as a means to prolong survival time [8]. As previously reported by Gutierrez [9] who evaluated 8249 cases of soft-tissue sarcoma, radiation therapy significantly increased the median overall survival of MFH patients by 3 months, and patients who received chemotherapy had significantly less overall benefits as compared with those who did not receive chemotherapy. Singer [10] also found that no significant improvement in survival but a decrease in quality of life when patients were treated with mesna, adriamycin, ifosfamide, and dacarbazine) and mesna, adriamycin, and ifosfamide chemotherapy for soft-tissue sarcomas, although more data is required to reach a conclusion. Patient prognosis mainly depends on the histological grade, size and location of the tumor, and the efficacy of resection, but typically, the prognosis is poorer for larger and deeper lesions [1].

Conclusion

We present an exceedingly rare case of primary MFH of the pancreas and liver metastasis with poor prognosis; this case had no specific symptoms and was difficult to diagnose before surgery. CT and MRI may be important diagnostic tools to determine whether a mass in the pancreas is a MFH or pancreatic cancer. Although MFH is very rare, as a clinical doctor, the possibility of primary MFH of the pancreas should be acknowledged when making a differential diagnosis for a mass in the pancreas, especially when the serum levels of CA19-9 are negative.

Conflicts of interest

The authors declare no potential conflicts of interest.

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DOI 10.1007/s10330-015-0075-y
Cite this article as: Zhai WL, Ye JW, Liang ZW, *et al*. A case of primary malignant fibrous histiocytoma of the pancreas with liver metastasis. Oncol Transl Med, 2015, 1: 261–263.