Case Report

Inflammatory Myofibroblastic Tumours of the Spleen and Liver

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Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm. Generally, these lesions have a benign behaviour, but the possibility of malignant transformation exists. We report the rare case of a 43-year-old woman with metachronous IMTs in the spleen and the liver. The patient was treated with laparoscopic splenectomy and partial hepatectomy. The patient recovered uneventfully. This case emphasizes the difficulties in diagnosis and the possibility of a metachronous occurrence. [Asian J Surg 2008;31(1):25–8]

Key Words: hepatectomy, inflammatory myofibroblastic tumour, liver neoplasm, splenectomy

Introduction

Inflammatory myofibroblastic tumour (IMT) is a rare condition. There are a variety of synonyms for this tumour including plasma cell granuloma, postinflammatory tumour, xanthomatous pseudotumour, inflammatory pseudotumour and inflammatory fibrosarcoma.1 By definition, the tumour is composed of dominant spindle cell proliferation with a variable inflammatory component. These spindle cells are now known to be myofibroblasts and this is the reason for the current designation for this disease. IMT may affect individuals of any age, but it has a predilection for children and young adults. It is most commonly found in the lung, but is also reported in the central nervous system, salivary glands, larynx, breasts, pancreas, spleen, lymph nodes, skin and liver. As the synonyms suggest, there is some confusion as to whether IMT is truly neoplastic or whether it represents a reactive proliferation of myofibroblastic cells. It is now generally accepted that IMT is a true neoplasm. These lesions have a benign behaviour, but the possibility of malignant transformation exists.2–4 We report the case of a 43-year-old woman with metachronous IMTs in the spleen and the liver.

Case report

A 43-year-old woman had a past history of laparoscopic splenectomy for splenic abscess in our hospital 5 years ago. Otherwise, she had a good past medical health. She was referred again to our hospital for an incidental finding of two liver tumours during routine health evaluation by abdominal ultrasonography. Physical examination did not show any stigmata of chronic liver disease or hepatomegaly. Bilirubin, liver transaminases and alkaline phosphatase levels were all normal. Hepatitis serology of HBsAg and anti-HCV, α-fetoprotein, and carcinoembryonic antigen were negative.

Computed tomography scan of the abdomen showed two contrast-enhanced masses measuring 6.3 × 5.5 × 5.3 cm at liver segments 6 and 7 and 2.8 × 1.8 × 1.7 cm at segment 8 (Figure 1). Hepatocellular carcinoma with a satellite nodule was suspected. However, magnetic resonance imaging showed radiologic features of benign liver lesions (Figure 2). Ultrasound-guided liver tumour biopsy showed features of IMT (Figure 3). For this, the histology of the previously resected splenic lesion was reviewed again by experienced pathologists. The diagnosis was revised to

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Figure 1. Contrast-enhanced computed tomography scan of the liver showed a lobulated mass at segments 6 and 7 and a satellite nodule at segment 8.

Figure 2. Magnetic resonance imaging of the two liver masses showed low to iso-intensity on T1-weighted images, and heterogeneous high intensity on T2-weighted images. After administration of Gd-DTPA, the masses showed delayed enhancement.
IMT of the spleen from the previous diagnosis of splenic abscess (Figure 4). The patient underwent resection of hepatic segments 6, 7 and 8 in June 2006 (Figure 5).

Microscopically, the tumours were characterized by a proliferation of spindle cells with intermingled inflammatory cells (lymphocytes, plasma cells, eosinophils). Numerous special stains were performed. The spindle cells were immunopositive for vimentin, smooth muscle actin and CD68. Negative staining was noted for EMA, ALK, desmin, CK, CD12, CD117, CD21, CD34, S100 and FVIII. There was no evidence of dysplasia or malignancy. Cellular mitosis was seen. The pathological diagnosis of IMT was made. The patient recovered well from surgery. She was well and without evidence of disease 6 months after the partial hepatectomy.

Discussion

Historically, the medical literature on IMT has been confined to small case series involving a single organ, with few reports encompassing multiple anatomic sites. Although the lung is the best known and the most common site, IMT occurs in diverse extrapulmonary locations. We reported a rare case of metachronous IMT of the spleen and the liver.

On gross examination, IMT tends to be circumscribed or multinodular, firm, white or tan, with a whorled fleshy, fibrotic or myxoid cut surface. The surface may show a variegated appearance with areas of necrosis and haemorrhage in a minority of cases. Microscopically, three basic histological patterns are recognized: (1) myxoid, vascular, and inflammatory areas resembling nodular fasciitis; (2) compact spindle cells with intermingled inflammatory cells (lymphocytes, plasma cells, eosinophils) resembling fibrous histiocyctoma; and (3) dense plate-like collagen resembling a desmoid or scar. Generally, there is little pleomorphism or mitotic activity. Immunohistochemistry is a valuable adjunct to light microscopic diagnosis. Vimentin is almost invariably positive in the spindle cells. Smooth muscle actin and muscle specific actin are present in the
The hypothesis of immunological origin of this disease. Desmin, CD68, pankeratin and p53 are positive in some cases. S100, CD21 and myoglobin are uniformly negative.

The aetiology and pathogenesis of IMT remain unknown, although the tumours were initially thought to represent a reactive inflammatory process. Aetiological causes, including infections, vascular causes and autoimmune disorders, have been hypothesized in their pathogenesis. Some authors have hypothesized that microorganisms from certain conditions, such as appendicitis, seed the hepatic parenchyma through the portal vein, creating an inflammatory reaction with obliterated phlebitis and granuloma formation. Some cases were reported to be Epstein-Barr virus (EBV)-positive inflammatory follicular dendritic cell tumours. However, this theory is doubtful because no evidence of infectious organisms could be demonstrated in most of the other reported series. In our patient, no acid-fast organisms, fungi, parasites, or bacteria could be identified and EBV was not detected on in situ hybridization. Vascular cause is another hypothesis and it is believed by some authors that these lesions may be due to intraparenchymatous haemorrhage secondary to trauma or coagulopathy. The high content of plasma cells in this lesion suggests the hypothesis of immunological origin of this disease.

The diagnosis of IMT is often difficult, despite the use of modern imaging and laboratory techniques, and generally these masses are often confused with other lesions, such as primary or secondary neoplasm, because the clinical presentations and morphological appearances are often nonspecific. For the diagnosis of IMT of the spleen, fine needle aspiration biopsy may cause haemorrhage and the results may be misleading due to the presence of inflammatory and mesenchymal cells. Therefore, most authors advocate surgery for the diagnosis and treatment of IMT of the spleen. Nowadays, laparoscopic splenectomy can be used for definitive diagnosis and symptomatic relief with minimal morbidity and mortality. In contrast, the treatment options for IMT of the liver are varied. Surgery, high-dose steroids, irradiation, and chemotherapy have been reported. Spontaneous regression has also been reported in some cases of IMT. Although some authors have suggested only simple observation or conservative therapy with steroids or nonsteroidal anti-inflammatory drugs if an accurate diagnosis of IMT is made, surgical resection is the preferred treatment for IMT of the liver, especially in patients for whom a definite histological diagnosis cannot be made preoperatively or by intraoperative frozen sections. The natural course of IMT without resection is unclear. A small proportion of patients with local recurrence or metastases after partial hepatectomy have been recorded. In fact, most patients reported in the medical literature recovered uneventfully after partial hepatectomy. Since the morbidity and mortality of partial hepatectomy in noncirrhotic livers is low, surgical resection with clear margins should be considered as the treatment of choice for IMT.

References