[症例報告]Malignant fibrous histiocytoma of the mesentery : A case report

Malignant fibrous histiocytoma of the mesentery : A case report

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ABSTRACT

A 46-year-old female was referred to the Ryukyu University Hospital for further evaluation of a large abdominal tumor. Physical examination showed a firm, fixed mass in the left side of the abdomen extending to 20 cm below the left costal margin. Diagnostic modalities using BE, CT, MRI, and angiography indicated that the tumor was malignant and arose from the mesentery. Exploratory laparotomy revealed a well-circumscribed tumor spreading to the distal transverse colon and the proximal jejunum in the mesentery. The tumor with the involved colon and jejunum was radically resected. Macroscopically, the tumor was ovoid and 21 cm in greatest diameter, and weighed 2,615 g. On cut section, the solid tumor was whitish and had an area of yellow, soft necrosis. Histologically, the tumor was diagnosed to be a malignant fibrous histiocytoma, and it was confirmed by immunohistochemical and electron microscopic studies. The patient was treated with adjuvant chemotherapy postoperatively and discharged with no evidence of recurrence. *Ryukyu Med. J., 16(2)75~78, 1996*

Key words: malignant fibrous histiocytoma, storiform-pleomorphic type, mesentery, prognosis

INTRODUCTION

Soft tissue sarcomas comprise 1% of all malignant neoplasms and malignant fibrous histiocytoma (MFH) accounts for around 20% of soft tissue sarcomas[1~5]. MFH originating in the retroperitoneum and mesentery is reported to occur in 5.7% to 16% of patients with this neoplasm[1,3,6]. It is a rare tumor and is seldom encountered in the career of the average gastroenterological surgeon. We recently encountered a patient with a large MFH of the mesentery, which was successfully removed and on which we presently report. The rarity has prompted us to present this case with a brief review of the literature regarding the major prognostic factors.

CASE REPORT

A 46-year-old female was referred to the Ryukyu University Hospital on May 22, 1995 for further evaluation for a large abdominal tumor with abdominal fullness and discomfort for a few months. She also developed episodes of occult hematuria and low-grade fever two months prior to this admission. Her past history revealed no remarkable illness and previous surgery.

Physical examination revealed a firm, fixed mass in the left upper abdomen extending to 20 cm below the left costal margin in the mid-clavicular line. Her laboratory data showed 8.6 g/dl of hemoglobin, 29.9% of hematocrit, and 2.91 mg/dl of C-reactive protein. Urinalysis showed occult hematuria. Other blood parameters including CEA, CA19-9, and LDH were within normal limits, as was the chest X ray. Flat and upright abdominal X ray showed a shadow of mass occupying the left abdominal cavity. A barium enema study revealed the compression and medial displacement of the left colon by the huge mass (Fig.1, A). Contrast-enhanced CT scan of the abdomen showed a well-circumscribed 18×16 cm solid tumor with a hypodense lesion, arising possibly from the mesentery or retroperitoneum (Fig.2, A). Coronal T2-weighted MRI revealed the tumor to be isointense with the liver, with a hyperintense lesion in the lower portion of the tumor (Fig.2, B). Angiogram of the superior mesenteric artery demonstrated that the middle colic artery and its branches were stretched and displaced around a large, hyper-vascular mass in the mesentery. The dilated middle colic branches supplied tumor vessels (Fig.1, B).

On June 6, the operation for the tumor was performed. The tumor was located in the left upper abdomen extending to the base of the mesentery with marked adhesion or invasion to the transverse colon and the

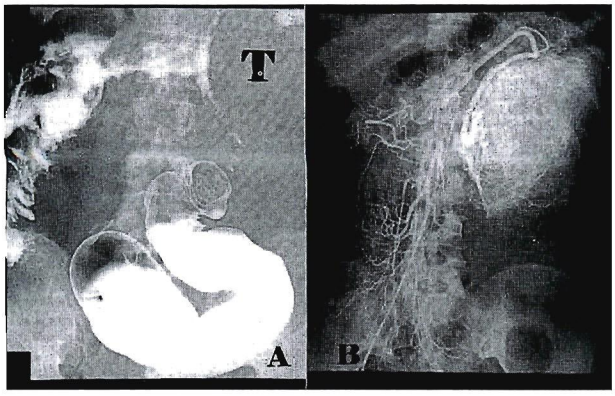

Fig.1 Barium enema study showing medial displacement of the left colon with irregular strictures by a tumor mass (T) (A). Angiogram of the superior mesenteric artery demonstrated stretching and displacement of the middle colic artery and its branches with tumor vessels (B).

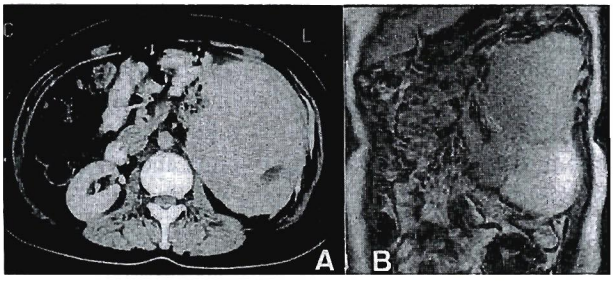

Fig.2 Contrast-enhanced CT scan of the abdomen showing a well circumscribed tumor mass with a hypodense zone (A). Coronal T2 weighted MRI revealing the tumor, isointense with liver, with a hyperintense lesion in the lower portion of the tumor (B).

proximal jejunum around the base of the mesentery. It was removed from the retroperitoneum with a clear free margin, but the transverse colon and the proximal jejunum were intensively adherent to the tumor. At the manual palpation, there were negative findings on the other abdominal viscera, omentum, and paraaortic lymphnodes. Extirpation of the tumor with the mesocolon, and partial resection of the distal transverse colon and the 20 cm proximal jejunum were then performed. The residual tumor was not detected in the abdomen.

The tumor was 21×16×13 cm in size and 2,615 g in weight. The cut section showed a whitish shining solid tumor with a yellow myxoid area (Fig.3). Microscopically, the solid part of the tumor was mainly composed of spindle-shaped cells with atypical ovoid nuclei, and arranged in a classical storiform or cartwheel pattern (Fig.4, a). The myxoid area of the tumor showed mononuclear or multinucleated giant cells in a loose band-like arrangement (Fig.4, b). These findings met the diagnostic criteria for MFH as proposed by Weiss and Enzinger[3]. The tumor cells infiltrated into the colon with osseous metaplasia and also infiltrated into the jejunum (Fig.5).

Immunohistochemical staining was positive for OKM1, α_1-antichymotrypsin, and S-100, but negative for myog-

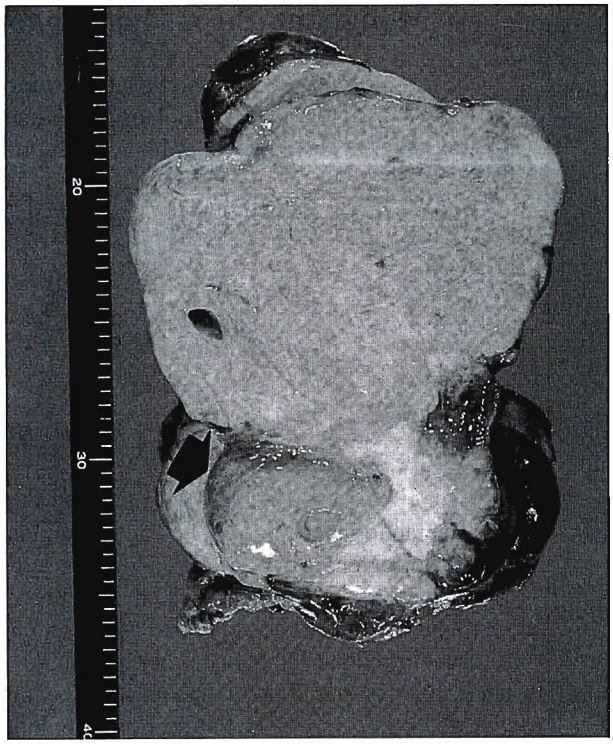

Fig.3 The cut section showing a whitish, solid tumor with an area of yellow,soft necrosis (arrow).

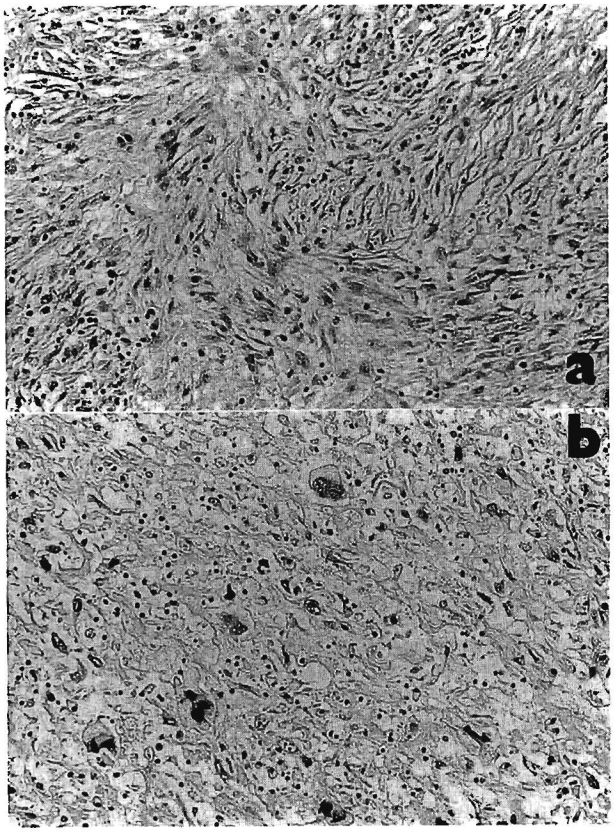

Fig.4 The solid part of the tumor composed of spindle-shaped cells with bizarre nuclei in a storiform arrangement (a: HE, ×50). The soft necrotic part showing mononucleated or binucleated giant cells with atypical nuclei in a diffuse, loose arrangement (b: HE, ×50).

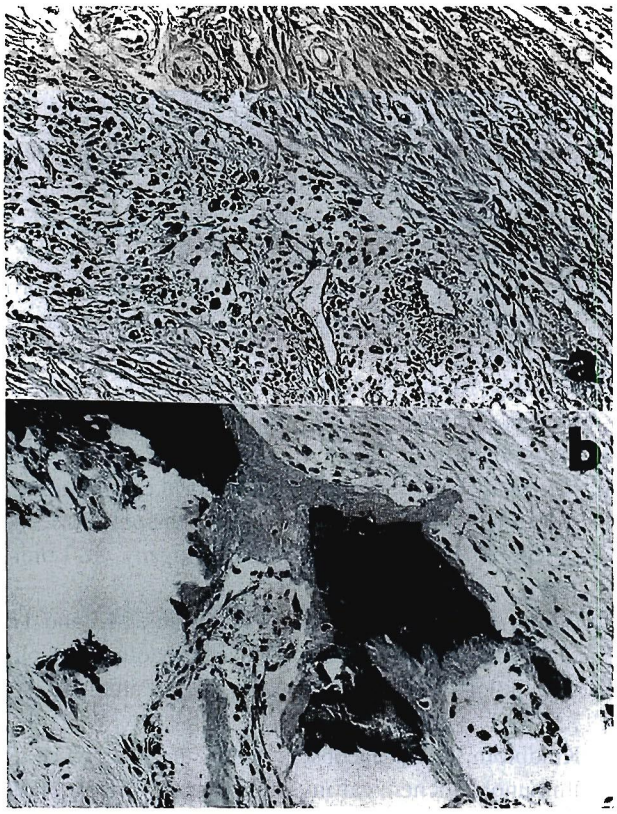

Fig.5 The involved colon showing infiltration of the tumor into the muscle fascicles (a) and osseous metaplasia in the tumor (b) (Both: HE, ×50).

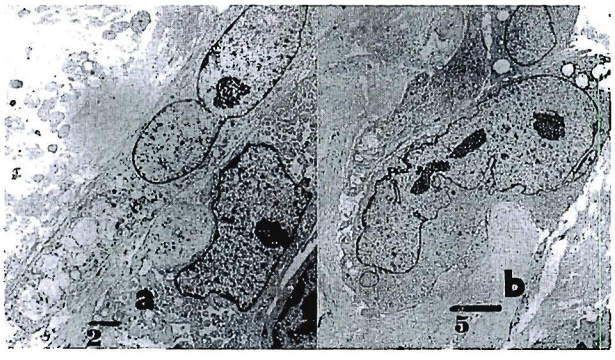

Fig.6 Electron microscopic examination revealing two main tumor cells; fibroblastic cells (a) (×2,000) and histiocytic cells (b) (×1,500).

lobin, desmin, and vimentin. Positive immunohistochemical demonstration of cell markers such as α_1-antichymotrypsin, OKM1 which are considered specific for histiocytes indicated histiocytic origin of the tumor[7,8].

Electron microscopic study revealed two types of main tumor cells, fibroblast-like cells and histiocyte-like cells in any portion of the tumor. The most predominant cell type was fibroblast-like cell, in mostly elongated or polygonal shape with smooth nuclei. The cytoplasm contained multiple cisternae of rough endoplasmic reticulum, well-developed Golgi zones and numerous mitochondria (Fig.6, a). The histiocyte-like cells varied in shape and were characterized by ruffled cell margins and reniform nuclei. The abundant cystoplasm contained variable numbers of lysosomes, abundant ribosomes, and moderate amounts of smooth endoplasmic reticulum vesicles (Fig.6, b).

The postoperative course was uneventful, and the pateint was treated with adjuvant chemotherapy (EPI, CDDP), and was discharged on the second postoperative month. At the time of discharge, abdominal CT scan showed normal findings.

DISCUSSION

Soft tissue sarcomas including MFH are usually grouped together because of perceived similarities in pathologic appearance, clinical presentation, and behavior[1]. Because these tumors are relatively rare, and this MFH has been recognized fairly recently, there have been relatively few major studies that examine any type of soft tissue sarcoma specifically and selectively[2~4].

MFH occur mostly in patients in the 5th to 7th decades of life, and occur more commonly in males than in females in an approximately 1.5 : 1 ratio[1~4]. Two thirds to three fourths of the MFH occur in the extremities, most commonly in the thigh, accounting for one third of all cases[2~4]. Other frequent locations include the trunk, shoulder region, and retroperitoneum[5]. Retroperitoneal or mesenteric MFH such as that of our case is reported to occur in 5.7% to 16% of all the MFHs[1,3,6].

In general, MFH has been considered as a highly aggressive tumor with a propensity of local growth, local recurrence and distant metastasis. The major prognostic factors for MFH are reported to be tumor size and histologic grade of the primary tumor. Bertoni et al. reported that the prognosis was worse in tumors larger than 5 cm in diameter, tumors that recurred early (less than 1 year) after surgery, and tumors that had inadequate surgical treatment[2]. The overall 5-year survival rate was only 36%[2]. Kearney et al., in a report of 167 cases of MFH, found the most important prognostic factors to be the depth of the tumor[4]. In a review of 200 cases of MFH, Weiss and Enzinger reported that larger and more deeply located tumors metastasized most frequently[3]. Pezzi et al. reported 227 cases of MFH and found that survival rates decreased progressively with increasing size of the primary tumor. Five-year survival rates were 82% for tumors smaller than 5 cm, 68% for tumors between 5 and 10 cm, and 51% for tumors larger than 10 cm[1]. The largest mean tumor size was found in the lesions arising in the retroperitoneum (mean, 16.2 cm), followed by the extremities (mean, 8.5 cm)[1].

These reported data lead us to believe that primary tumor size significantly correlates with survival rates. In our case, the tumor size was 21 cm in greatest diameter with involvement of the adjacent intestine. Consequently, our patient would pursue a poor outcome although the tumor was radically resected without residual tumor.

With regard to the relation of histologic grade to survival rates, Pezzi et al. found that there was a significant difference in the survival curves[1]. Patients with intermediate-grade tumors showed a significantly improved survival rates (a 80% of 5-year survival rate) as compared with those with high-grade tumors (60%). However, the study by Kearney et al. revealed no difference in prognosis between the different histologic subtypes[4]. MFH is histologically characterized by tumors mainly composed of cells with histiocytic and fibroblastic characteristics mixed in various proportions and usually classified into five types : (1) the storiform-pleomorphic type ; (2) the myxoid type known as myxofibrosarcoma ; (3) the giant cell type, being malignant giant cell tumors of the soft tissue ; (4) the inflammatory type, such as xanthosarcoma and malignant xanthogranuloma ; and (5) the angiomatoid type. The present tumor was diagnosed to be type 1. In clinical practice, it is generally believed that patients with a higher-grade tumor have significantly less chance of cure than those with a lower-grade tumor. Unfortunately there is little agreement on the histologic grading system and features of the tumor which best represent the degree of differentiation. In a study of 227 cases of MFH, Pezzi et al. reported that the tumors were assigned to one of two grade groups (an intermediate or high-grade group) based predominantly on their degree of cellularity and a significant difference in the survival rates between the two grade groups was evident[1]. The tumor in our case was assigned to a high-grade group. As a result this patient has possessed at least two unfavorable prognostic factors for MFH.

This relationship between grade, size and prognosis for MFH has some therapeutic implications. Adjuvant chemotherapy remains controversial for soft tissue sarcomas, with some randomized clinical studies showing a clear advantage in overall or disease-free survival[9,10], whereas others show no benefit for adjuvant chemotherapy[11]. Adjuvant chemotherapy is most likely to be beneficial for subgroups of patients with the highest risk of distant metastasis and death resulting from disease. Accordingly, this patient was thought to be a more suitable candidate for adjuvant chemotherapy. Postoperatively, our patient was treated with adjuvant chemotherapy.

After discharge, regular outpatient follow-up including ultrasonography and CT scan are recommended due to a high potential of local recurrence and distant metastasis.

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