[症例報告]Retropentoneal malignant fibrous histiocytoma : Acasereport

Retroperitoneal malignant fibrous histiocytoma : A case report

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ABSTRACT

Malignant fibrous histiocytoma (MFH) is the most common among the malignant bone and soft tissue tumors. However, retroperitoneal cases of MFH are occasional. A 70-year-old man was hospitalized with loss of appetite and left hypochondralgia. Prognosis showed a tumor in the left upper abdomen by ultrasonography, CT and MRI. We could not accurately diagnose and detect the origin of the tumor preoperatively since definitive diagnosis depends on histopathological findings. Early discovery of retroperitoneal MFH is difficult and its prognosis is poor. Surgical excision is the first treatment choice, however, other additional effective treatments, such as chemotherapy, radiotherapy and immunotherapy are necessary. *Ryukyu Med. J., 16(3)127~130, 1996*

Key words: malignant fibrous histiocytoma, retroperitoneal case, surgical excision

INTRODUCTION

Malignant Fibrous Histiocytoma (MFH)[1,2] has the highest incidence among the malignant bone and soft tissue tumors. It is however, rare to encounter a retroperitoneal case of MFH in daily practice of medicine. This paper reports a huge tumor that was adjacent to the intraperitoneal and retroperitoneal organs, of which preoperative diagnosis was difficult.

CASE REPORT

A 70-year-old male patient visited Naha City Hospital with complaints of appetite loss and left hypochondralgia which had been persistent for two months since the middle of July 1995. Physical examination revealed a palpable tumor in the left upper abdomen. He underwent an operation for gastric ulcer approximately 40 years earlier. Family history revealed no special problems. On admission, he was 165 cm tall and weighed 45 kg. The palpebral conjunctiva was slightly anemic but the bulbar conjunctiva was not icteric. No superficial lymph nodes were palpable. There was tenderness in the left hypochondrium and an infant head sized tumor was palpated in the left upper abdomen. Laboratory examination showed no abnormalities except for mild anemia. Tumor markers (CEA·CA19-9) were within normal ranges.

Abdominal X-ray disclosed calcification at the site of the tumor. Ultrasonography showed that the tumor had a clear boundary and smooth surface. Gastric fiberscope examination revealed extrinsic oppression of the gastric wall. The mucosa was normal with Group 1 cytology. The capacity of the stomach was extremely small. The extrinsic oppression was caused by the tumor at the lesser curvature of the body portion which caused severe deformity of the stomach as shown by barium meal. Computed Tomography (CT) showed that the tumor (13×15 cm) was adjacent to the gastric posterior wall and the pancreas. Density of the tumor was irregular, and showed partial calcification and cystic patterns.

Using Magnetic Resonance Imaging (MRI) on T2-weight portrait, the major portion of the tumor showed cystic patterns. The portion showing high intensity in the MRI (T1) suggested bleeding (Fig.1). In the study of selective celiac arteriography, no malignant findings such as irregularity of vessels, vascularization or tumor stain were recognized, although feeding vessels diverging to the tumor from the left gastric artery (Fig.2) were noticed.

Laparotomy was carried out with provisional diagnosis of gastric submucosal, pancreatocystic or retroperitoneal tumor. There was slight adhesion between the tumor and greater omentum or peritoneum. No ascites was observed. The tumor had a thick capsule, smooth and soft surface at the site of the lesser curvature of the stomach and behind the transverse colon. We found it originating in the retroperitoneum, which compressed the pancreas forward with the lower margin reaching to both kidneys.

We were able to perform ablation of the tumor smoothly. However, the tumor had partially adhered to the spleen and the tail portion of the pancreas. Therefore, we

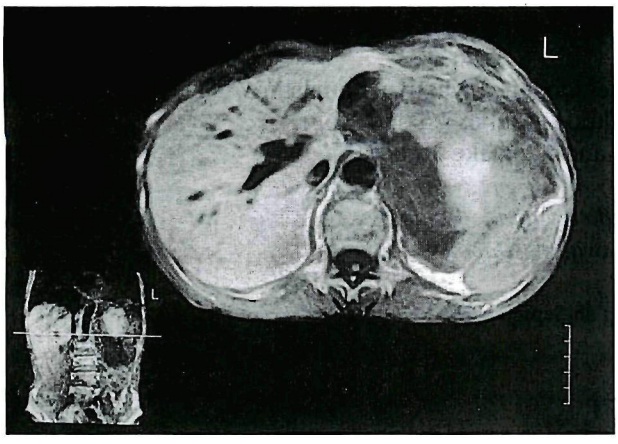

Fig.1 MRI (T1) demonstrating a huge mass within high intensity suggesting bleeding and cystic patterns in the upper left quadrant of the abdomen.

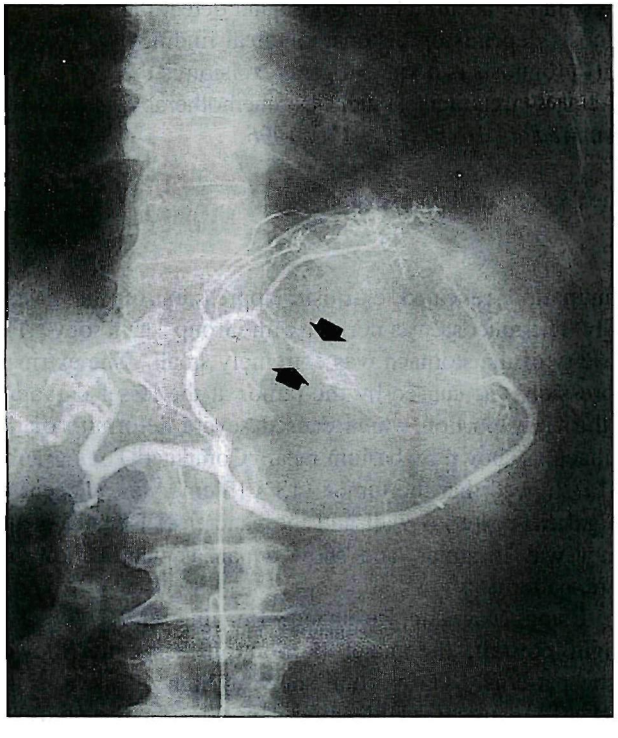

Fig.2 Celiac arteriography showing the feeding artery (arrow) diverging to a tumor from the left gastric artery.

performed resection of the tumor together with the spleen and the tail portion of the pancreas. The dominant vessel from the gastric artery that we had noticed preoperatively was not observed. No lymph node swelling around the tumor was noticed, and no visible metastases were recognized. The tumor was 21×20×15 cm in size and weighed 1,655 g. The inside of the tumor looked like a clear vegetable gelatin with some cartilage. Parts of the cystic lesion contained white semitransparent and mucoid substance (Fig.3).

Histopathologically the tumor consisted of spindle shaped cells with storiform pattern (Fig.4), atypical mononucleal and multinucleal cells, myxomatous parts and

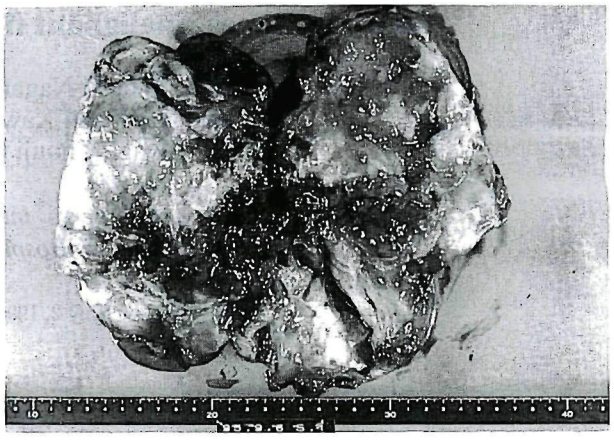

Fig.3 Macrophotographys of the resected specimen showing its cut surface.

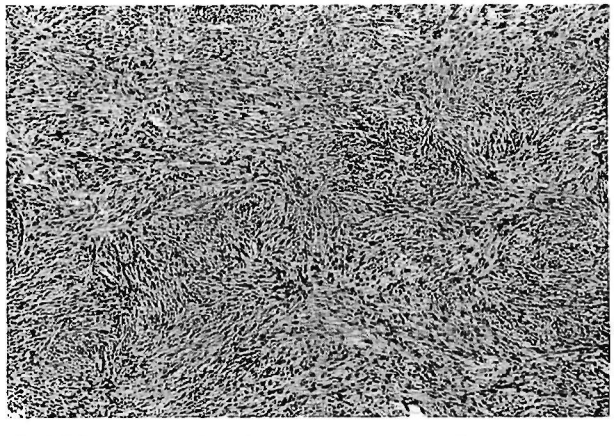

Fig.4 Microphotography of the tumor showing storiform pattern with a predominant of spindle-shaped cells (H&E staining. ×40).

bleeding. The staining of S-100 protein was negative. Histopathological diagnosis showed the tumor as MFH.

The patient was discharged twenty days after the operation with seemingly good progress. However, he was rehosptalized with reccurence of the tumor on the abdominal wall seventy-nine days after the operation, and passed away of sepsis due to peritonitis caused by intraperitoneal reccurence.

DISCUSSION

O'Brien & Staut[3] state that MFH has the highest frequency among the malignant bone and soft tissue tumors. Males are affected more than females. The patients in the ages of 50s to 70s account for approximately two-thirds of the total. MFH occurs mostly in the limbs, followed by the peritoneum and abdomen[1]. Among the malignant retroperitoneal tumors, MFH has the highest frequency of approximately 20% followed by leiomyosarcoma and liposarcoma[1]. Main symptoms of MFH occuring in the limbs include a painless swelling and a continuous enlargement[5]. In the retroperitoneal cases, subjective symptoms accompanied with the tumor growth are hard to

detect but abdominal distension and weariness are often observed in the patients[6]. Therefore, early discovery of the tumor is difficult until it becomes palpable[7]. Prognosis of MFH is poor. The retroperitoneal cases reflect 27.4% of survival rate over a five-year-period, while all cases of MFH reflect 47.7%[1].

In typical cases, the tumor surface is smooth, multinodular and capsulated[1]. The sectional property of the tumor is solid, and rarely shows mucoid pattern. When the tumor becomes large, cystic, necrotic and bloody changes occur[5]. Regardless of those changes, there are no specific features for MFH that are detectable by ultrasonography, CT, MRI and angiography[8]. Therefore, we must rely on histopathological findings discriminating it from other diseases[9]. If the tumor becomes huge, like in this case, it is almost impossible to detect a primary lesion preoperatively by CT or MRI. In this case, angiography suggested the feeding artery diverged from the left gastric artery. However we could not determine the primary lesion because it was in the retroperitoneum instead of in the stomach.

Histogenesis of MFH is unclear. However, there are several theories such as those of histiocytic origin[10] and undifferentiated mesenchymal cell origin[11]. Histological examination of MFH usually discloses mixture of fibroblast-like spindle cells and histiocyte-like round cells. Given various kinds of ratios of those cells, it is an undifferentiated polymorphic sarcoma. From the histlogical findings, it is subdivided into storiform-pleomorphic, myxoid, giant cell, inflammatory and angiomatoid types[5]. The present case was a storiform pleomorphic type, because of the recognizable spindle and fibroblast cells showing a storiform pattern.

Immunohistological stains with vimentin[12] as a mesenchymal tumor maker and alpha 1-antitripsin as a histiocytic tumor marker often show positive results. Those findings are not peculiar to MFH, but are helpful in diagnosis.

Surgical excision is the first choice of treatment for MFH and its local recurrence. Because of its frequent metastases, it is important to perform wide resection of the tumor including the connective tissue, the fascia and the muscles around the tumor and regional lymph nodes.

Hashimoto[1] reports a survival rate over a period of five years of 87% in the cases treated with wide excision in comparison with that of 50% in those with simple extraction. When the tumor becomes large, without symptoms in the retroperitoneal cases, curative resection is much more difficult. Dalton[4] states that the survival rate over a period of five years in those patients with tumor sizes of more than 5 cm is 56% and 36% in those with infiltrating tumors. There is no relationship between the degrees of atypicality or extent of karyomitosis of tumor tissue and the degrees of biological malignancy of the tumor in MFH. It is assumed that the cases with large tumors and of retroperitoneal origin have poor prognosis with frequent hematogenic metastases[6]. Remote metastases are often seen in lungs, lymph nodes, liver and the bone via hematogenic routes[1,2,5]. As an auxilliary treatment, radiotherapy is effective in some cases that are mainly composed of fibroblasts in spindle shapes. Chemotherapy with Cyclophosphamide, Vincristine, Adriamycin, Methotrexate and Cisplatin is reported to be effective[13~15]. However, there is controversy regarding the effectiveness of those therapies. Constructive surgical tumor excision involving other organs is indispensable for a better prognosis. In addition, it is necessary to establish an effective treatment such as chemotherapy[13~15], radiotherapy[16] and immunotherapy[17] beside wide excision of the tumor.

Weiss[5] reported many advanced cases of MFH, of which a local recurrent rate is 44% and metastatic rate is 42 %. According to Hashimoto[1], metastases had occured in 61.5% of the surgically treated patients within less than one year from their first surgery. There is, however, a report recognizing long term survival by performing repeated excisional operations for the reccurent tumors[18]. However, in this study, due to the poor general status of the patient after detecting intraperitoneal recurence, a reoperation could not be performed.

We believe that strict observation is important in detecting local recurrence, metastases, and that surgical excision with a possible procedure in reccurent cases within a few years after the initial operation is necessary.

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