Case Report

Surgical Resection of Primary Liposarcoma of the Anterior Mediastinum

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We report on the rare and surgical treatment of a case of primary mediastinal liposarcoma. A 64-year-old male complained of hoarseness for one month and was admitted to our hospital because of an abnormal shadow, which was postulated to be an anterior mediastinal tumor on a chest computed tomography (CT) scan. Horizontal T1-weighted magnetic resonance imaging (MRI) showed an anterior mediastinal round mass with a signal intensity similar to that of subcutaneous fat, which was 6.5 cm in diameter. An operation similar to extended thymectomy was perfomed through a median sternotomy. Histological examination of the resected specimen revealed that the tumor was composed of well-differentiated liposarcoma and pleomorphic malignant fibrous histiocytoma and the tumor was diagnosed as a de-differentiated liposarcoma. We discuss it with reference to a collective review of the Japanese literature for surgical cases of primary liposarcoma of the mediastinum. (Ann Thorac Cardiovasc Surg 2008; 14: 38–41)

Key words: mediastinal liposarcoma, extended thymectomy, de-differentiated liposarcoma

Introduction

Primary mediastinal liposarcoma is a very rare variant of mediastinal neoplasms, constituting less than 1% of all mediastinal malignancies and about 9% of primary sarcomas of the mediastinum.[1,2] We report a rare operative case of anterior mediastinal liposarcoma through a median sternotomy and discuss it with reference to a collective review of patients reported in the literature.

Case Report

A 64-year-old male, previously healthy, complained of hoarseness for one month and was admitted to our hospital because of an abnormal shadow, which was postulated to be an anterior mediastinal tumor on a chest computed tomography (CT) scan. Horizontal T1-weighted magnetic resonance imaging (MRI) showed an anterior mediastinal round mass with a signal intensity similar to that of subcutaneous fat, which was 6.5 cm in diameter (Fig. 1). An operation similar to extended thymectomy was perfomed through a median sternotomy. There was no invasion of the surrounding tissue, inspite of pleural adhesions on the cranial side of the tumor, enabling us to remove the tumor en bloc. The resected specimen showed a soft, rubbery, multilobular, and encapsulated yellow and gray mass (Fig. 2) and histological examination of this tumor revealed that it originated from the thymus and was composed of well-differentiated liposarcoma and pleomorphic malignant fibrous histiocytoma (Fig. 3). Therefore, the tumor was diagnosed as a de-differentiated liposarcoma. The patient had an uneventful postoperative course and was discharged on the 20th postoperative day. Six postoperative cycles of chemotherapy (doxorubicin hydrochloride, 10 mg/day for 3 days; ifosfamide, 3,500 mg/day for 5 days) were carried out. The patient remains free of disease 14 months after surgery.

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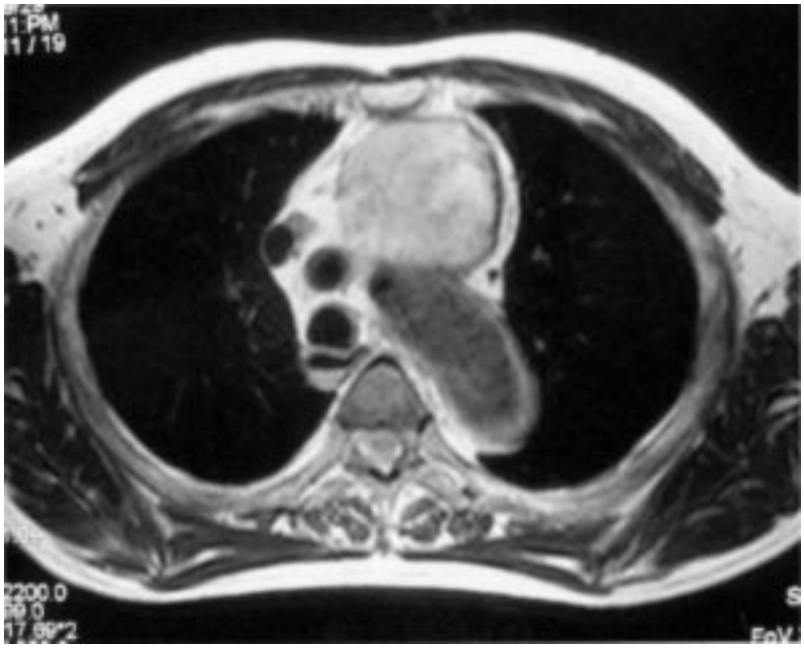

Fig. 1. Horizontal T1-weighted magnetic resonance imaging (MRI) showed an anterior mediastinal round mass with a signal intensity similar to that of subcutaneous fat, which was 6.5 cm in diameter.

Discussion

Primary liposarcoma of the mediastinumis a very rare malignancy that remain asymptomatic until it reaches a large size, and most are diagnosed by chest radiography. Common presenting symptoms include chest pain, dyspnea, wheezing, cough, and weight loss. CT scan and MRI are accurate in identifying fatty tumors, and characteristic findings of liposarcoma are inhomogenous soft tissue densities and intensities equal to that of fat. However, they do not originate from adipose tissue and usually belong to a primitive mesenchymal cell line, which has the property of lipogenesis. Pathologic subtypes of liposarcoma show multiple variation, i.e., well-differentiated, myxoid, pleomorphic, de-differentiated, mixed, and round cell types,[1,7] and myxoid liposarcoma accounts for 40 to 50% of the histological subtypes.

After reviewing the Japanese literature for surgical cases of primary liposarcoma of the mediastinum since 1996, only 15 were found, including our case (Table 1).[3–12] As shown in Table 1, the mean age was 56±18 years, with an age range of 12 to 79 years. The mean maximum size of the tumor was 14.7±6.9 cm, with a size range of 6 to 30 cm. These tumors can attain massive sizes and can arise in all mediastinal compartments, most commonly in the

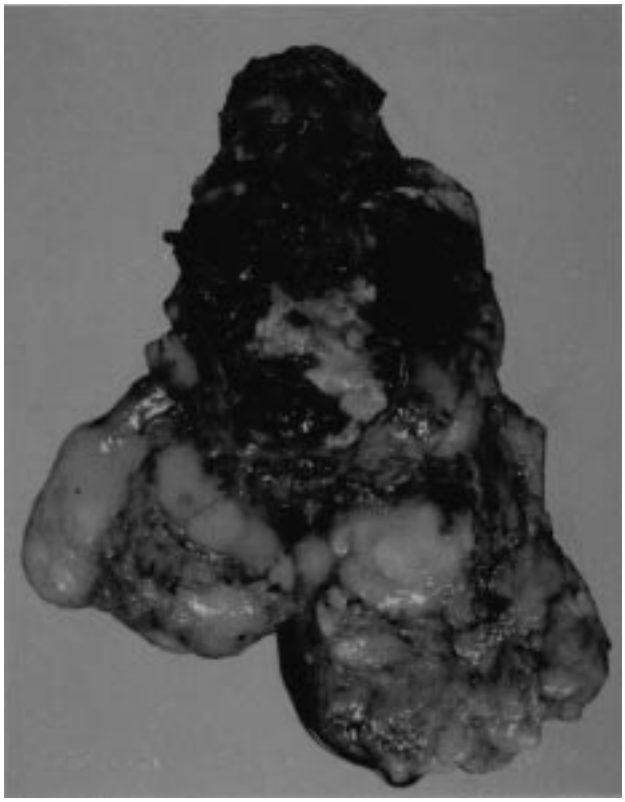

Fig. 2. Resected specimen originating from the thymus showed a soft, rubbery, multilobular, and encapsulated yellow and gray mass.

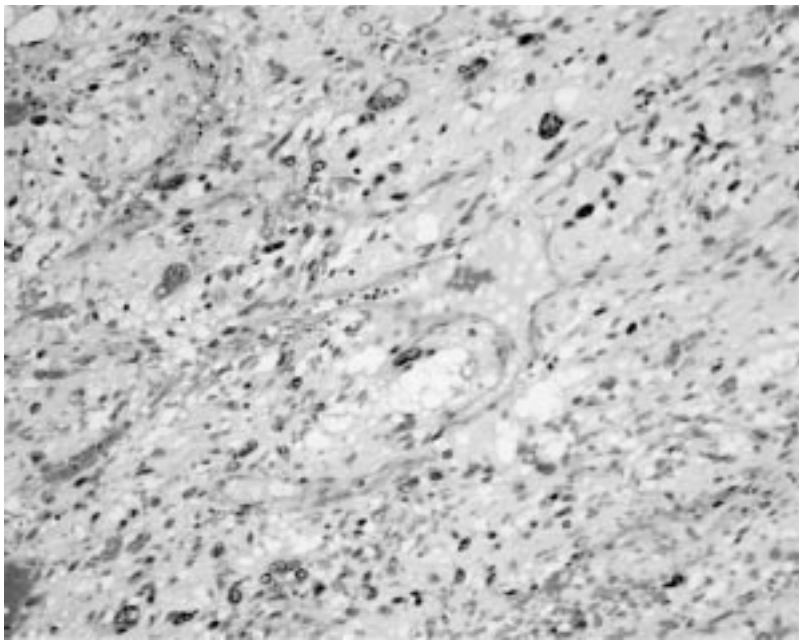

Fig. 3. Histological examination showed this tumor was composed of well-differentiated liposarcoma and pleomorphic malignant fibrous histiocytoma, and it was diagnosed as a de-differentiated liposarcoma.

Table 1. Review of the Japanese literature for surgical cases of primary liposarcoma of the mediastinum since 1996

Author (year)	Age/Sex	Size (cm) (Location)	Pathological type	Therapy	Outcome (months)
1) Kiyama et al. (1996)[3]	50/M	9×6 (anterior)	well-differentiated	S	36 alive
2) Tanaka et al. (1997)[4]	64/M	8×20×6 (posterior)	myxoid	S	3 alive
3) Tori et al. (1998)[5]	62/F	19×15×6 (anterior)	well-differentiated	S	28 alive
4) Koizumi et al. (1999)[6]	77/F	right 8×5, left 10×8 (bilateral posterior)	well-differentiated	S	3 alive
5) Shimada et al. (2000)[7]	79/M	11×8×7 (anterior)	mixed	S	13 dead
6) Sakamaki et al. (2001)[8]	76/M	12×12×9 (posterior)	well-differentiated	S	24 alive
7) Mase et al. (2002)[9]	48/M	18×15×5 (anterior)	well-differentiated	S	8 alive
8) Tian (2002)[10]	48/–	15 (posterior)	myxoid	S+R+C	65 dead
9) Tian (2002)[10]	40/–	10 (posterior)	myxoid	S+R	10 dead
10) Tian (2002)[10]	39/–	30 (posterior)	well differentiated	S+C	48 alive
11) Tian (2002)[10]	12/–	12 (anterior)	myxoid	S+C	5 dead
12) Tian (2002)[10]	75/–	25 (bilateral anterior)	myxoid	S	23 alive
13) Ohta et al. (2004)[11]	59/F	– (anterior)	well-differentiated	S (3 times)	192 alive
14) Inaba et al. (2004)[12]	47/M	9 (anterior)	well-differentiated	S (2 times)+R	51 alive
15) Hirai et al. (2008)	64/M	6 (anterior)	de-differentiated	S+C	14 alive

anterior or posterior. The survival rate seemed to depend on the pathological type. All 8 patients (100%) with the well-differentiated type were alive after a mean interval of 35 months, including 2 of 8 patients (25%) that had local recurrence. On the other hand, 2 of 5 patients (40%) with the myxoid type were alive after a mean interval of 13 months and 3 of 5 patients (60%) died after a mean interval of 26 months. McLean et al. reported that myxoid liposarcoma tends to disseminate to serosal surfaces of the pleura, pericardium, and diaphragm, either alone or in combination with the involvement of other viscera.[13] Kiyama et al. also reviewed the 21 cases in the Japanese literature and reported that the overall 5-year survival rate was 38.1%, 4 of 6 patients (67%) with well-differentiated liposarcoma were alive after a mean interval of 5.5

years, and none of the other types of patients were alive in the same interval.[3)]

In general, radiotherapy and chemotherapy are believed to be ineffective therapeutic modalities for survival. At present, therefore, the best treatment for mediastinal liposarcoma should be complete surgical resection. Our review also found that nine cases (60%) received surgical resection without adjuvant irradiation and/or chemotherapy. In particular, in the 8 patients with well-differentiated liposarcoma, 6 patients received only surgical resection, with only 2 (25%) receiving adjuvant chemotherapy or radiotherapy, and no local and systemic recurrence has occurred. However, we selected adjuvant chemotherapy postoperatively, although complete resection was also possible in our case, by informed consent due to the pathological finding of de-differentiated liposarcoma composed of pleomorphic malignant fibrous histiocytoma, a highly malignant tumor with a high local recurrence and metastasis rate. It was said that doxorubicin is the single most active agent with a response rate of 15 to 30%, and that improved response rates are evident when doxorubicin is combined with other agents.[14)] Furthermore, as tissue adhesion caused by radiation therapy might make it difficult to perform aggressive surgical intervention when local recurrence occurs postoperatively, we selected only adjuvant chemotherapy.

Primary liposarcoma is a relatively rare neoplasm, arising most commonly in the lower extremities and retroperitoneum, and primary mediastinal liposarcoma is very rare. As surgical treatment of mediastinal liposarcoma is very rare, we report and discuss it with reference to a collective review of the literature.

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