

A Case of Leiomyosarcoma of the Diaphragm

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A patient with a rare leiomyosarcoma of the diaphragm was encountered. A 74-year-old woman was admitted complaining of left shoulder pain and hypochondralgia. A mass was found on the left side of the diaphragm on imaging studies. Laparoscopic biopsy was performed and the diagnosis of leiomyosarcoma was established histopathologically. To improve her complaint, removal of the mass was attempted, which resulted in compromise resection. (Ann Thorac Cardiovasc Surg 2001; 7: 297–300)

Key words: diaphragm, leiomyosarcoma

Introduction

Leiomyosarcoma of the diaphragm is very rare. Due to the lack of symptoms, and unresponsiveness to various therapeutic modalities, the prognosis of leiomyosarcoma is very poor. Recently we encountered such a patient, and herein report the case.

Case Report

A 74-year-old woman consulted her physician with complaints of epigastric pain, oppressiveness in the chest and appetite loss for three months. Upper gastrointestinal series and abdominal ultrasonography were done and her condition was provisionally diagnosed as reflux esophagitis and cholelithiasis. She was then referred to our hospital for further evaluation and management. On admission, she had spontaneous pain in the left hypochondriac region and the left shoulder; the abdomen was flat and soft, with no tenderness, and no mass was palpated. No abnormality was found in a complete blood count, biochemical studies and tumor markers. Auscultatory findings of the chest were normal. A slight decline was

observed in respiratory function. Blood gas analysis showed slightly decreased PO₂ of 62.9 torr in room air. A spirogram showed almost normal data; vital capacity of 1.82 L, % vital capacity of 92.9%, forced expiratory volume in one second of 1.37 L, and % forced expiratory volume in one second of 75.3%. On chest radiography, a mass was observed in the left lower lung field, overlapping the shadow of the heart and the diaphragm, and the mediastinum was pushed towards the right (Fig. 1). Computed tomography (CT) revealed a mass of low density of heterogeneous nature, affecting the left thoracic cavity through the lateral segment of the liver in the abdomen; pleural effusion was also revealed in the left lung (Fig. 2a, b). On magnetic resonance imaging (MRI), a large mass was seen, contacting the chest wall, heart and liver, presenting low signal intensity at T1 enhancement and iso-intensity at T2 enhancement (Fig. 3). Angiography presented no abnormal findings.

Laparoscopic surgery was performed. In the left upper quadrant of the abdomen, a large mass was found arising from the left diaphragm and protruding into the abdominal cavity. It was suspected to have invaded the lateral segment of the liver. While performing a biopsy, the mass was revealed to crumble and have a tendency to bleed. Histopathological examination revealed proliferation of highly atypical spindle cells, and immunohistochemical staining revealed them to be positive for anti-actin antibody. Hence the mass was diagnosed as leiomyosarcoma.

To alleviate the patient's persistent symptoms, extir-

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Fig. 1. Pre-operative chest X-ray shows a tumor shadow overlapping the heart and diaphragm. Mediastinum shifts to the right.

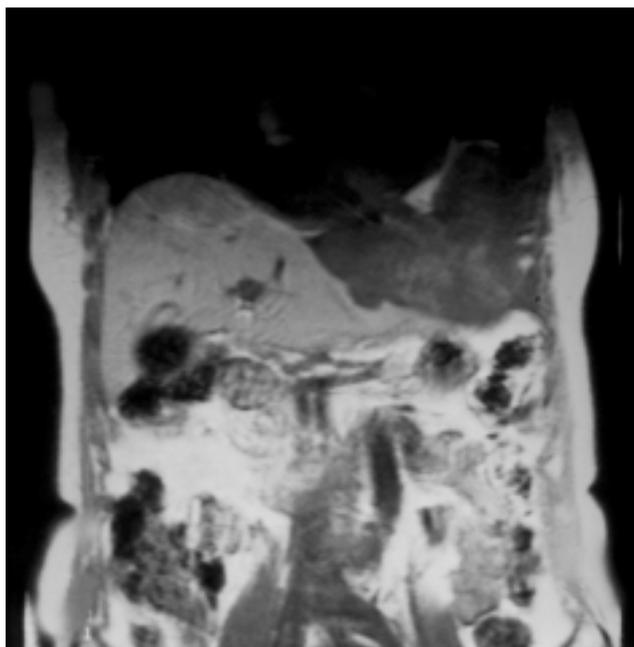


Fig. 3. Pre-operative magnetic resonance imaging of chest shows a 11.5×7.6×11.0 cm tumor in contact with chest wall, heart, and liver.

pation of the tumor was attempted. The patient was placed in the hemi-right lateral position, and thoracolaparotomy was performed via a thoracoabdominal continual diagonal incision in the 7th intercostal space. The tumor was found to occupy the anterior area of the diaphragm, and it had directly invaded the thoracic wall, left lung, and lateral segment of the liver and pericardium. The left

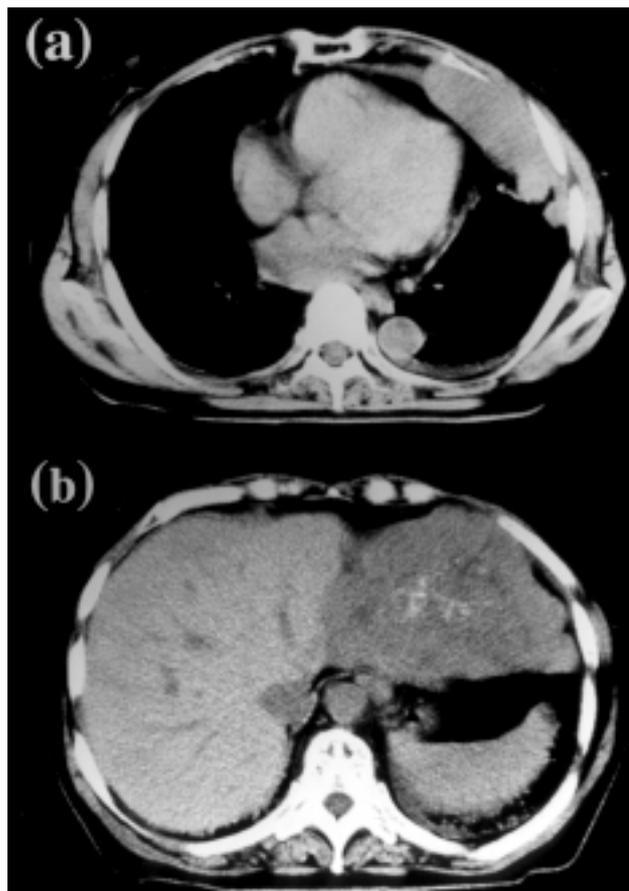


Fig. 2. a: Pre-operative chest computed tomography shows tumor adherent to chest wall.
b: Pre-operative abdominal computed tomography shows a tumor in contact with the liver.

phrenic nerve was completely involved in the tumor mass. Tumor invasion to a part of the right ventricle of the heart was also suspected, and radical operation seemed impossible. A compromise tumor resection was performed.

The resected material measured 15×13×12 cm, weighed 655 g, and was elastic and hard in consistency. A cross section showed a solid, irregular, white lesion combined with scattered foci of intratumoral hemorrhage (Fig. 4). Histologic examination detected frequent mitoses along with atypical spindle cell proliferation in the hematoxylin and eosin stained specimen, from which the diagnosis of high-grade, malignant leiomyosarcoma was established (Fig. 5).

The postoperative course was uneventful except for requiring respirator management for 11 days. The patient was discharged from hospital on the 33rd postoperative day, with somewhat improved lung function; PO₂ of 83.4 torr in room air, vital capacity of 1.20 L, % vital

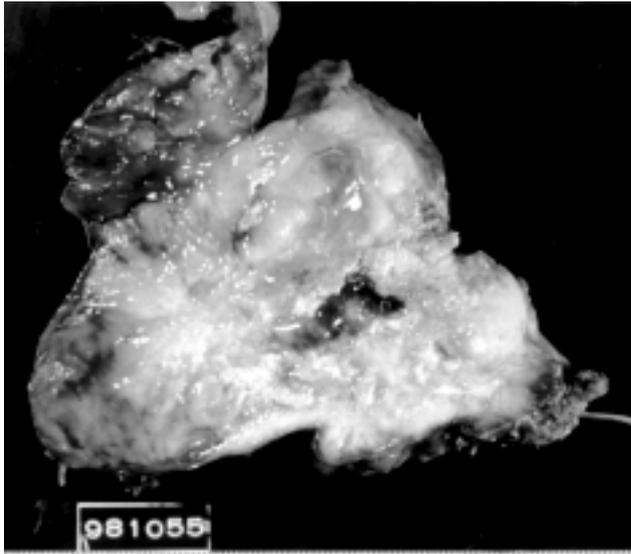


Fig. 4. Macroscopic appearance of the resected tumor, 15×13×12 cm in size, 655 g in weight, and elastic and hard in consistency. Cross section shows a white solid component with foci of hemorrhagic necrosis.

capacity of 60.3%, forced expiratory volume in one second of 1.05 L, and % forced expiratory volume in one second of 90.5%.

Unfortunately, the tumor recurred in the liver and the mediastinum, and the patient died 9 months after onset and 6 months after operation.

Discussion

Leiomyosarcoma develops relatively frequently in the uterus and digestive tract, and somewhat less frequently

in the skin, ovary, retroperitoneum and mesentery. On the other hand, primary leiomyosarcoma of the diaphragm is extremely rare. It was first reported by Kirschbaum in 1935, and so far only 7 cases have been reported to the best of our knowledge.¹⁻⁷⁾

As these tumors are mostly asymptomatic or occasionally present nonspecific symptoms, leiomyosarcomas of the diaphragm often grow large before they are detected. The rarity of this tumor makes its early identification much harder.

It is often found on chest radiography first, which reveals, however, non-specific findings such as deformation or elevation of the diaphragmatic shadow.⁸⁾ For establishment of diagnosis, US, CT and MRI are useful. In particular, MRI is useful in localizing the tumor and determining the relationship with other organs since it can give sagittal and coronal images. Diagnostic criteria for the qualitative diagnosis of a diaphragm tumor have not yet been established due to the small number of patients. The only qualitative diagnostic method is biopsy, although it is often difficult. In the present case, diagnosis was established by biopsy under laparoscopic guidance primarily for the treatment of cholelithiasis.

In treating leiomyosarcoma, resection is the most promising approach. Both chemotherapy and radiotherapy are less effective. Parker⁵⁾ and Strauch⁷⁾ reported that a complete cure could be obtained only by surgery in leiomyosarcoma of the diaphragm. Except for the case in a 2-month-old child of Koudriavstev,⁴⁾ the treatment and prognosis of the previous seven cases were as follows. Only surgical resection was done in four cases. Two patients died at 9 months and 14 months. Two other pa-

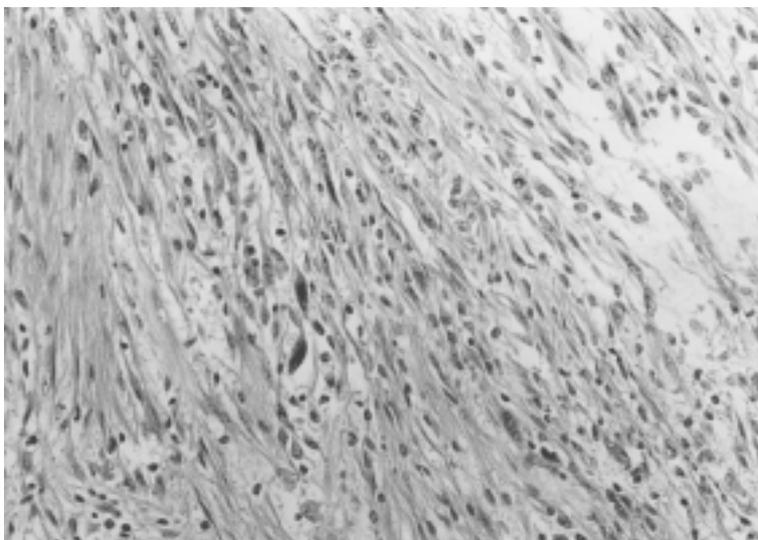


Fig. 5. Microscopic appearance of the resected tumor showing mitosis. (hematoxylin and eosin stain, ×400)

tients were still alive at 12 months and 24 months. Only radiotherapy was done in two cases. One died within 3 months and the other in 26 months. In only one case⁶ was combination chemotherapy done for relapse after surgery, and the patient was still alive after 51 months.

Actually, physical signs are mostly absent for tumors of the diaphragm. Therefore such tumors are diagnosed late and are already large. Once the tumor invades the surrounding tissue, curative operation becomes difficult and the prognosis with only surgical treatment is poor as in the present case. On the other hand, Blondeel⁶ treated a patient with combination chemotherapy for relapse after surgery. The chemotherapy consisted of iphosphamide (5 g/m² over 3 days) and adriamycin (50 mg/m² day 1) repeated every 3 weeks. In total, the patient received six cycles, resulting in very good partial remission and was alive after 51 months. Thus, in treating for leiomyosarcoma of the diaphragm, we should consider not only operation but also chemotherapy for relapse after surgery or incomplete resection. We would thus obtain a better prognosis.

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