[症例報告]Leiomyosarcoma of the chest wall : A case report

Leiomyosarcoma of the chest wall : A case report

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ABSTRACT

Leiomyosarcoma of the chest wall is a rare, but well recognozed clinical entity. We describe a case with leiomyosarcoma which was diagnosed by the use of immunohistochemical and electron microscopic studies. A 81-year-old woman developed a large firm tumor in her right lateral chest wall in October, 1994. A chest X-ray showed osteolytic changes in the 8th rib embedded in the tumor. A CT scan revealed a tumor arising from the chest wall along with a destroyed rib, thus indicating the tumor to be malignant. A radical resection of the tumor leaving a-3 cm free margin in the chest wall was performed. Reconstruction of the chest wall defect was performed using prosthetic material (Marlex mesh). The tumor measured 6×6×4 cm in size. The cut surface was gray-white in color and firm with a whorled appearance. The pathological examination (routine histology, immunohistochemistry, and electron microscopy) confirmed the diagnosisof leiomyosarcoma arising from the chest wall. The patient is doing well, disease-free and currently in her 5th month after surgery. *Ryukyu Med. J., 16(3)131~134, 1996*

Key words: chest wall tumor, leiomyosarcoma, immunohistochemical study, electron microscopic study

INTRODUCTION

Leiomyosarcoma is a relatively rare form of cancer. According to Togashi[1], in 1993 only fourteen cases with leiomyosarcoma of the chest wall, including four cases in Japan, could be found in the literature. The most common clinical presentation is a painful, enlarged mass. Almost all patients die within the first five years of diagnosis. Subsequently, the prognosis is very poor.

CASE REPORT

A 81-year-old woman was referred to our hospital for the management of a chest wall tumor on November 19, 1994. Approximately, one month prior to her admission, she developed an enlarged mass with dull pain in her right lateral chest wall below the breast. Physical examination revealed a nontender firm tumor, measuring 6×6 cm in size, located in her right chest wall. She had no evidence of any axillary node swelling.

A routine blood analysis revealed mild anemia and a urinalysis with mild renal dysfunction. A chest roentgenogram showed an oval tumor in the right chest wall without any abnormal shadows in either lung. A CT scan of the chest showed a solid tumor (isodense to the liver parenchyma), involving the 6th to 8th ribs (Fig.1). The tumor extended intrathoracically and involved the whole thickness of the chest wall. The tumor involving the 6th to 8th ribs was resected with a margin of 3 cm around the tumor.

The ensuing large chest wall defect, measuring 10×10.5 cm in size, was reconstructed using Marlex mesh (Fig. 2). She did not undergo adjuvant radiochemotherapy postoperatively. Her postoperative course was uneventful and without any evidence of recurrence 5 months after surgery.

PATHOLOGICAL FINDINGS

The tumor resected from the chest wall revealed that the tumor arose from the chest wall and thereafter extended to all layers of the chest wall. It measured 6×6×4 cm in size, was firm and had a monotonous contour. The cut surface was yellowish-white, and showed a whorled appear-

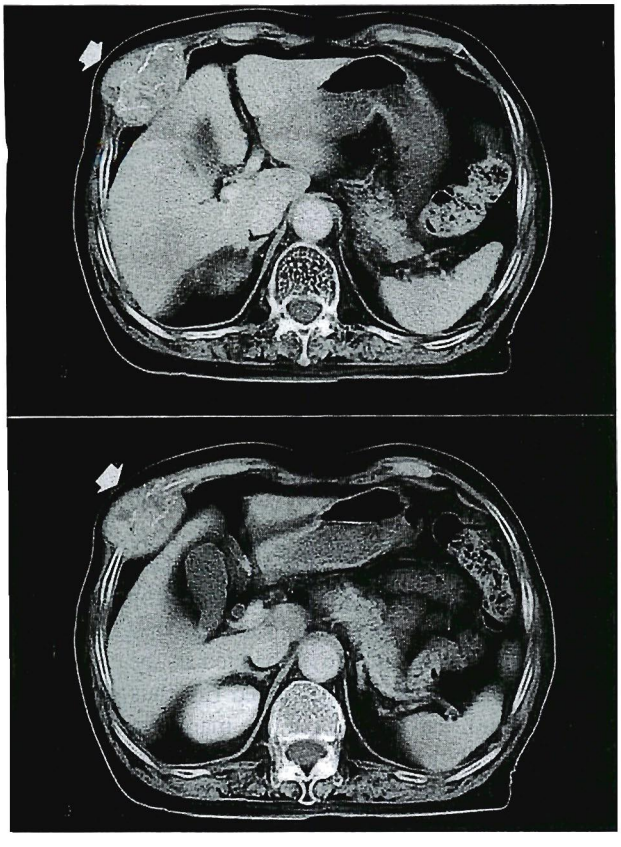

Fig.1 CT scans of the chest wall showing an oval tumor in the right chest wall with associated destruction of the rib (arrows).

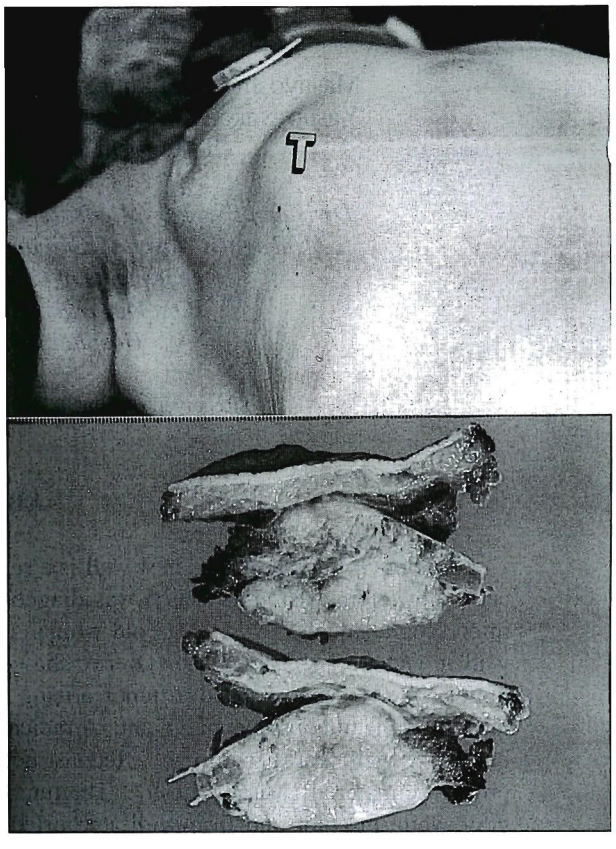

Fig.3 Macrophotographs revealing an semilunar tumor mass (T) in the chest wall at surgery (top) and a whitish-yellow, firm tumor with a whorled appearance on the cut surface (bottom).

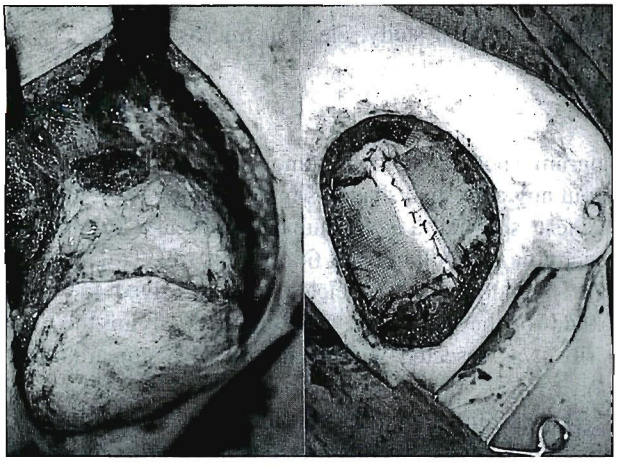

Fig.2 Operative macrophotographs showing the resection of the tumor together with the chest wall (left) and the reconstruction of the chest wall defect using a prosthetic material (right).

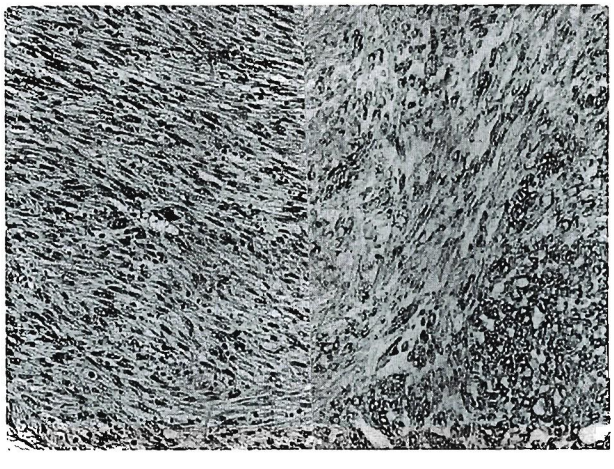

Fig.4 Microphotographs of the tumor showing interlacing bundles of spindle cells with marked hyperchromatism and frequent mitotic figures (left; HE, ×50) and positive staining for muscle-specific actin (right; ×50).

ance with some hemorrhage spots (Fig.3).

The tumor consisted of interlacing bundles of spindle-shaped cells with cigar-shaped nuclei and scanty cytoplasm arranged in a whorled pattern. Some areas of hemorrhage and necrosis were noted. Marked nuclear pleomorphism and hyperchromatism were observed, while mitotic figures were also frequently evident with more than five per highpower field. The tumor cells were both positive for cytoplasmic (Azan-Mallory stain) and immunohistochemical (anti α-smooth muscle actin DAKO, Kyoto) staining (Fig.4).

Electron microscopic examination revealed a distorted tumor cell, postfixed in 1% osmium tetraoxide, and embedded in Epon. Section coupled with intracytoplasmic bundles of microfilaments with a vague focal density and an

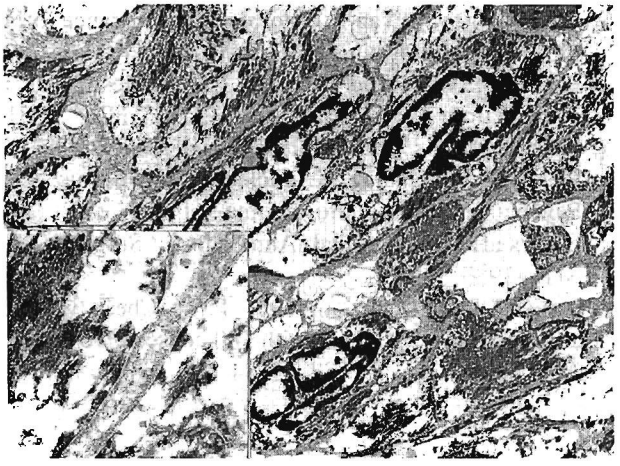

Fig.5 Electron microphotographs of the tumor revealing cellular distortion and the presence of intracytoplasmic bundles of microfilaments showing a vague density (×2,500). The inset on the lower left shows a magnification of these findings (×12,000).

elongated-shape, deeply grooved nuclei (Fig.5). The histologic ultrastructural findings of the tumor were consistent with leiomyosarcoma.

DISCUSSION

Primary tumors of the chest wall, especially leiomyosarcoma, are extremely rare[1-3]. Therefore, it is necessary that in addition to routine histopathological studies, histologic typing employing immunohistochemical and electron microscopic examinations should be done.

With a monotonous population of closely packed spindle cells in the histologic sections, the main differential diagnoses in this case included such other primary sarcomas of the chest wall as fibrosarcoma and peripheral nerve sheath tumors[2]. However, the uniform and cytoplasmic fibrillary staining using Azan-Mallory and immunohistochemical stains for muscle-specific actin in our case distinguished this type of tumor from other primary sarcomas of the chest wall[4]. Moreover, at the ultrastructural level, intracytoplasmic bundles of microfilaments with focal density were noted in the tumor cells[5]. These features are characteristic of leiomyosarcoma.

Smooth muscle tumors in the soft tissue, such as that found in the chest wall, are very uncommon and often arise from blood vessels. Smooth muscle cells constitute a significant portion of the blood vessel wall[3], but the origin of the above described tumor remains unclear. The most common sites in which smooth-muscle tumors develop are the subcutaneous soft tissues in the walls of the gastrointestinal, urinary and genital tracts[6]. The retroperitoneum is not a frequent site of leiomyomatous tumor development[6].

Chest wall tumors generally present as an enlarged mass, and are also often painful. Our patient suffered from such a painful mass. Diagnostic modalities including conventional roentgenography and CT scan revealed a tumor in the chest wall with pulmonary involvement. A CT scan revealed a tumor arising from the chest wall with involvement of the ribs, indicative that it is malignant.

Chest wall tumors suspected of being a primary tumor should be diagnosed by excision rather than by an incisional or needle biopsy. When such tumors are diagnosed to be malignant, wide excision should be considered appropriate. The extent of resection, however, should not be compromised because of an inability to close the chest wall defect[7]. Opinions differ as to what constitutes a wide resection. Many patients with malignant tumors have at least a 2-cm margin resected around the tumor, while others have a 4-cm margin. The extent of the resected margin does not influence survival[2,8,9], although a different rate of tumor recurrence was evident in the resected group and the non-resected group.

Certainly, many surgeons are of the opinion that a margin grossly free from the tumor by several centimeters would normally be an adequate resection[2]. Although this may be sufficient for low-grade malignant primary tumors, higher-grade tumors have the potential to spread within the chest wall and along the pleura. Consequently, an excision of those higher-grade tumors with a 2-cm margin would not be an adequate resection. Based on these data, it is widely accepted that a resection of all primary malignant neoplasms should include all of the involved soft tissue or bone as well as a 4-cm margin of normal tissue on all sides[2,8].

Chest wall reconstruction should include the stabilization of the bony thorax and the coverage of any soft tissue defects. The decision not to reconstruct the skeleton depends on the size and location of the defect. Defects measuring less than 5 cm in greatest diameter anywhere on the thorax can usually be left without reconstruction. Larger defects, however, should be reconstructed. In general, the stabilization of the bony thorax can be accomplished with such prosthetic material as Prolene mesh or 2 mm Gore-Tex Soft-Tissue Patch[7,8]. We used Marlex mesh without any respiratory insufficiency. Soft tissue reconstruction is best accomplished by muscle transposition. The pectoralis major, serratus anterior, latissimus dorsi and rectus abdominis muscles all have an axial blood supply which permits the elevation and rotation of these muscles[7].

In conclusion, the key to successful treatment of primary chest wall tumors remains an early diagnosis combined with an aggressive surgical resection. This procedure can now generally be performed in one stage operation, with minimal respiratory insufficiency, and with low operative mortality.

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