A Case of Retroperitoneal Malignant Mesenchymoma

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ABSTRACT

A rare case of malignant mesenchymoma of a retroperitoneal lesion, composed of liposarcoma and osteosarcoma, is reported. For complete resection of the tumor, two surgical operations were performed. The first operative material showed a mass measuring 20 x 20 x 10 cm, weighing 1607g, arising from the soft tissue of the left retroperitoneum and the tumor had a smooth surface and elastic-hard consistency. The secondary operative materials exhibited a mass measuring 10 x 5 x 3 cm, weighing 268g. The tumor was composed of soft gelatinous tissues and adhered to the tail of the pancreas but was separate from the spleen. More than 3 years after the secondary operation, no recurrence has been observed. Immunohistochemical examinations showed that S-100 protein and non-specific enolase were present in the liposarcomatous area and that vimentin was positive in the osteosarcomatous area.

Key words : Malignant mesenchymoma, Retroperitoneum, Liposarcoma, Osteosarcoma

INTRODUCTION

Malignant mesenchymoma is a rare soft tissue tumor showing two or more separate and distinct types of malignant mesenchymal differentiation to an undifferentiated sarcomatous component. The clinical and pathological features of a case of malignant mesenchymoma arising from a retroperitoneal lesion around the left kidney are presented.

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Case Summary

A 78-year-old man who complained of epigastric pain was admitted to Higashi-Sapporo Hospital on June 5, 1995. Physical examination showed a surgical operation scar for an appendectomy about 40 years earlier and a child-head sized mass was felt in the left abdomen. He had experienced angina pectoris about 15 years previously and was operated on for hyperplasia of the prostate in 1990. Laboratory examinations showed no abnormal data. Computed tomography demonstrated a large low-density mass with large calcified areas in the retroperitoneal region (Fig. 1A and B). An operation was carried out on June 28 and the large mass was easily separated and removed. After the operation, a small low-density mass was found to remain between the pancreas and the spleen (Fig. 1C). Therefore, a second opera-

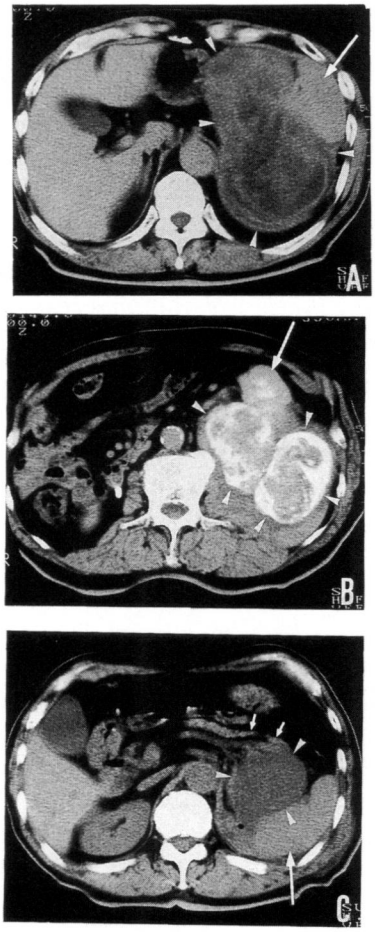

Figure 1
Abdominal CT of the retroperitoneal tumor. (A) Low-density area of the retroperitoneal tumor (arrowheads) pressing the spleen (an arrow). (B) Two high-density nodules surrounding with low density areas (arrowheads). Enhanced left kidney (an arrow) is in the ventral position of the tumor. (C) Two weeks after the first operation, a remnant of the tumor (arrowheads) is located between the tail of pancreas (small arrow) and the spleen (large arrow).

tion was conducted on July 26, 1995. The small mass that adhered to the tail of the pancreas was removed. Neither chemotherapy nor radiation was done after the operation. Although about 3.5 years have passed, he is free of the disease.

Materials and Methods

Tumor tissues obtained by surgery were fixed in 10% neutral formaldehyde and embedded in paraffin according to conventional procedures. Twenty-three blocks of the tumors were examined and 5-μm-thick sections were stained with hematoxylin and eosin (H&E). Some sections were stained with periodic acid Schiff (PAS), Alcian blue, and Sudan III. Immunohistochemistry was carried out using the avidin-biotin complex method (Vectastain ABC Elite Kit). Primary antibodies used are shown in Table 1.

Table 1 Histochemical Examinations

Staining	Company	Liposarcomatous area	Osteosarcomatous area
Keratin (PKK1)	Labsystems	(-)	(-)
Vimentin	DAKO	(+/-)	(++)
Desmin	DAKO	(-)	(-)
Smooth Muscle Actin	PROGEN	(-)	(-)
Factor VIII Related Antigen	DAKO	(-)	(-)
S-100	Nichirei	(+)	(-)
Non-specific Enolase	Nichirei	(+)	(-)
Sudan III		(+)	(-)

Pathological Findings

Gross pathological examination of the first operative material showed a mass measuring 20 x 20 x 10 cm, weighing 1607g. The tumor had a smooth surface and elastic-hard consistency, and neither invasion nor adhesion was observed. The cut surface of the mass showed that the tumor was composed

of at least two different tissue components; one consisted of small gelatinous nodules, whereas the other was of hard calcificated large nodules (Fig. 2).

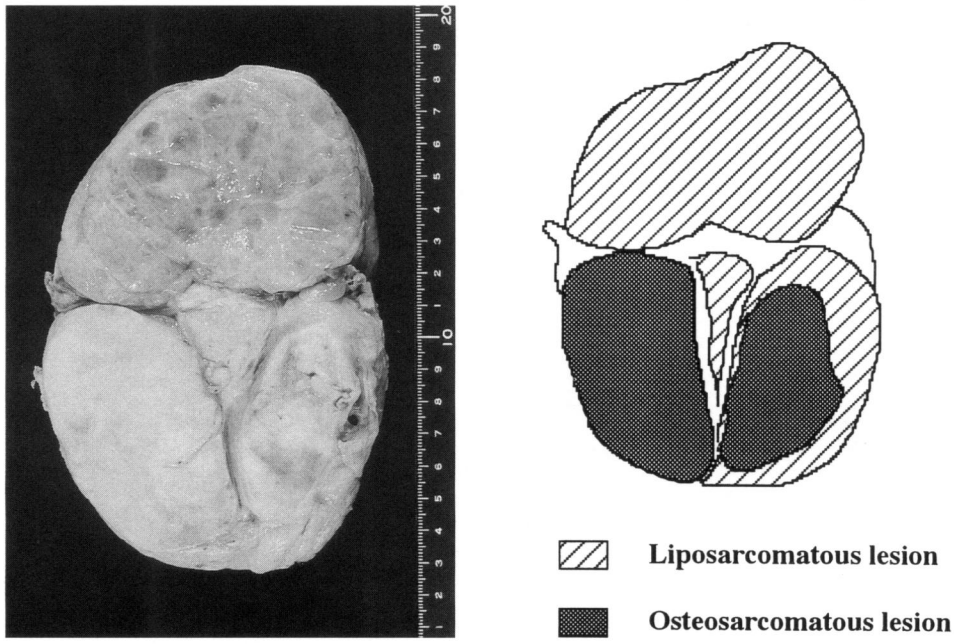

Figure 2 Gross appearance (left) and schematic drawing (right) of a frontal section of the tumor.

Microscopical diagnosis was malignant mesenchymoma composed of two-different mesenchymal elements, liposarcoma (Fig. 3A) and osteosarcoma (Fig. 3B). The predominant component was of liposarcoma with a mixed pattern ranging from well-differentiated lipoma-like areas to myxoid with a capillary vascular pattern (Fig. 3C). The myxoid liposarcomatous area was separated by fibrous tissues and capillary vessels (Fig. 3A), and showed proliferation of small oval lipoblasts and collagen tissue with an abundance of myxoid material (Alcian blue-positive) and vessels with positive findings for factor VIII related antigen and α-smooth muscle actin (Table 1). Multivacuolated lipoblasts were rarely observed within lipomatous portions and the fibrous area (Fig. 3E). Some signet-ring type lipoblasts that were immunohistochemically positive for anti-S-100 antibody were observed (Fig. 3F). The presence of osteoid surrounded by malignant osteoblasts was observed (Fig. 3D). The osteoblasts were positive for anti-vimentin antibody. The osteoid was sometimes calcificated and surrounded by osteoclast-like multinucleated giant cells.

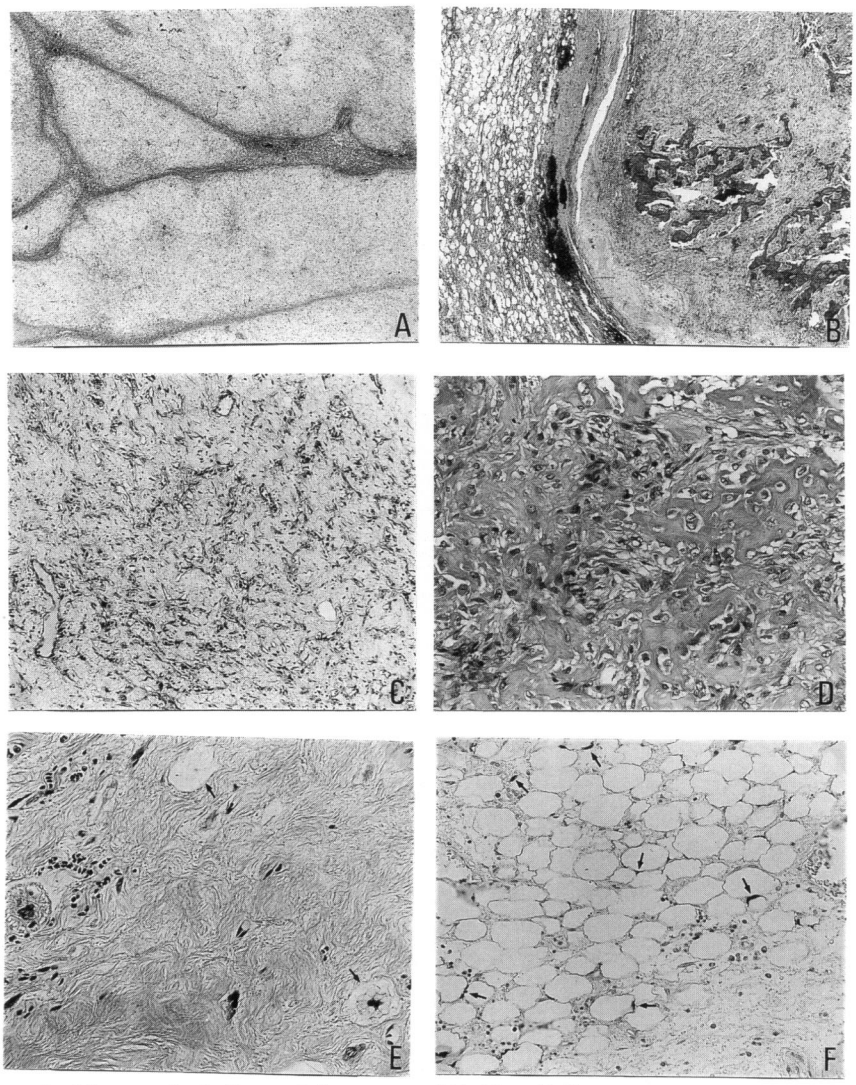

Figure 3 Microscopic findings of the tumor. (A) Myxoid liposarcomatous area separated by fibrous tissues and capillary vessels (H&E). x16.5 (B) Osteosarcomatous area with calcifications surrounded by a liposarcomatous area (H&E). x16.5 (C) Myxoid liposarcoma with prominent capillary vascular pattern (H&E). x82.5 (D) Osteoids were formed and osteoblasts surrounded them (H&E). x165 (E) Multivacuolated lipoblasts in the myxomatous area (arrows; H&E). x235 (F) Immunohistochemistry for S-100 in an area of well-differentiated liposarcoma. Signet-ring cell type lipoblasts are positive (arrows). x165

Macroscopic examination of the secondarily resected materials revealed a mass measuling 10 x 5 x 3 cm, weighing 268g. The tumor was composed

of soft gelatinous tissues and adhered to the tail of pancreas but was separate from the spleen. The cut surface of the mass showed that most of the tumor consisted of small gelatinous nodules. Microscopically, the tumor also consisted of two different mesenchymal elements; the major part was liposarcoma and the remnant osteosarcoma. Although the secondary operative material adhered to the tail of the pancreas, microscopic examination revealed that the thin septum consisted of fibrous tissues separating sarcomatous tissues from the pancreatic tissue. No epithelial elements were found in either tumor.

Discussion

Malignant mesenchymoma is a rare soft tissue neoplasm first described by Gilmour[1] in 1943 and later defined by Stout[2] as a tumor composed of two or more unrelated malignant mesenchymal components in addition to any undifferentiated fibrosarcomatous element. The experience with these tumors at the Armed Forces Institute of Pathology suggests that "they can be divided into two closely related categories: (1) tumors that are clearly diagnostic of malignant mesenchymoma and are characterized by the presence of coexisting myosarcomatous (rhabdomyosarcomatous or leiomyosarcomatous) and liposarcomatous elements in the same neoplasm, often in addition to a fibrosarcoma-like spindle cell component, and (2) tumors that show, in addition to a specific and clearly recognizable type of sarcoma, foci of malignant cartilaginous or osseous tissue."[3] Most cases have occurred in the deep tissues of the retroperitoneum or thigh and most patients have been older than 55 years.[3] In the present case, typical coexistence of liposarcoma and osteosarcoma was observed, and the age of this patient was 78, exactly fulfilling the criteria. In addition, the tumor was probably derived from the retroperitoneum because it existed within the retroperitoneum and was clearly separated from the pancreas and the kidney. The histogenesis of this rare entity remains uncertain. Because the majority of the tumors affect older patients, it seems more likely that they arise from primitive and uncommitted mesenchymal elements that have differentiated along multiple cell lines.[3]

It was reported that bone and cartilage was occasionally present in atypical lipomatous tumors[4,5] and that focal cartilaginous, leiomyomatous, or osseous mataplasia occurred in myxoid and well-differentiated liposarcoma.[6,7] Furthermore, malignant fibrous histiocytoma combined with fibrosarcoma, leiomyosarcoma, or rhabdomyosarcoma was reported to be present in retroperitoneal liposarcoma.[8] In the present case, although the appearance of a part of osteosarcoma has a possibility to be an osseous metaplasia of dedifferentiated liposarcoma, parts of osteosarcoma and liposarcoma were clearly

separate and each element almost equally ocupied in the tumor of the first resetion. Therefore, in the case that the coexistence of sarcomas differentiating to clearly different tissues is observed in a tumor like this, it may be appropriate to consider as a malignant mesenchymoma than as components of the metaplasia of liposarcoma.

It is generally thought that malignant mesenchymoma is a highly malignant neoplasm with a poor prognosis, or at least that the prognosis is related to the grade and type of tissue elements present, being least favorable in the presence of a high-grade tumor or rhabdomyosarcoma. However, Newman and Fletcher[9] suggested that malignant mesenchymoma might not be as aggressive as its high-grade histology would imply. Recently, Brady et al.[10] reported that their eight cases of malignant mesenchymoma showed a particularly aggressive form of soft tissue sarcoma. Although the prognosis of this tumor is still controversial, the prognosis of the subtypes is best with basically liposarcomatous components. Surgical removal is the preferred treatment. In the present case, the tumors were completely removed by surgery and the predominant component of the tumors was liposarcoma. Therefore, recurrence has not been observed in this case although more than 3.5 years have passed after the operation.

Acknowledgments

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