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Case Report

Retroperitoneal Liposarcoma With Leiomyosarcomatous Differentiation

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We herein describe a 60-year-old Japanese man with a giant retroperitoneal liposarcoma undergoing leiomyosarcomatous differentiation. He was admitted to our hospital because of a 5-month history of dysphagia and abdominal distention. Abdominal computed tomography showed a giant tumor that occupied the entire retroperitoneal space. The majority of the mass was lipomatous and low density; both a heterogenous and solid mass were also present. A giant retroperitoneal liposarcoma was diagnosed, and tumor resection was performed. At surgery, the tumor was mostly isolated from the retroperitoneum and other organs. Histopathologically, the tumor comprised well-differentiated and dedifferentiated liposarcoma with heterologous differentiation of the leiomyosarcomatous components, which is a rare phenomenon in liposarcoma. The patient was alive 3 years after the first treatment, although he has had 3 local recurrences (approximately one recurrence yearly) and has been treated by repeated resection and radiotherapy.

Key words: Retroperitoneum – Liposarcoma – Leiomyosarcomatous differentiation

Dedifferentiated liposarcoma (DL) is one of the most frequent sarcomas of the retroperitoneum. It is defined by the association of an atypical lipomatous tumor, namely, areas of well-differentiated liposarcoma (WDL), with a dedifferentiated component. WDL is composed of mature adipocytes and atypical stromal cells with an enlarged, hyperchromatic nucleus. Usually, the dedifferentiated part of the

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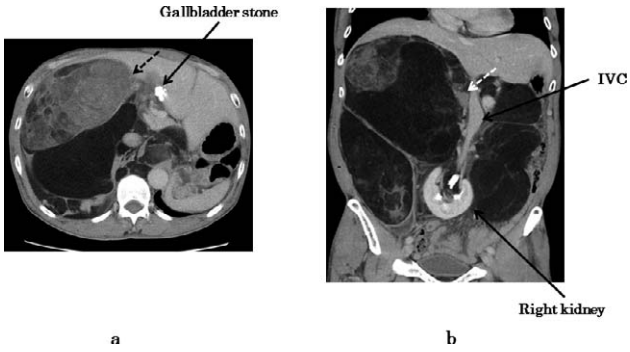


Fig. 1 Abdominal CT images (a, horizontal; b, coronal) indicated that a large lipomatous mass occupied the retroperitoneal space with a lower shift of the right kidney (black arrow). (a) A heterogenous mass with a higher density than fat (black dotted arrow) was observed in the right lower part of the liver along with a gallbladder stone (black arrow). (b) A clearly enhancing solid mass (white dotted arrow) was observed in the right upper part of the IVC (black arrow).

liposarcoma is composed of either a spindle/pleomorphic high-grade sarcoma or a mixoid/spindle cell low-grade sarcoma. The WDL component may be easily overlooked, and DL may thus be mistaken for another high-grade sarcoma. It has been reported in one study that approximately 5% of the dedifferentiated component showed heterologous differentiation, such as leiomyosarcoma, rhabdomyosarcoma, osteosarcoma, and angiosarcoma.[1] A less common phenomenon is the occurrence of WDL with leiomyosarcomatous (LMS) differentiation.[2] Limited to the retroperitoneum, only 8 cases of liposarcoma with LMS components have been reported.[3–7] We herein report a case of retroperitoneal liposarcoma comprising WDL and DL, with LMS components, treated by surgical resection.

Case Report

A 60-year-old Japanese man was admitted to our hospital in May 2010 because of a 5-month history of dysphagia and abdominal distention. Blood test results, including tumor marker levels, were within normal limits. Abdominal computed tomography (CT) revealed that a large, lobulated mass occupied the retroperitoneal space with a lower shift of the right kidney (Fig. 1a and 1b). The majority of the mass had a lipomatous density; although an 18-cm-diameter heterogenous mass with a higher density was present in the right lower part of the liver (Fig. 1a). Moreover, a clearly enhancing 2-cm-diameter solid mass was present in the right upper part of the inferior vena

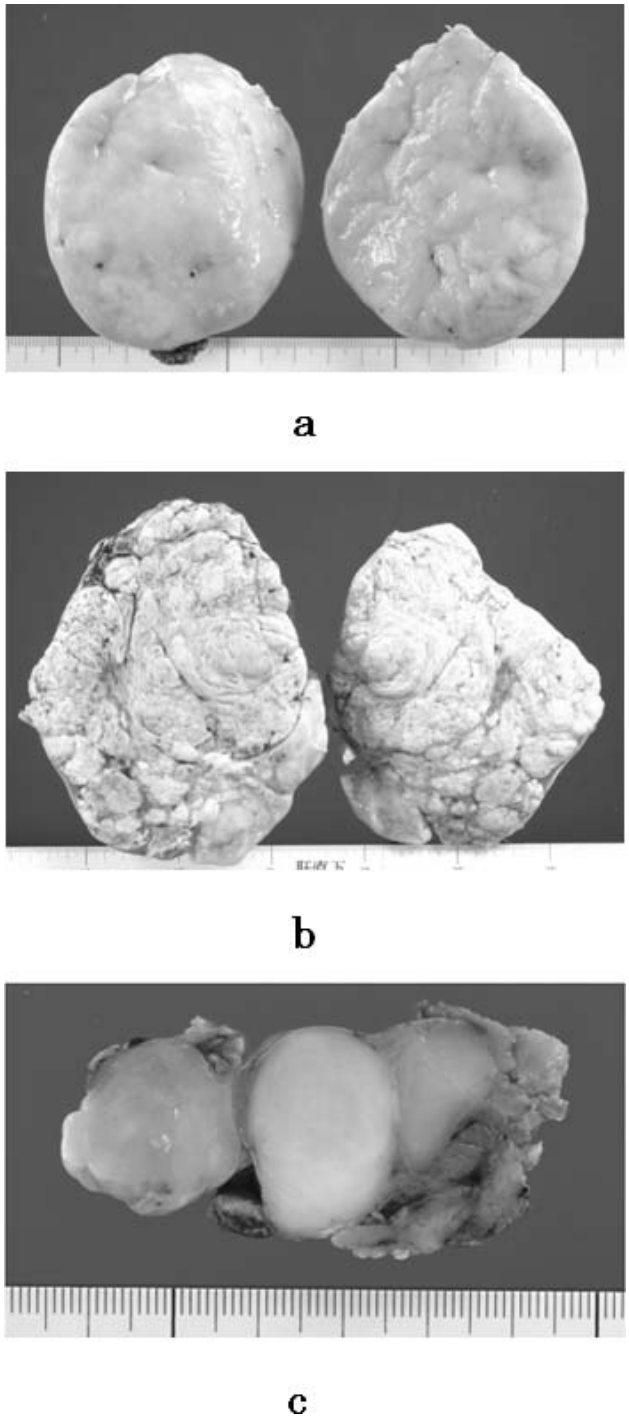
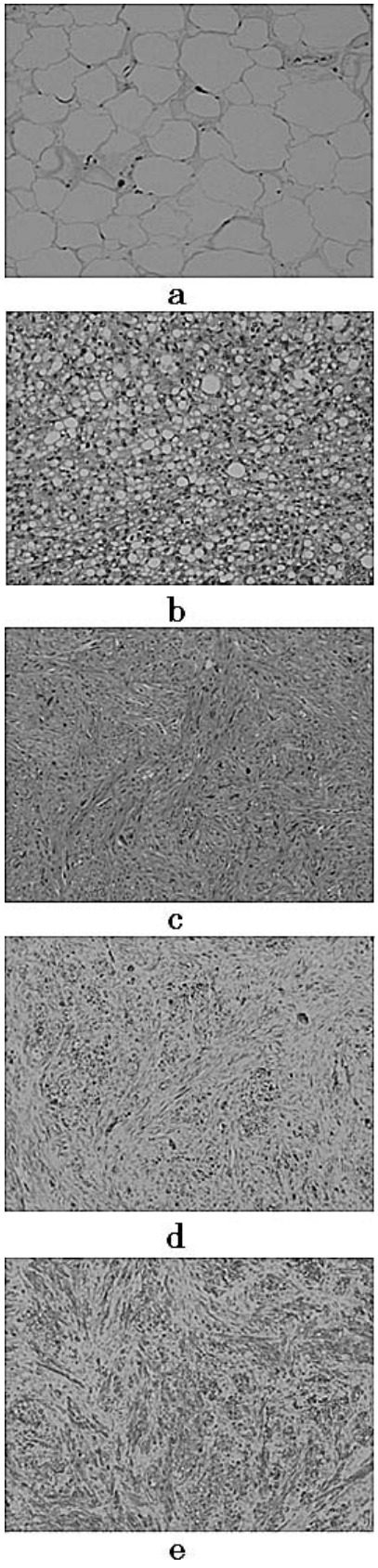


Fig. 2 Sectioned slices of separately resected specimens. (a) Most nodules were pale yellow to yellow-tan and uniform in shape. (b) The nodule in the right lower part of the liver was yellow-tan and lobulated. (c) The nodule in the right upper part of the IVC was white and uniform in shape.



cava (IVC) (Fig. 1b). We diagnosed a giant retroperitoneal liposarcoma and performed a tumor resection in June 2010. At surgery, the tumor occupied the entire abdominal cavity, but it was mainly present in the right upper part of the abdomen. The tumor was mostly isolated from the retroperitoneum and other organs, but detachment was needed only for the fixation near the right upper part of the lumbar vertebrae. The whole tumor measured about 40 cm in the maximum diameter, with a weight of 11 kg, was resected. Macroscopically, most of the nodules were pale yellow to yellow-tan and uniform in shape (Fig. 2a), whereas the nodule in the right lower part of the liver was yellow-tan and lobulated (Fig. 2b), and in the right upper part of the IVC, the nodule was white and uniform in shape (Fig. 2c).

Histologically, both the pale yellow and yellow-tan nodules included WDL to DL tissue (Fig. 3a), whereas the yellow-tan lobulated nodule was a DL (Fig. 3b). Moreover, the white nodule had eosinophilic intersecting fascicles composed of slender or slightly plump cells with spindle, oval, or pleomorphic nuclei (Fig. 3c). Immunohistochemically, the cells were strongly positive for desmin (Fig. 3d) and alpha-smooth muscle actin (Fig. 3e) and negative for S-100 protein (data not shown). These histologic and immunohistochemical results indicated LMS differentiation.

The postoperative course was uneventful, and the patient was discharged 12 days after surgery. Although he was alive for about 3 years from the first treatment, he had 3 local recurrences (approximately one recurrence per year) near the right upper part of the lumbar vertebrae. Thus, he was twice treated by repeated resection, including right nephrectomy. The histologic results of the resected specimens were almost identical to those of the first histologic examination. However, because the tumor invaded the IVC at the third recurrence, we judged resection to be impossible. He could not receive chemotherapy because of a low renal function after the right nephrectomy. Since then, he has been treated by radiotherapy.

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Fig. 3 Microscopic examination. (a) Both the pale yellow and yellow-tan nodules comprised WDL to DL tissue (H&E; ×100). (b) The yellow-tan lobulated nodule was a DL (H&E; ×100). (c) The white nodule had eosinophilic intersecting fascicles composed of slender or plump cells with spindle, oval, or pleomorphic nuclei (H&E; ×100). (d, e) Strong immunohistochemical reactions for desmin and alpha-smooth muscle actin were observed in the cells of the white nodule (×100). H&E, hematoxylin and eosin.

Discussion

Retroperitoneal liposarcomas account for 15% to 30% of all liposarcomas and 35% of retroperitoneal sarcomas.[8,9] They represent a relatively well-differentiated liposarcoma, while a minority of retroperitoneal liposarcomas are DLs.[3–7] Morphologically, the clinical behavior of this histologic subtype of liposarcoma is expected to be more aggressive, which has been well documented.[2,4] Approximately 5% of the dedifferentiated components showed heterologous differentiation.[1] LMS differentiation is quite rare, and only 8 cases of retroperitoneal liposarcoma with LMS components have been reported to the best of our knowledge.[3–7]

Malignant mesenchymoma, first described by Stout in 1948, is a malignant soft tissue tumor with 2 or more distinct mesenchymal components.[10] Strict diagnostic criteria for malignant mesenchymoma currently require that each component is sufficiently differentiated histologically. The malignant components that normally fall into this category are liposarcoma, leiomyosarcoma, rhabdomyosarcoma, osteosarcoma, chondrosarcoma, and angiosarcoma. However, liposarcoma occasionally shows heterologous differentiation, such as LMS differentiation.[1] After review of 8 cases of liposarcoma with LMS differentiation limited to the retroperitoneum,[3–7] the histologic results of liposarcoma revealed WDL in 3 cases, DL in 4 cases, and both DL and WDL in 1 case. Moreover, LMS differentiation was observed in the primary tumor in 3 cases, the recurrent tumor in 4 cases, and both the primary and recurrent tumor in 1 case. In our case, LMS differentiation was seen with DL and WDL or in the primary and recurrent tumor. These findings suggest that liposarcoma in itself may have the potential for LMS differentiation regardless of the histologic type or time at which it occurs. However, the mechanisms of LMS differentiation remain to be elucidated.

Malignant mesenchymomas, including retroperitoneal liposarcomas, are generally considered to be highly malignant with a poor prognosis.[11] Although the behavior of liposarcomas with LMS differentiation has not been clearly defined, mostly because of their rarity, the outcome of patients with liposarcomas with LMS differentiation reportedly does not differ from that of patients with conventional liposarcomas not limited to the retroperitoneum.[1] It is known that the treatment of liposarcoma with LMS differentiation is surgical resection, which is the same as the treatment of conventional liposarcoma. Because conventional radiotherapy or chemotherapy for liposarcoma is ineffective in improving the prognosis, surgical resection is usually the first choice. Therefore, we performed repeated resection for the primary and recurrent tumors in this case. However, the outcome was not improved by surgery alone. Therefore, the efficacy of multimodal therapy, such as radiotherapy or chemotherapy plus surgery, should be examined in the future.

In summary, we reported a rare case of retroperitoneal liposarcoma with LMS components, which is valuable in terms of examining the heterologous differentiation of liposarcoma.

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