

CASE REPORT

A case of perinephric liposarcoma which recurred ten years later from the initial operation

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Abstract : A 58-year old man was referred to our hospital for treatment of an abdominal mass. As for him, tumor resection with right nephrectomy had been performed ten years ago for a giant well-differentiated perinephric liposarcoma. CT examination showed a huge tumor shadow in the abdominal cavity. Abdominal MRI examination showed a 15×8 cm tumor with almost high signal intensity on the T2 weighted images. At lapalotomy, a large bulky retroperitoneal tumor pointed out before an operation was found. Surgical extirpation of the tumor was performed. Besides, several tumors of the thumb head size were detected into right retroperitoneal fatty tissue. The right side mesocolon and the tumors were not able to exfoliate, therefore right hemicolectomy was performed. Histological features showed dedifferentiated liposarcoma. The postoperative course was uneventful. But eight months after surgery, he was admitted again for treatment of a 4×3 cm retroperitoneal tumor. Extirpation of the tumor was performed. Histological finding of this tumor also showed dedifferentiated liposarcoma. Dedifferentiation, occurring in 15% of the well-differentiated liposarcomas, sometimes may develop later. Long-term detailed follow-up is necessary for well-differentiated liposarcoma. *J. Med. Invest.* 58 : 154-158, February, 2011

Keywords : *perinephric liposarcoma, retroperitoneal liposarcoma, dedifferentiated liposarcoma, well-differentiated liposarcoma*

INTRODUCTION

Soft tissue sarcomas are uncommon tumors that represent only 1 to 1.5% of all malignancies (1). Of these tumors, only 15% are located in the retroperitoneum. We herein describe a case of perinephric liposarcoma which recurred ten years later from the initial operation. At the first operation, histology

of the tumor was well-differentiated type. By contrast, the recurrent histological type of the tumors showed dedifferentiation.

CASE REPORT

A 58-year old man was referred to our hospital for treatment of an abdominal mass. As for him, tumor resection with right nephrectomy for a 22×19×16 cm well-differentiated right perinephric liposarcoma had been performed ten years ago (Fig. 1). Abdominal computed tomography (CT) examination was performed every year after initial

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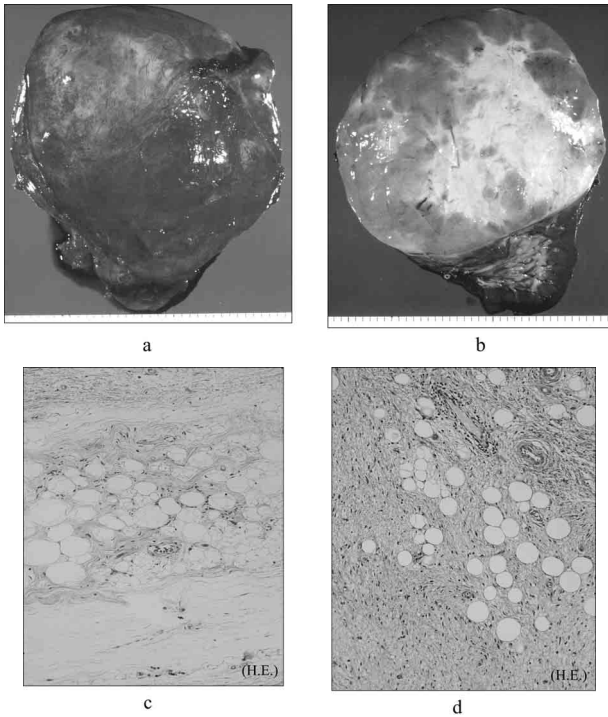


Fig. 1 : The excised tumor size was 22×19×16 cm (1-a, 1-b). Pathological examination revealed a well-differentiated liposarcoma (HE) (1-c, 1-d).

surgery. Until 1 year before this admission, CT showed no recurrence sign. Laboratory findings at this admission showed no abnormalities on peripheral blood and serum examination except slight renal function disorder. Abdominal CT examination showed a huge tumor shadow having well-defined margins in the abdominal cavity (Fig. 2). Abdominal

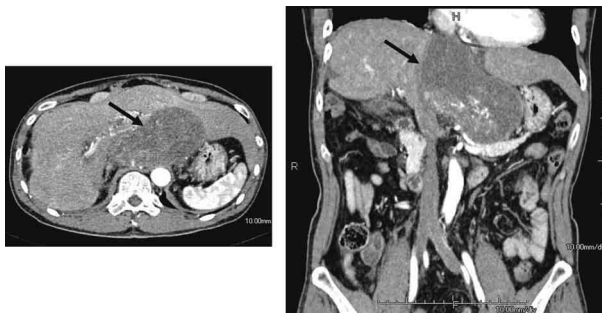


Fig. 2 : CT demonstrated a huge tumor shadow having well-defined margins in the abdominal cavity (arrow).

magnetic resonance imaging (MRI) examination showed a 15×8 cm tumor with almost low signal intensity on the T1 and with almost high signal intensity on the T2 weighted images (Fig. 3). Furthermore, the tumor had the part where showed high signal intensity on the T1 and T2 weighted images in MRI. That part regarded as the adipose constituent. The area accompanied with homogeneous low signal intensity on the T1 weighted image in MRI

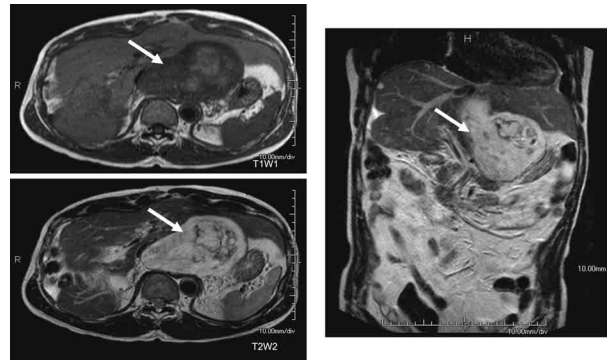


Fig 3. MRI demonstrated a 15×8 cm tumor with almost low signal intensity on the T1 and with almost high signal intensity on the T2 weighted images (arrow).

was regarded as a myxomatous component. Considering the above facts, we presumed that well differentiated liposarcoma had converted to the other differentiation type. Though the definitive diagnosis was not provided, the tumor was thought to be a recurrence of liposarcoma. At laparotomy, a bulky tumor was found in the retroperitoneal adipose tissue of the left upper abdominal region. The tumor showed expansive growth without any invasion into surrounding structures such as the liver, pancreas, or inferior vena cava. Extirpation of the tumor was performed. The tumor size was 15×9×8 cm (Fig. 4). Besides, several tumors of the thumb head size were detected into right retroperitoneal fatty tissue. The right side mesocolon and the tumors were not

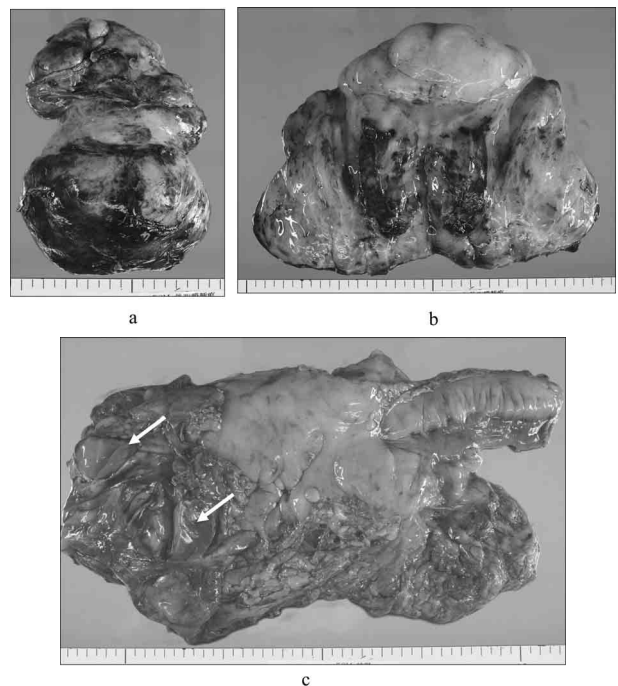


Fig. 4 : The excised tumor size was 15×9×8 cm (4-a). Cut surface (4-b). Besides, several tumors of the thumb head size were detected into right retroperitoneal fatty tissue (4-c) (arrow).

able to exfoliate, therefore right hemicolectomy was performed. Histological features of the tumors showed dedifferentiated liposarcoma (DDLPS) (Fig. 5). The postoperative course was uneventful and the

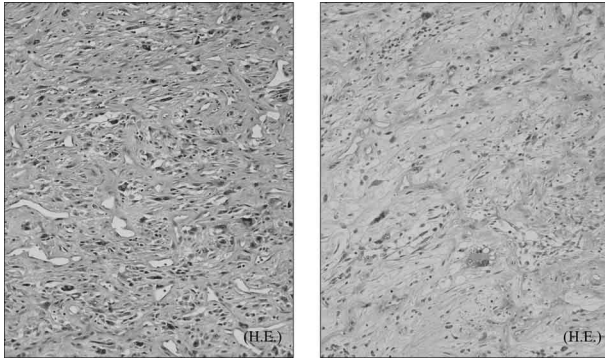


Fig 5 : Pathological examination revealed dedifferentiated liposarcoma (HE).

patient was discharged soon. But eight months after surgery, he was admitted again for treatment of a 4×3 cm tumor in the retroperitoneal region (Fig. 6). Extirpation of the tumor was performed. Histological features of this tumor showed local recurrence of the DDLPS (Fig. 7). He got well immediately and left the hospital.

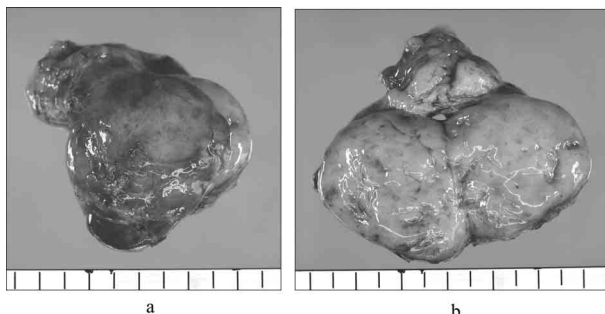


Fig. 6 : The excised tumor size was 4×3 cm (6-a). Cut surface (6-b).

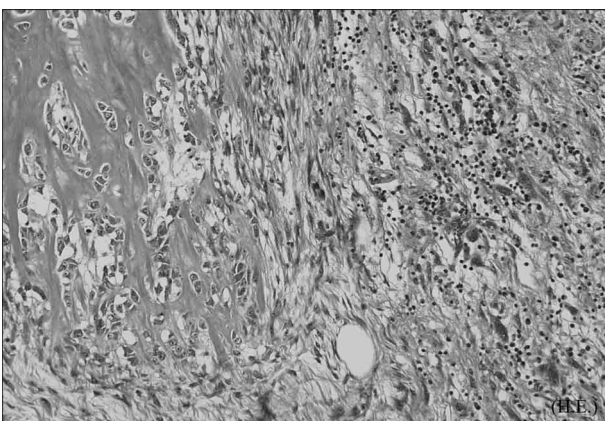


Fig. 7 : Pathological examination revealed a dedifferentiated tumor (HE).

DISCUSSION

Liposarcomas, corresponding to 19% of all soft tissue sarcomas, are malignancies of adipose tissue (2). They are the most frequent histopathological variety (41%) of the retroperitoneal sarcoma (3). The World Health Organization classification of soft tissue tumors has classified liposarcoma into five main subgroups : well-differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes, myxoid, round cell, pleomorphic and dedifferentiated (4). More than 90% of the retroperitoneal liposarcomas are well-differentiated type (56%) and dedifferentiated type (37%) (5).

Dedifferentiation is defined as the presence of nonlipogenic high grade areas within the well-differentiated liposarcoma (WDLPS). Dedifferentiation is rare, occurring in 15% of the WDLPS (6). About 90% of DDLPS arise de novo, while 10% occur in recurrence. According to current literature, WDLPS and DDLPS share the same basic genetic abnormality characterized by a simple genomic profile with a 12q14-15 amplification involving MDM2 gene (7). The risk of dedifferentiation is higher in the retroperitoneum and is probably a time-dependent phenomenon (8-10). In our present case, initial histological type was a well-differentiation, whereas recurrent histological type was a dedifferentiation. It took for a long time, more than about 9 years, till the histological change from well-differentiation to dedifferentiation occurred. At the moment histological change occurred, the recurrent tumor grew rapidly.

As for treatment of liposarcoma, surgical resection is considered the mainstay of curative treatment, and complete surgical resection with negative margins is required as the goal of therapy for most patients. However, liposarcoma forms a pseudo capsule without having a capsule histologically, so that a border is indistinct. Therefore, an aggressive surgical technique with en-bloc multiorgan resection is necessary in order to achieve negative margins (5, 11-14). The most frequent organ resected is the kidney like our first resection was so. Even with aggressive surgical approaches, local recurrence remains a common type of failure (13). As to our case, to keep negative margins, we performed right nephrectomy at initial surgery, and right hemicolectomy at second surgery. Despite aggressive surgical procedures performed mentioned above, we failed in the local control 2 times, and the third surgery performed against the recurrent tumor. Fortunately,

the patient is alive without recurrence after the third operation. If a resectable local recurrence tumor appears in future, we shall extirpate the tumor.

Some articles report that retroperitoneal WDLPS does not recur after an operation (15-18). It seems WDLPS has favorable prognoses. However, about these cases, postoperative follow-up survey period is a short term. Like our present case, long term follow-up for at least 10~20 years has revealed that WDLPS in the retroperitoneum recur regionally in almost 100% of cases even if the tumor seems to have been completely resected (7).DDLPS in the retroperitoneum also recur in almost 100% of cases and often cause the patient's death (7). In our case, DDLPS recurred regionally in a short term. The interval of the second operation and the third one was only eight months.

The effectiveness of chemotherapy and radiation therapy for both primary and metastatic liposarcomas is still controversial (19-22). Survival benefits have not been demonstrated (23). Hence, we are using no chemotherapy or radiation therapy. But if a non-resectable local recurrence lesion appears in future, we will consider radiotherapy such as heavy particle beam for a purpose of the regional control (24).

In conclusion, we have experienced a case of huge perinephric WDLPS which recurred ten years later from the initial operation. Once WDLPS recurs, recurrence may cause the tumor to evolve into a higher grade of sarcoma or to dedifferentiate. A long-term follow-up after surgery is mandatory due to high rates of recurrence.

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