Surgical Treatment of Retroperitoneal Liposarcoma

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ABSTRACT

Background Retroperitoneal liposarcoma (RL) is a relatively rare tumor and is usually found at the advanced stage. Chemotherapy or radiotherapy for this tumor is not yet defined, and if operable, surgery is the treatment of choice. Complete resection of tumor with wide margins including excision of other organs has been recommended. However, many patients suffer from deterioration of the quality of a postoperative life. In the present study, we retrospectively analyzed the ideal surgical procedures for treating RL.

Methods RL patients treated at our institute between 2003 and 2013 amounted to 10. RL was primary in 5 patients and recurrent in the rest 5. We analyzed cases of the 10 patients retrospectively.

Results Tumor resection was performed for 9 patients, 7 of whom underwent complete tumor resection. RL was well-differentiated in 6 patients and dedifferentiated in 4. We analyzed the overall survival of 10 patients, and the relapse free survival of the operated 9 patients. Patients with well-differentiated RL showed better survival than those with dedifferentiated RL. Even the recurrent RL was huge, complete tumor resection could be performed in the well-differentiated type, but it was difficult in the dedifferentiated type.

Conclusion In the recurrent huge RL, the chance of a margin-negative resection remains low, but surgery remains the treatment of choice. Tumor resection with preserving important organs may improve patients' quality of postoperative life and survival.

Key words dedifferentiation; quality of life; recurrence retroperitoneal liposarcoma; surgery

Retroperitoneal sarcomas are relatively rare. Among them, liposarcoma is one of the most common subtypes. Retroperitoneal liposarcoma (RL) is classified into 5 subtypes according to the World Health Organization Classification: well-differentiated, dedifferentiated, myxoid/round cell, pleomorphic and mixed-type.[1] The peak incidence is in the 6th decade of life[2] and the incidence is almost equal in men and women with a slight male predominance.[3]

RL is often asymptomatic and considerably large when initially diagnosed. Complete resection for curative intent is the treatment of choice, and a negative margin should be achieved to improve survival, even if resection of adjacent organs is needed.[4–8] But, the results of surgery for RL have been quite inferior to those of extremity sarcomas because of the large size at operation and difficulty in resecting the tumor due to its anatomical location. Furthermore, many patients suffer from high frequency of local recurrence of tumor after such radical and complete operation. Neoadjuvant and adjuvant treatments with chemotherapy or radiotherapy have also not shown any consistent benefit.[9, 10] This paper shows the surgical treatment results and prognosis of RL in a single institute of Japan.

MATERIALS AND METHODS

Between 2003 and 2013, a total of 5,590 patients visited our institute. Retroperitoneal tumors were detected in 43 patients (0.8%). Among them, 10 patients (10/43, 23.3%) had RL. Also, malignant fibrous histiocytomas and leiomyosarcomas were found in 10 and 7 patients, respectively, and metastatic tumors, in 3. Recent findings on the genetic field suggest that most lesions diagnosed as malignant fibrous histiocytomas are in fact dedifferentiated liposarcomas.[11, 12] However, diagnosis of disease known as malignant fibrous histiocytoma is not esablished now. Thus, in the present study, we registered only the tumors histologically proved to be liposarcoma.

The patients' records, operative notes, histopathological reports and imaging studies were reviewed retrospectively. Tumor size and tumor invasion were visualized by enhanced computed tomography and/or magnetic resonance imaging. Clinicopathological characteristics, treatment procedures and prognoses were analyzed in the 10 patients.

RESULTS

Patients' characteristics are indicated in Table 1. On visiting our department, 5 patients had primary lesions, and another 5, recurrent lesions. Well-differentiated RL was detected in 6 patients, and dedifferentiated RL, in 4.

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Abbreviation: RL, retroperitoneal liposarcoma

The follow-up period of patients ranged from 3 to 164 months. The mean survival time of 10 patients with RL was 155 months, and that of 6 patients with well-differentiated RL (157 months) was significantly longer than that of 4 with dedifferentiated RL (135 months) ($P = 0.027$, Fig. 1).

Operation (resection of RL) was performed for 9 patients, but tumor resection was avoided for 1 patient because of tumor invasion to the greater psoas muscle and lumbar bone. The 9 patients had received tumor resections for the average of 3 times (range: 2–10 times). At the initial operation, complete tumor resection was performed for 7 patients, but incomplete tumor resection for the rest 2 due to tumor invasion to neighboring organs. The average time interval between the initial resection and the 2nd (relapse free survival) in the 9 patients was 54 months (range: 9–114 months). The mean relapse free survival of 6 patients with well-differentiated RL (64 months) was longer than that of the rest 3 with dedifferentiated RL (28 months), but the difference was not significant ($P = 0.27$, Fig. 2).

The following is the course of disease with the operation procedures for recent cases of 2 patients with recurrent RL. Patient 1 was a 53-year-old female. About 10 years ago, she underwent complete resection for well-differentiated RL at a hospital. Thereafter, huge recurrent RL took place, which was resected 1 year ago at another hospital. But, because the tumor involved the left ureter and the left external iliac artery, the operation was done with incomplete extraction of the tumor. According to the rapid growth of the residual tumor, she was admitted to our institute. Abdominal enhanced computed tomography indicated dislocation of the left ureter and the left external iliac artery due to tumor invasion. Also, tumor invasion to the left iliopsoas muscle was observed (Fig. 3). At the time of operation, we found a huge tumor occupying almost the whole abdominal cavity (Fig. 4). At first, we inserted bilateral ureter stents, preserving the bilateral common iliac arteries and veins, bilateral internal and external iliac arteries and veins pulse bilateral ureters, and performed complete extraction of the tumor. The extracted tumor weighed 6,500 g, the operation time was 9.5 h and the amount of intraoperative bleeding was 2,820 mL. Patient 2 was a 65-year-old female. She underwent incomplete resection of dedifferentiated RL at a hospital 2 years ago, because tumor invaded to bilateral ureters and bilateral iliac arteries and veins. But, the residual tumor enlarged gradually and she was moved to our institute. For this patient also, bilateral ureter stents were inserted at first. Almost the whole tumor was extracted with resection of the left internal iliac artery and vein. However, the tumor invaded to the inferior vena cava deeply, incomplete extraction was partly done. The extracted tumor weighed 6,000 g, operation time was 12.5 h and the intraoperative bleed-

Table 1. Demographic details of 10 patients with retroperitoneal liposarcom (RL)

Age at initial diagnosis, mean (range), yr		57.5 (44–73)
Sex	Male/female	1/9
Variant	Well-differentiated	6
	Dedifferentiated	4
	Myxoid/round cell	0
	Pleomorphic	0
	Mixed type	0
Cases	Primary/recurrent	5/5
Complete tumor resection		7
	Primary cases	3/5
	Recurrent cases	4/5

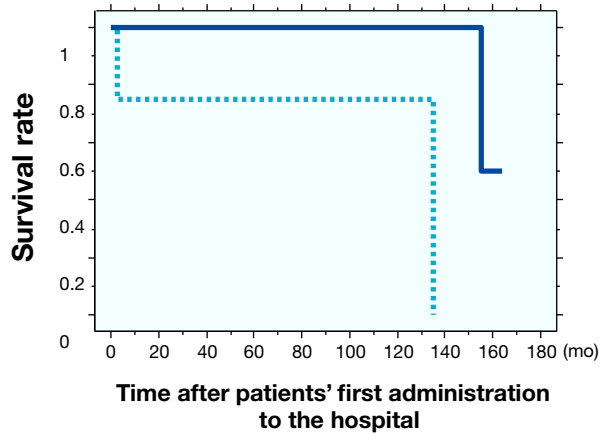

Fig. 1. Overall survival of 6 patients with well-differentiated RL (solid line) is significantly better than that of 4 patients with dedifferentiated RL (dotted line, $P = 0.027$). RL, retroperitoneal liposarcoma.

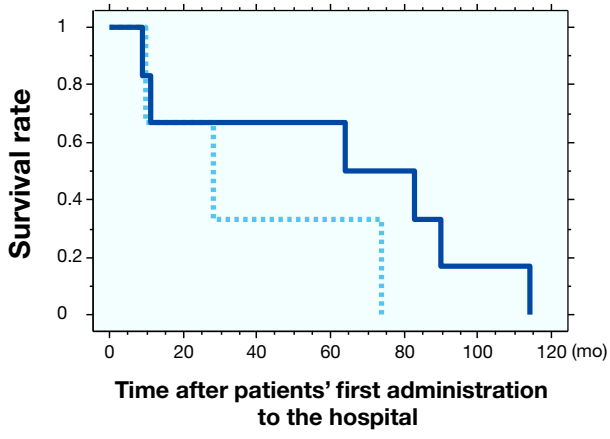

Fig. 2. Relapse free survival of 6 patients with well-differentiated RL (solid line) is better than that of 3 patients with dedifferentiated type (dotted line), but the difference was not significant ($P = 0.27$). RL, retroperitoneal liposarcoma.

ing amount in Patient 2 was 2,590 mL. In each of the 2 patients, even though a huge tumor involved important organs such as ureter and external iliac artery and vein, we could perform careful and prudent operation which led complete tumor extraction with preserving important organs.

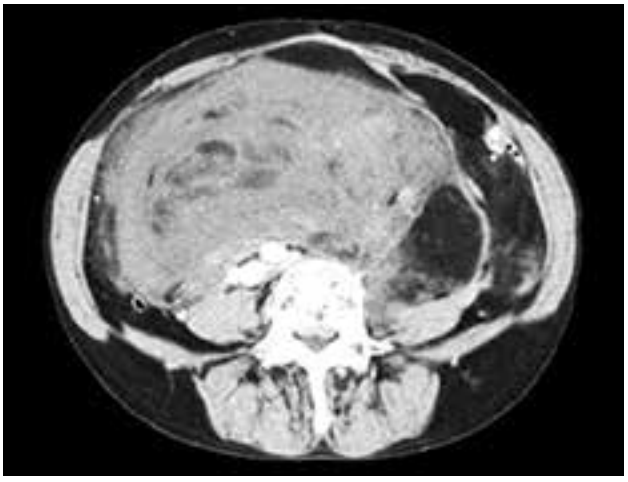

Fig. 3. Abdominal enhanced computed tomography indicates dislocation of the left ureter and the left external iliac artery due to tumor invasion.

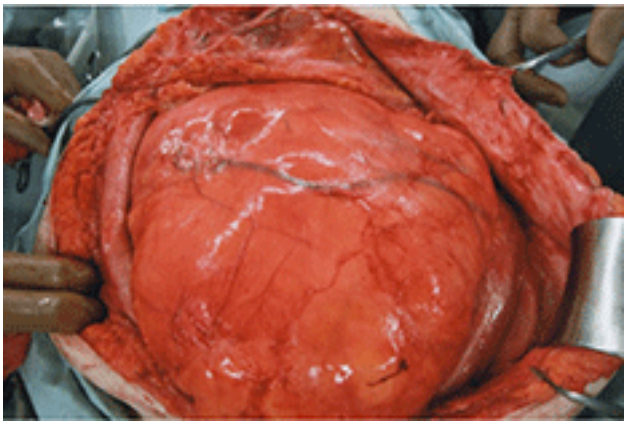

Fig. 4. The huge tumor occupies the whole abdominal cavity.

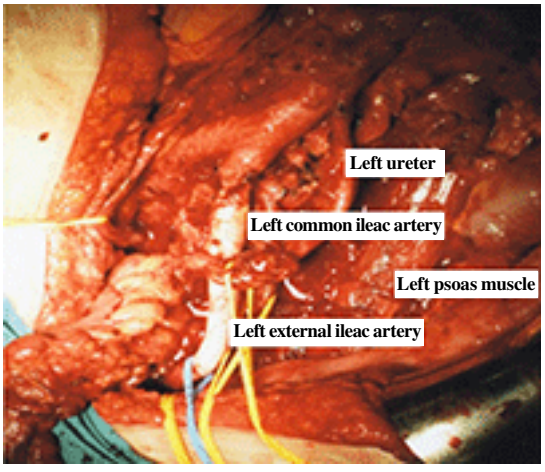

Fig. 5. Complete resection of tumor with preserving important organs was performed.

DISCUSSION

Since RL has no characteristic symptom, patients often enter hospital with a very huge RL. Complete resection of RL with a surrounding margin of normal tissue is the only way to cure the patients, because the effectiveness of radiotherapy and chemotherapy for RL has not been established. A few retrospective studies reported that patients who had received adjuvant radiotherapy had a significant improvement in local failure free survival compared to patients who underwent surgery alone.[13, 14] On the other hand, Ballo et al.[15] found no benefits from radiotherapy for RL. Moreover, they emphasized that a significant amount of toxicities of radiotherapy causes neuropathy, hydronephrosis, ureteral fistula and bowel obstruction. Thus, we have to consider that radiotherapy to the retroperitoneum is challenging because of the large field size and presence of important visceral structures like the kidney, liver, bowels and spinal cord. Also, few retrospective and prospective studies indicated that neoadjuvant or adjuvant chemotherapy showed the survival benefit of patients with RL. Moreover, they concluded that chemotherapy worsened the performance status of the patients.[16, 17]

To achieve a negative margin depends on the relation of tumor to major vascular structures and to invasion of adjacent visceral organs. Complete resection rates in several series for primary RL vary from 43% to 95%.[18–21] The rate will be worse in patients with recurrent huge RL. In our series, the complete resection rate of primary RL was 78%. To achieve a negative margin, Kumar et al.[22] recommended combined resection of one or more organs abutting the tumor such as kidney, colon, adrenals, spleen and pancreas rather than peeling off the organ from the tumor. But we know that many RL patients who underwent combined resection suffer from conditions lacking important organs. Moreover, huge RL often involves inferior vena cava, abdominal aorta, common ileac artery or vein, or bilateral ureters. For such patients, peeling off technique should be recommended to preserve the important organs.

The well-differentiated liposarcomas grow slowly, but dedifferentiated sarcomas grow faster and have a higher ability to metastasize than well-differentiated liposarcomas. We found that well-differentiated RL had better outcomes in terms of recurrence, rate of metastasis and overall survival than dedifferentiated RL. Moreover, considerable attention is paid to the observation that well-differentiated liposarcoma may convert to dedifferentiated liposarcoma after one or more recur-

rences.[23, 24] In the present study, we found that dedifferentiated RL more hardly invaded to neighboring organs (aorta, vena cava or ureter) than well-differentiated RL. So, the peeling off operation was more difficult in dedifferentiated RL.

Local recurrence of RL after surgery is frequent and the rate was reported from 50% to 85%.[25, 26] RL recurs in a huge form, but surgery remains the treatment of choice. Although the chance of a margin-negative resection remains low, adequate resection is associated with an improved survival and hence surgery is preferred for recurrent disease. Also, we can consider adjuvant radiotherapy to improve the relapse free survival of patients with recurrent RL. Radiotherapy to the retroperitoneum is challenging and the radiation dose should be in minimum to avoid the risk of radiation injury.

The authors declare no conflict of interest.

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