Original Article: Clinical Investigation

Demographics, management and treatment outcomes of benign and malignant retroperitoneal tumors in Japan

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Abbreviations & Acronyms
CT = computed tomography
DDLS = dedifferentiated liposarcoma
LS = liposarcoma
MRI = magnetic resonance imaging
OS = overall survival
WDLS = well-differentiated liposarcoma

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*See Appendix.

Objectives: To show the demographics, type of treatment and clinical outcomes of patients with retroperitoneal tumors in Japan.

Methods: We carried out a retrospective analysis of patients with retroperitoneal tumors treated between 2000 and 2012 at 12 university hospitals in Japan. Histology was re-evaluated using the 2013 World Health Organization classification.

Results: A total of 167 patients were included in the analysis. The number of diagnosed patients increased over the 12-year study period. Liposarcoma and schwannoma were the most common histological types among intermediate/malignant and benign tumors, respectively. The intermediate/malignant tumors were larger and were more frequently found in older people. Surgical resection was the primary treatment for 151 patients. The median survival duration for patients with malignant tumors was 91 months, and was significantly shorter than that for patients with benign and intermediate tumors (P < 0.01). R2 resection was associated with significantly shorter survival than R0/R1 resection for malignant tumors (P < 0.01), but not for intermediate. Grossly complete resection of the recurrent tumors improved survival.

Conclusion: The number of patients diagnosed with retroperitoneal tumors increased over time. R2 resection of primary tumors was found to be associated with poor prognosis in malignant tumors, but not in intermediate tumors. Complete surgical resection of recurrent tumors was associated with a better oncological outcome.

Key words: demographics, Japan, retroperitoneal tumor, soft-tissue tumor, treatment outcome.

Introduction

Primary retroperitoneal soft-tissue tumors are rare and heterogeneous in histology and malignant potential. The most common location of soft-tissue sarcomas is the extremities, and retroperitoneal sarcomas account for 9–15% of all adult soft-tissue sarcomas.1–2 Several large studies investigating patients with retroperitoneal tumors in Western countries have been published3–16 but there are only a few studies from Asian countries.17,18 In addition, the majority of the previous works focused only on sarcomas. There is a paucity of data on clinical features, management, and treatment outcomes of patients with benign and malignant retroperitoneal tumors in Asia. We collected data on retroperitoneal tumors from 12 university hospitals, and clarified the trend of incidence, demographics, tumor characteristics, management and prognoses of patients with retroperitoneal tumors in Japan.

Methods

Patients

The present multi-institutional study was carried out with the approval of all the institutional review boards involved.
We carried out a retrospective analysis of patients newly diagnosed with primary retroperitoneal tumors between January 2000 and December 2012 from 12 university hospitals in Japan. A tumor was considered primary when it had not been previously treated. All tumors arising from the retroperitoneal space were included, except the following: all cancers (primary or metastatic), lymphomas, inflammatory tumors, and tumors arising from the gut or its mesentery, kidney, adrenal, prostate, ovary and uterus. Patients whose pathological specimens were unavailable for central pathological examination were also excluded from the analysis.

Pathological assessment
A single pathologist with expertise in the field of soft-tissue tumors re-evaluated the available pathological specimens according to the 4th edition of the World Health Organization classification, and the biological potential of tumors was categorized into benign, intermediate (locally aggressive, rarely metastasizing) and malignant. Pathological diagnoses were made based on hematoxylin–eosin-stained specimens. Resection margins were categorized as either macroscopically complete resection (R0/R1) or gross residual tumor (R2); R0 and R1 were not differentiated because not all tumors were available for microscopic evaluation of margin status.

Statistical analysis
OS was defined as the time from surgery to death from any cause or to the last follow up. In patients who had not undergone surgery, OS duration was calculated from the date of commencement of the first-line treatment. The time to disease-specific death was defined as the time from surgery or from commencement of the first-line treatment to death caused by the disease or to the last follow up. Local recurrence and distal recurrence were defined as recurrence at or near the location of the primary tumor and recurrence at a location distant from the primary site, respectively. Intergroup differences were analyzed using the Mann–Whitney U-test, ANOVA with Tukey’s test and χ²-test. Survival curves were generated using the Kaplan–Meier method and compared using the log–rank test. Statistical software StatMate (GraphPad Software, Inc., La Jolla, CA, USA) and SPSS version 20 (IBM, Armonk, NY, USA) were used for analysis. P-values of <0.05 were considered significant.

Results
Patients and tumor characteristics
In total, 204 patients were diagnosed with retroperitoneal tumors during the study period; 37 patients did not meet all inclusion criteria and were excluded, leaving 167 patients eligible for analyses (Fig. 1). Initial patient characteristics are shown in Table 1.

In total, 71, 49, and 47 patients had benign, intermediate and malignant tumors, respectively. The most common histological type was LS, followed by schwannoma, paraganglioma and leiomyosarcoma. LSs comprised 57.3% of intermediate and malignant tumors. Among the benign tumors, schwannoma was the most common histological type; it was found in 36.6% of patients (26/71). Tumors measuring <5, 5–14.9, and ≥15 cm were found in 32, 93 and 29 patients, respectively. The sizes of intermediate/malignant tumors ranged from 2.2 to 34 cm (median 10 cm), and were significantly larger (P < 0.001) than those of benign tumors, ranging from 2 to 18.7 cm (median 6 cm). In particular, LSs were significantly larger than other tumors (P < 0.05; Fig. 2).

The proportion of intermediate and malignant tumors significantly increased according to age, with 66.7% (28/42) found in patients aged ≥70 years, and 35.3% (6/17) in patients aged <30 years (P = 0.023; Fig. 3).

The number of patients, symptoms, histology, and tumor sizes were compared between the first (January 2000–June 2006) and second halves (July 2006–December 2012) of the study period. The number of patients diagnosed with retroperitoneal tumors increased over time (Fig. 4), with 67 and 100 patients diagnosed in the first and second halves of the study period, respectively. Tumor size was not significantly different between the first and second halves of the study period. The proportion of symptomatic patients slightly decreased (61.2% vs 53.0%) in the second half period, although the difference did not reach the statistical difference. The proportion of intermediate and malignant tumors increased in the second half of the study period (P = 0.048; Table 2).

Treatment
A total of 151 (90.4%) patients underwent surgical resection. Resection margins were R0/R1 in 134 (88.7%) patients, whereas the rest (n = 17 patients) had gross residual tumors (R2). A total of 16 patients did not undergo surgery because of non-resectable tumors or serious comorbidities. Of them, six patients received chemotherapy as primary treatment, and 10 received only the best supportive care.

Survival
OS and disease-specific survival data are shown in Figure 5. With a median follow-up duration of 25 months (range 1–223 months), 4, 7, and 19 patients with benign, intermediate and malignant tumors, respectively, died. All four patients with benign tumors died of other causes. Of seven patients with intermediate tumors, five died of tumors and the other two died of other causes. Of 19 patients with malignant tumors, 18 died of tumors and another patient died of other cause. The median OS for patients with malignant tumors was 91 months, and that for those with benign and intermediate tumors was not reached. The 5-year OS rates for patients with benign, intermediate, and
malignant tumors were 90.4%, 91.6% and 56.8%, respectively. Patients with malignant tumors had significantly shorter OS than those with benign and intermediate tumors ($P < 0.01$); however, OS between patients with benign and intermediate tumors was not significantly different ($P = 0.262$). The median disease-specific survival duration for patients with malignant tumors was 91 months. The 5-year disease-specific survival rates were 100%, 91.6%, and 56.8% for patients with benign, intermediate and malignant tumors, respectively.

### Table 1 Demographics of the 167 patients

| Variable                        | Value (% or range) |
|---------------------------------|--------------------|
| Sex                             |                    |
| Male                            | 74 (44.3%)         |
| Female                          | 93 (55.7%)         |
| Median age at diagnosis, years (range) | 59 (6–84)         |
| Retroperitoneal location        |                    |
| Abdomen                         | 147 (88.0%)        |
| Pelvis                          | 20 (12.0%)         |
| Tumor size (cm)                 |                    |
| <5                              | 32 (19.2%)         |
| 5–15                            | 93 (55.7%)         |
| >15                             | 29 (17.4%)         |
| Unknown                         | 13 (7.8%)          |
| Symptom                         |                    |
| None                            | 73 (43.7%)         |
| Abdominal pain                  | 26 (15.6%)         |
| Palpable mass                   | 17 (10.2%)         |
| Others                          | 49 (29.3%)         |
| Unknown                         | 2 (1.2%)           |
| Biological potential            |                    |
| Benign                          | 71 (42.5%)         |
| Intermediate                    | 49 (29.3%)         |
| Malignant                       | 47 (28.1%)         |
| Histology                       |                    |
| DDLS/myxoid LS                  | 29 (17.4%)         |
| WDLS                            | 26 (15.6%)         |
| Schwannoma                      | 26 (15.6%)         |
| Paraganglioma                   | 11 (6.6%)          |
| Leiomyosarcoma                  | 10 (6.0%)          |
| Ganglioneuroma                  | 10 (6.0%)          |
| Solitary fibrous tumor          | 8 (4.8%)           |
| Mature teratoma                 | 4 (2.4%)           |
| Others                          | 43                 |
| Benign                          | 24 (14.4%)         |
| Intermediate/malignant          | 19 (11.4%)         |
| Multifocality                   |                    |
| Solitary                        | 157 (94.0%)        |
| Multiple                        | 10 (6.0%)          |
| Lymph node metastases†          |                    |
| Yes                             | 4 (4.2%)           |
| No                              | 91 (94.8%)         |
| Unknown                         | 1 (1.0%)           |
| Visceral metastases†            |                    |
| Yes                             | 13 (13.5%)         |
| No                              | 83 (86.5%)         |
| Primary treatment               |                    |
| Surgical resection              | 151 (90.4%)        |
| Chemotherapy                    | 6 (3.6%)           |
| Best supportive care            | 10 (6.0%)          |
| Resection margins               |                    |
| R0–1                            | 134 (88.7%)        |
| R2                              | 17 (11.3%)         |
| No. organs resected             |                    |
| 0                               | 89 (58.9%)         |
| 1                               | 35 (23.2%)         |
| >1                              | 27 (17.9%)         |

† In intermediate and malignant tumors.

### Table 2 Tumor size, symptom, and biological potential in the first and second halves

| Period | First half          | Second half         | P      |
|--------|---------------------|---------------------|--------|
|        | Median size, cm (range) | 8.6 (2–20)            | 7.9 (2.2–3.4) | 0.622 |
| Symptoms (yes/no) | 41/26               | 53/47               | 0.295 |
| Benign/Intermediate and malignant | 32/35               | 39/61               | 0.048 |

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Disease-specific survivals were significantly different among patients with benign, intermediate and malignant tumors (benign vs intermediate, \(P = 0.008\); intermediate vs malignant, \(P = 0.003\)).

Figure 6 shows OS based on the resection margin status. R0/R1 resection was achieved in 73.5% (36/49) and 72.7% (32/44) of patients with intermediate and malignant tumors, respectively. In patients with intermediate tumors (Fig. 6a), the 5-year OS rates for R0/R1 and for R2 resections were both 100%. Median OS was not significantly different between R0/R1 and R2 resections (\(P = 0.372\)). Median OS for patients who did not undergo surgery was 26 months, and the survival of these patients was significantly worse than that of those who underwent surgery (no surgery vs R0/R1, \(P < 0.001\); vs R2, \(P = 0.014\)). In patients with malignant tumors, the median OS duration and 5-year survival rate were 91 months and 65% for R0/R1 resection, and 18 months and 0% for R2 resection, respectively. Survival of patients with R2 resection was significantly worse than that of those with R0/R1 resection (\(P = 0.0006\)), and was not significantly different from patients who did not undergo surgery (\(P = 0.988\)).

**Recurrence**

Follow-up data were missing for seven out of 134 patients who underwent complete resection of their primary tumors (R0/R1); thus, tumor recurrence was evaluated in the remaining 127 patients. Recurrence was observed in three, eight, and 24 patients who had benign, intermediate and malignant tumors, respectively (Table 3). Of these, the most common histological type of primary tumor was DDLS/Myxoid LS, followed by leiomyosarcoma. The initial location of recurrence was local only for 26 patients (74.3%). Six patients developed distant metastasis only (17.1%), and both local and distant were three (8.6%). Of nine patients with distant metastases, five (55.6%) had leiomyosarcomas. Tumor recurrence was detected on follow-up imaging studies in 27 patients (23 using CT and four using MRI) without any recurrence-related symptoms.

The 5-year recurrence-free rates were 89.6%, 92.8%, and 19.3% for benign, intermediate and malignant tumors, respectively. Median recurrence-free durations for intermediate and malignant tumors were 77 and 26 months, respectively (Fig. 7a). The differences in recurrence-free survivals among benign, intermediate and malignant tumors were significant (benign vs intermediate, \(P = 0.018\); intermediate vs malignant, \(P < 0.0001\)). Three patients with benign tumors did not receive any treatment for tumor recurrence. Of the 32 patients with recurrent intermediate and malignant tumors, treatment and follow-up data were available for 25 patients. Of them, 15 underwent surgery for recurrent tumors; 12 and three patients achieved R0/R1 and R2 resections, respectively. Six patients received chemotherapy with ifosfamide, doxorubicin,
vincristine or methotrexate. The remaining four patients received only the best supportive care after recurrence. With the median follow-up period of 27 months (range 2–177 months) after treatment for recurrence, 5-year OS rates were 80.0% and 45.9% for intermediate and malignant tumors, respectively. The median OS was 60 months for primary malignant tumors and not reached for primary intermediate tumors (Fig. 7b). The 5-year survival rate for patients with R0/R1 resection for recurrence of intermediate and malignant tumors was 68.1%, and the median OS was not reached. The 5-year survival rate and median OS for the patient group of R2 resection and no surgery (chemotherapy and best supportive care) were 38.5% and 31 months, respectively, and OS for this group was significantly worse than that for patients with R0/1 resection \( (P = 0.035; \text{Fig. } 7c) \).

**Discussion**

To the best of our knowledge, this is the first study outlining the demographics, treatments and prognosis of retroperitoneal tumors in Japan. In contrast to most of the previous studies on soft-tissue tumors, the present study included benign tumors. Benign tumors comprised 42.2% of all studied retroperitoneal tumors, and were more common than intermediate or malignant tumors. Schwannoma is usually diagnosed in patients aged 21–40 years, and it was the most common benign tumor in our cohort.20 Benign tumors were more frequently found in the younger population, and 64.7% of our patients aged younger than 30 years had benign tumors. LS was the most common histological type, and comprised 57.7% of intermediate and malignant tumors, similar to that reported in other studies.3–18,21 Leiomyosarcoma comprised 10.4% of intermediate and malignant tumors, and was slightly less common compared with that reported in other

| Table 3 | Recurrent tumors |
|---------|-----------------|
| Variable | Value [%] |
| Histology |  |
| DDLS/myxoid LS | 16 (45.7) |
| Leiomyosarcoma | 8 (22.9) |
| WDLs | 7 (20.0) |
| Paraganglioma | 1 (2.9) |
| Benign tumor | 3 (8.6) |
| Symptoms |  |
| Yes | 8 (22.9) |
| No | 27 (77.1) |
| Recurrence site |  |
| Local only | 26 (74.3) |
| Distant only | 6 (17.1) |
| Local and distant | 3 (8.6) |
| Primary treatment |  |
| No treatment | 3 (8.6)† |
| Surgical resection | 15 (42.9) |
| R0–1 | 12 |
| R2 | 3 |
| Chemotherapy | 6 (17.1) |
| Best supportive care | 4 (11.4) |
| Missing | 7 (20) |

Total n = 35. †All three had benign tumors.
studies, where leiomyosarcomas comprised 16–40% of retroperitoneal sarcomas.3–18,21

The incidence of retroperitoneal tumors increased over the duration of our study period, particularly from 2007 to 2012. The development of diagnostic technology including imaging studies and sensitive tumor markers enable the detection of asymptomatic and small-sized tumors. The proportion of asymptomatic patients slightly increased in the second half of the study period. Although no useful tumor markers for retroperitoneal tumors have been identified, the development and prevalence of imaging studies might be helpful to detect incidental tumors. The proportion of benign tumors decreased in the second half of the study period. Although the reasons are unknown, recent advances of imaging studies, such as MRI or positron emission tomography, might be associated with this trend. When tumors are diagnosed as benign by MRI or positron emission tomography, patients might not undergo surgery or biopsy. These patients were excluded from the present study for lack of pathological diagnosis.

Tumor histology, grade, size and treatment are strong determinants of local recurrence and survival in patients with retroperitoneal tumors.5–16,18,21 Patients with malignant tumors had significantly worse overall, disease-specific and recurrence-free survival rates than those with benign and intermediate tumors. Patients with intermediate tumors had significantly lower disease-specific and recurrence-free survival rates than those with benign tumors, but the OS rates were not significantly different between the two. Therefore, the new World Health Organization classification was well correlated to biological tumor potential, and is useful in predicting the prognosis of retroperitoneal tumors.

Surgical resection is still the gold standard of treatment. In our cohort, nearly 90% of patients underwent surgery, and treatment outcomes were comparable with the previous studies.10–14 It is anticipated that complete resection, including resection of contiguous organs when necessary, can improve oncological outcome. Although the superiority of R0 to R1 in oncological outcome remains to be determined,5,13,15 most studies showed that a grossly positive margin (R2) was a strong indicator of unfavorable clinical outcomes, in terms of recurrence and survival, compared with the macroscopic clearance of resection margin (R0/R1).5–11,13–16,18,21 These studies analyzed the prognostic importance of margin status without considering the malignant potential of tumors. In the present study, overall and disease-specific survival rates were significantly poorer after R2 resection for malignant tumors than after R0/R1, and were similar to those for patients who had not undergone surgery. In intermediate tumors, however, survival for R2 resection was identical to that for R0/R1. Therefore, surgery should be carefully considered for patients with large malignant tumors when complete resection is difficult to achieve. In intermediate tumors, however, debulking and cytoreductive surgeries might be indicated for patients with large tumors that are difficult to resect completely.

Chemotherapy and radiation therapy were rarely carried out as a primary treatment. Nussbaum et al. suggested the potential efficacy of radiation therapy for retroperitoneal sarcomas in a matched case–control analysis.22 Ongoing (NCT01344018) and future clinical trials might prove the survival benefit of radiation therapy.

Local recurrence was more frequent than distant metastasis as the initial recurrence in retroperitoneal tumors. The predominant histological types in local and distant recurrences were LS and leiomyosarcoma, respectively. In a large-scale study, 80% of local recurrences were LS, and 53% of distant metastases were leiomyosarcoma.23 During follow up after tumor resection, we need to consider these differences in recurrence sites by histological types.

OS for patients with R0/R1 resection of recurrent tumors was significantly better than that for patients who did not undergo surgery. Complete resection of a recurrent tumor is a powerful predictor of outcome after local recurrence.5,6,8,11,13,14,23 To increase tumor resectability and improvement of prognosis, early detection of recurrence is necessary. In the present study, follow-up imaging studies helped detect asymptomatic tumor recurrence in 77% of the patients. Patients with small and early recurrent tumors are generally asymptomatic. Therefore, follow-up imaging studies are mandatory, although the optimal imaging modality and study intervals are yet to be determined.

The present study had several limitations. First, this was a retrospective and multi-institutional study, and results are based on a relatively small number of patients. To minimize the possibility of interinstitutional bias, we selected 12 university hospitals in Japan, where diagnostic methods and treatment strategies are almost similar. Furthermore, pathological examinations were carried out using a central review system. Second, long-term data were not mature for patients who had undergone surgical resection for low malignant potential tumors, which require long-term follow up for recurrence and progression. Recently, an international collaboration to obtain a consensus on the management of adult retroperitoneal sarcoma has been established.24 The accumulation of data from large-scale, prospective studies can provide the optimal management strategies for rare and heterogeneous retroperitoneal tumors.

In conclusion, the present study is the first to show the demographics and prognosis of patients with benign and malignant retroperitoneal tumors in Japan, and the study data might be generalizable for Japanese as well as other Asian patients. The number of diagnosed patients increased with time. The World Health Organization classification of histology was useful in predicting the prognosis. R2 resection of primary tumors was associated with poor prognosis in malignant tumors, but not in intermediate tumors. En bloc surgical resection of recurrent tumors was associated with a better oncological outcome, and postoperative follow up is mandatory to detect recurrences at a resectable stage.

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Conflict of interest

None declared.
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Appendix

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Editorial Comment

Editorial Comment from Dr Taguchi to Demographics, management and treatment outcomes of benign and malignant retroperitoneal tumors in Japan

Retroperitoneal soft tissue sarcomas are rare mesenchymal tumors that account for approximately 0.15% of all malignancies and comprise a wide variety of histological types.1 Because of their extreme rarity, studies on retroperitoneal tumors are generally lacking. Although some large-scale studies have been carried out in Western countries, few have been carried out in Asian countries.2 The present study by Fujimoto et al. is valuable in that it describes intimate data on the demographics, treatments and prognoses of retroperitoneal tumors, using a multi-institutional cohort of 167 Japanese patients.3 The study included both intermediate/malignant and benign tumors, which might compromise the purity of the study cohort, but could reflect the real-world situation instead.

Interestingly, the authors reported that the incidence of retroperitoneal tumors increased over time, and speculated

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