Original article

Retroperitoneal soft tissue Sarcomas: retrospective cohort study

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Abstract:

Objective*:** Soft Tissue Sarcomas are rare mesenchymal tumors with many subtypes. Clean margin wide resection is recommended for treatment.In this study, the location, histopathological features, clinical and demographic features, recurrence and prognosis of retroperitoneal sarcomas were investigated. ***Methods: The demographic, histopathological and immunohistochemical data of 18 patients who were operated on with the diagnosis of retroperitoneal mass between March 2016 and June 2021 were evaluated retrospectively. ***Results:*** 18 patients were included in the study. 10 patients were male and 8 patients were female. The mean age was 57 (23-81), the median age was 55. While 14 patients were primary sarcoma, 4 patients were recurrent sarcoma. The most common complaint was abdominal pain with 38,87%. The average follow-up time was 26 (0-55) months. The postoperative mean hospitalstay was 5,83 (2-8) days. The average size of the tumor was 19,81 (6,5-36) cm. A total of 8 different histopathological sarcoma types were detected. The most common histology was Dedifferentiated Liposarcoma (44,45%). R0 resection in 10 patients, R1 resection in 6 patients and R2 resection in 2 patient were performed. Organ resection was performed in 6 patients due to organ invasion. During follow-up, 10 patients had a local recurrence and underwent reresection and 6 patients died. The 30-day mortality number was 0. The mean time to detect relapse was 9,3 (1-55) months. Metastasis developed in 2 (11, 11%) patients. ***Conclusion:***Although we are a low-volume hospital for retroperitoneal sarcomas, our results are similar to those in the literature.

Keywords: Soft tissue sarcoma;Retroperitoneal Tumor; Surgery; Multidisciplinary Sarcoma Team

Bangladesh Journal of Medical Science Vol. 21 No. 04 October'22 Page : 829-835
DOI: https://doi.org/10.3329/bjms.v21i4.60258

Introduction

Cancer is the one of the most important cause of mortality all around the world [1]. Soft tissue sarcomas (STS) are rare cancers of mesenchymal origin and can occur in different anatomical locations[2]. STS are most common in the extremities[2,3]. Origin tissue and anatomical region are important in classification [2,4]. There are many histological subtypes. The most common histopathological subtypes are liposarcoma

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and leiomyosarcoma[4,5]. The World Health Organization (WHO) classifies soft tissue tumors into 12 subtypes according to their origin in the 2020 soft tissue sarcoma classification[6,7]. Intra-abdominal and retroperitoneal sarcomas cause nonspecific symptoms, so they are detected as large mass lesions at the time of diagnosis[8,9]. Diagnosis is usually made by CT, MRI or USG. Some studies report a low risk of needle tract metastasis in the biopsy[10].

The anatomical region where STS develops is important in terms of keeping the surgical clean border during resection. Although there are different treatment modalities, the most effective treatment in suitable cases is surgical resection with wide margins[11,12,13]. In vessel or organ invasion may prevent extensive resection. For this reason, other treatment options such as chemotherapy and radiotherapy come to the fore[11,12].

FLNC (Fédération Nationale des Centres de Lutte Contre le Cancer) system is used in STS grading and TNM (tumor, lymph node, metastasis) system is used in staging[14,15]. Today, in evaluating the prognosis of STS, age, size, histological subtype, grade and surgical margin positivity are used as evaluation criteria[16,17]. It has been defined in different markers to determine the prognosis.CD34, S100 protein, SMA, desmin, myogenin, MDM2, CDK4, STAT6, ALK, CD99, H3K27me3, NKX2.2, TLE1, melanocytic markers cyclin D1, cytokeratin, SOX10 and EMA etc. are useful immunohistochemical markers for sarcoma classification[18-20]. Ki-67 protein is used as an important immunohistochemical marker to determine the proliferation level of the tumor cell[21].

In this study, the location, histopathological features, clinical and demographic characteristics, recurrence and prognosis of intraabdominal-retroperitoneal sarcomas were investigated.

Material And Method:

The demographic, histopathological and immunohistochemical data of the patients who were operated on with the diagnosis of intraabdominal mass from March 2016 and June 2021 were retrospectively evaluated. This clinical trial wasapproved by our hospitals Ethical Committe.

Patients diagnosed with pathological sarcoma subclass were included in the study. Patients younger than 18 years were excluded from the study. Visceral organ-origin sarcomas and extremity sarcomas were also excluded from the study. GIST's were included in sarcoma subtypes by WHO in 2013. However, they were not included in the study due to their solid organ origin.

The data of 18 patients' whore remained after the exclusion criteria were analyzed retrospectively. Pathology samples of the patients were re-examined by a single pathologist.

The demographic analysis was used for statistics. This study has been reported in line with the STROCSS criteria [22].

Ethical Approval

Require ethical permissions had been taken from ethical committee of Istanbul Medeniyet University Goztepe Prof. Dr. SuleymanYalcin City Hospital.

Results

A total of 18 patients were included in the study. While 14 patients were primary sarcoma, 4 patients were recurrent sarcoma. 4 patients who were operated for recurrent sarcoma had their previous surgeries performed in different hospitals. 10 patients were male and 8 patients were female. The mean age was 57 (23-81), the median age was 55. The mean follow-up time was 26 (0-55) months. Postoperative mean hospitalization was 5,83 (2-8) days. The mean size of the tumor was 19,81 (6,5-36) cm. The most common complaint was abdominal pain with %38,87. Application complaints are nonspecific and given in the table1. A total of 8 different histopathological sarcoma types were detected. The most common histology was Dedifferentiated Liposarcoma with a rate of 44,45%. Histopathological details are given in table 2. R0 resection in 10 patients, R1 resection in 6 patients, and R2 resection in 2 patients were performed. The organ resection was performed in 8 patients due to organ invasion. Surgical site infection developed in one patient during follow-up was treated with an oral antibiotic. Small bowel resection anastomosis was performed in 1 patient due to enteroatmospheric fistula. 1 patient was treated with percutaneous drainage because of intra-abdominal abscess and empyema in the thorax. Local recurrence was observed in 10 patients and death 6 patients during follow-up. The mean recurrence detection time was 9,3 (1-55) mounts. 8 of 10 patients who developed recurrence were reoperated. Of the 8 patients who reoperated, 2 recurrences developed and were reoperated. One of these two patients again relapsed and were reoperated. Metastasis developed in 2 (11,11%) patients. Thirty-day mortality was 0.

Discussion

Although retroperitoneal soft tissue sarcomas can be observed at any age in the literature, most of the cases are observed in middle and advanced ages. Also, there is no difference between genders in terms of frequency. The median age in the study was 59, and patients were equally distributed in both sexes. The aetiology of STS development is not fully known. However, exposure to certain chemical agents increases the risk of developing soft tissue sarcoma. For example, phenoxy acetic acid derivatives are chlorophenols, thorotrast, vinyl chloride and arsenic. No exposure to chemicals was detected in the study group. Also, due to acute lymphocytic leukaemia in childhood, the risk of developing sarcoma has increased in later ages who received chemotherapy containing alkylating agents (cyclophosphamide, melphalan, procarbazine, nitrosourea, chlorambucil)[23].

All patients in the study presented with nonspecific complaints. Abdominal pain, back pain, constipation, urinary symptoms are some of the complaints of retroperitoneal sarcoma patients. Patients with retroperitoneal soft tissue sarcoma are generally asymptomatic[2,11,13,24]. Therefore, palpable mass is palpated in many patients at the time of diagnosis. There was only 1 patient with a mass of less than 5 cm in the study. This mass was detected as a recurrence during control. After the tumor reaches a large size, specific symptoms due to compression or invasion of surrounding structures begin to develop. Symptoms often develop due to compression or invasion of the colon, ileum and ureter[2,8,13]. In this study, the symptom and size of the STS were similar to the literature.

Treatment in STS should have a multi-disciplinary approach. According to the general opinion, they should be treated in specialized centres. When patients are treated with a multidisciplinary team in reference centres, compliance with treatment guidelines is increased[25,26]. In the study of Randall et al. Evaluating the errors in the diagnosis of soft tissue sarcoma in 104 patients treated in a non-specialized centre, they found that 37% of 104 histological diagnoses were wrong[27]. Our clinic also examines sarcoma cases with a multidisciplinary team (MDT).

The primary treatment for retroperitoneal soft tissue sarcomas is surgery. The most important factor in the development of recurrence in the long term is incomplete surgical resections. Despite the use of adjuvant-neoadjuvant therapies, the most important

Table 1: Clinical Features of Cohort

		N	%
Sex	Male Female	10 8	55,55 44,45
Age		57(23-81)	
Disease Resected	Primary Recurrent	14 4	77,78 22,22
Complaint	No Abdomınal Pain Hematuria Back Pain Palpable Mass Weakness Routine Visit	3 7 1 1 4 1 1	16,67 38,87 5,56 5,56 22,22 5,56 5,56
Recurrence		10	55,55
Reoperation	Total 1 2 3	8 6 1 1	44,45 33,33 5,56 5,56
Metastasis	Total Lung Liver	2 2 1	11,11 11,11 5,56
Resected Organ **Resection Procedure**	Total Hemicolectomy Lıver Segment 5-6 Resection Low Anterior Rezeksiyon Nephrectomy Appendectomy Splenectomy Distal Pancreatectomy Diaphragm Resection	8 3 1 1 2 1 2 2 1	44,45 16,67 5,56 5,56 11,11 5,56 11,11 11,11 5,56

factor affecting long-term survival is R0 surgical resections[3,11,13,28]. However, aggressive surgical procedures may be required for R0 resections due to the size of the tumor, invasion of surrounding tissues and organs. In the study of Bonvalot et al. including 347 patients, 75% of the cases required additional organ resection to provide a negative surgical margin. In the study of Bonvalot et al., nephrectomy was performed most frequently, whereas colectomy was performed at the second frequency[28]. In this study, a total of 13 organ resections were performed in 8 patients. Colon, rectum, splenic, appendix, diaphragm, kidney, pancreas and liver were resected organs. The colon was the most frequently resected organ.

Villano et al. in the analysis of the hospital volume-outcome relationship, 13 RPS operation cases per year were determined as the minimum volume threshold, and institutions meeting this criterion were evaluated as high-volume hospitals (HVH). In our hospital, the mean of resection with the diagnosis of RPS during the study period was 3.9 cases per year that did not meet the criteria for HVH[29]. In the study of Gronchi et al. with 1007 patients, the complete resection rate(R0+R1) was 95%[30]. In the study conducted by Strauss et al. with 200 patients, the complete resection rate was 85%[11]. In our study, the complete resection rate was %88.89.

In the study series by Russo et al., nephrectomy was applied to 20% of the patients. When the patient group with a positive microscopic surgical margin was compared with the patient group who underwent nephrectomy, it was found that the 5-year survival significantly increased in the group undergoing aggressive surgery[31]. In a study of 165 patients by Stoeckle et al., it was found that incomplete surgery increased mortality by 2.8 times[32].

In a study by Hassan et al., between 1983 and 1995, 48 (63%) of 76 patients had adjacent organ resection. More than one organ resection was performed in 22 patients[9]. In the study by Patkar et al., 43 (43%) of 100 patients underwent organ resection[33]. In this study, organ resection was performed in 8 (44,45%) patients. But the number of our patients was too low for comparison.

The TARPS study group, in 2017, in a multicenter study of 1007 patients, found a 30-day mortality rate of 1.8% and a reoperation rate of 10.5% in patients with primary RPS[34]. In this study, the reoperation rate was 44,45% and there was no 30-day mortality. But the number of our patients was too low for comparison.

		N	%
Mortality	Totally	6	33,33
	0-30 day	0	0
	Day>30	6	33,33
Complication	Totally	3	16,67
	Fistula	1	5,56
	Surgical Site İnfection	1	5,56
	Retroperitoneal Abscess	1	5,56
	Empyema	1	5,56

Limitation

The fact that the study is retrospective and the small number of patients is a limitation.

Conclusion

Soft tissue sarcomas are seen in less than 1% of all malignant diseases. This study contains the results of a university hospital in Turkey. In this study, statistical data could not be obtained due to the small number of patients. Although we are a low-volume hospital for retroperitoneal sarcomas, our results are similar to those in the literature. MDT improves the results of sarcoma surgery.

Authorship Contributions

Data gathering and idea owner of this study: MSO, OA

Study design: MA, MSO, OA

Data gathering: ML, NG, ANTY

Writing and submitting manuscript: MA, MSO, OE

Editing and approval of final draft: MSO, OA

Conflicts of Interest: There is no conflicts of interest.

Sources of Funding: We have no funding for our research.

Provenance and Peer Review: Not commissioned, externally peer-reviewed

Table 2: Histopathological Features

		N	%
FNCLCC Grade	1	4	22,22
	2	5	27,78
	3	9	50
Histological Subtype	Liposarcoma, Well Differentiated	4	22,22
	Liposarcoma, Dedifferentiated	8	44,45
	Leiomyosarcoma	1	11,11
	Undifferentiated Pleomorphic Sarcoma	1	5,56
	ExtraskeletalChondrosarcoma	1	5,56
	Synovial Sarcoma	1	5,56
	Mixed-type liposarcoma	1	5,56
	Desmoplastic Small Round Cell Tumor	1	5,56
Resection Clearance	R0	10	55,55
	R1	6	33,33
	R2	2	16,67
LymphovascularInvasion	Yes	2	11,11
	No	16	88,89
Necrosis	No	8	44,45
	<%50	5	27,78
	>%50	5	27,78
Mitosis	0-9	13	72,22
	10-19	4	22,22
	>20	1	5,56
KI-67	0-9	8	44,45
	10-49	7	33,33
	>50	3	16,67

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