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Original Article

Retroperitoneal Soft Tissue Sarcoma: An Analysis of Surgical and Adjuvant Chemotherapy

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Abstract

Introduction: Soft tissue sarcomas are a rare and heterogeneous group of tumors that arise from the supporting skeletal tissue.

Aim: To study the epidemiology, various presentation, and the response to the surgery and adjuvant chemotherapy.

Materials and Methods: A prospective observational study on retroperitoneal sarcoma was conducted in the Department of Surgical Oncology.

Results: A total of 21 patients were taken in the study and the age incidence is mainly in the 5th decade as with other study. The major histopathological examination of retroperitoneal sarcoma was liposarcoma followed by leiomyosarcoma.

Conclusion: Most of the tumors occur in the 5th decade, and surgery was the mainstay of treatment. Only primitive neuroectodermal and rhabdomyosarcoma showed good results with chemotherapy.

Key words: Retroperitoneal sarcoma resection of tumor, Soft tissue sarcoma, Tumor grade

INTRODUCTION

Soft tissue sarcomas are the most frequent sarcomas. They are a rare and heterogeneous group of tumors that arise from the supporting extraskeletal tissues.[1] Soft tissue sarcomas are disease of adulthood, occurring most commonly in persons between 30 and 60 years of age. The sole exception is rhabdomyosarcoma, which occurs in young children. Each of the various soft tissue sarcomas has a unique morphology, biological behavior, and characteristics.[2] The clinical, radiographic, and surgical management of most soft tissue sarcomas is identical, regardless of histogenesis. The treatment of soft tissue sarcoma has become multidisciplinary, as advances in biology, imaging, surgery, chemotherapy, and radiotherapy have improved the outlook for these patients who have these malignancies.[3] Fifteen percent of adult soft tissue sarcomas occur in the retroperitoneum. Most retroperitoneal tumors are malignant, and about one-third are soft tissue sarcomas. The most common sarcomas occurring in the retroperitoneum are liposarcomas, malignant fibrous histiocytomas, and leiomyosarcomas. The size at presentation depends on the location. Tumors in the proximal extremities and retroperitoneum are often quite large, whereas distal extremity tumors are often small. The anatomic site of the primary disease represents an important variable that influences treatment and outcome. Soft tissue sarcomas of the extremities account for about 50% of all sarcomas, gastrointestinal (GI) sarcomas for 25%, retroperitoneal sarcomas for 15-20%, and head and neck for 9%.[4]

Aim

The aim of the study was to study the stage of the disease at presentation, incidence of various pathological types, and incidence of the grade of the tumor.

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MATERIALS AND METHODS

This prospective observational study was conducted in the Department of Surgical Oncology, Government

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Royapettah Hospital, Chennai. Patients admitted for retroperitoneal sarcoma were included in the study. The institutional ethics committee approval and informed consent were obtained. Histories such as abdominal mass, its duration, presence of pain and its duration, other symptoms, and family history were recorded. Previous history of surgery, biopsy if any, and treatment were taken. Physical examination was done to note site, size of swelling, and presence or absence of metastases. Chest X-ray, computed tomography (CT) abdomen, and CT chest were taken in all patients. Histopathology, grade, and margin status were noted. Histopathology is compared with previous reports.

RESULTS

Retroperitoneal sarcoma consisted of 0.16% of all cancers admitted in the institute (Table 1).

Retroperitoneal sarcoma forms 9.85% of all soft tissue sarcomas (Table 2).

In this study, incidence of retroperitoneal sarcoma was seen more in males than in females in a ratio of males females 2.5:1 (Table 3).

According to literature,[4] the most common histopathologic types in the retroperitoneum are liposarcoma (40%), leiomyosarcoma (25%), malignant peripheral nerve sheath tumor (MPNST), and fibrosarcoma. Approximately 55% of retroperitoneal liposarcomas are well differentiated and low grade, with tumors in roughly 40% of patients showing dedifferentiated, high-grade histologic features at primary presentation. In this study, the results are the same as that of literature with liposarcoma (52.38%) being the most common histopathology (Table 4).

As per literature, the peak incidence is in the 5th decade of life although they can occur in any age group.[5] The study findings bide with literature results with peak incidence in 5th decade, accounting about 33.33% of the all age groups (Table 5).

Most of the retroperitoneal sarcoma are large at the time of presentation and are considered as deep tumors according to the AJCC staging. All belong to T2b. The average size of the tumor at presentation 15.19 cm. The median size of retroperitoneal sarcoma is 25 cm. As per literature, most of the retroperitoneal sarcoma was seen in Stage III accounting to 62%. Only one case was found to be in Stage IV in this study. At the time of presentation, the majority were primary tumors but few of them were recurrent (Table 6).

Out of the two recurrent cases, one of them was a case of 2nd time recurrence and the other 3rd time recurrence (Table 7).

Table 1: Prevalence of retroperitoneal sarcoma

Cancers	Number (%)
Retroperitoneal sarcoma	21 (0.16)
Other	12990 (99.83)
Total	13011 (100)

Table 2: Distribution of soft tissue sarcoma

Cancers	Number (%)
Retroperitoneal sarcoma	21 (9.85)
Other soft tissue sarcoma	192 (90.14)
Total	213 (100)
Sex ratio	

Table 3: Gender wise distribution

Sex	Frequency (%)
Male	15 (71.42)
Female	6 (28.57)
Total	21 (100)

Table 4: Pathological presentation

Histopathology	Frequency (%)
Liposarcoma	11 (52.38)
Leiomyosarcoma	4 (19)
PNET	3 (14.3)
Rhabdomyosarcoma	1 (4.7)
MPNST	2 (9.5)
Total	21 (100)

MPNST: Malignant peripheral nerve sheath tumour, PNET: Primitive neuroectodermal tumour

Table 5: Age wise distribution

Age groups	Frequency (%)
<20	2 (9.5)
21-30	4 (19)
31-40	1 (4.7)
41-50	7 (33.33)
51-60	3 (14.3)
61-70	3 (14.3)
>70	1 (4.7)
Total	21 (100)

Table 6: Stage wise presentation

Stage	Frequency (%)
IA	-
IB	7 (33.33)
IIA	-
IIB	-
III	13 (62)
IV	1 (4.7)

Management and work up plan of the recurrences were managed like as for primary tumors. Most patients present with an asymptomatic abdominal mass. On occasion, pain is present, and less common symptoms include GI bleeding, incomplete obstruction, and neurologic symptoms related to retroperitoneal invasion or pressure on neurovascular structures. Weight loss is uncommon, and incidental diagnosis is the norm. Some of the patients came with more than one complaint. Abdominal pain was the major complaint most of the patients in this study accounting about 47.6%. Neurologic symptoms were found to be high 42.85% when compared to the literature values 27% (Table 8).

In this study, only one case was incompletely resected and in literature incomplete resection is acceptable only in case of a retroperitoneal sarcoma with well-differentiated liposarcoma as the histopathology. In such cases, the long-term survival is significantly increased, whereas in other cases, incomplete resection has same survival rates as those without surgery. Out of the 8 cases which had complete resection, 3 cases were positive for margins. These cases were subjected to postoperative radiotherapy (Tables 9 and 10).

Table 7: Recurrence presentation

Presentation	Frequency (%)
Primary	19 (90.47)
Recurrence	2 (9.52)
Total	21 (100)

Table 8: Symptoms at presentation

Symptoms	Frequency (%)
Abdominal pain	10 (47.6)
Abdominal mass	6 (28.5)
GI bleed	1 (4.7)
Neurologic	9 (42.85)

GI: Gastrointestinal

Table 9: Management

Treatment	Frequency (%)
Complete resection	8 (35)
Incomplete resection	1 (4.7)
Inoperable	6 (28.5)
Metastasis	1 (4.7)
Chemotherapy without surgery	5 (23.8)

Table 10: Margin of resection

Margin	Frequency (%)
Positive	3 (37.5)
Negative	5 (62.5)
Total	8 (100)

In a study of 28 patients with liposarcomas, adjacent organ resection was carried out in more than half the cases, with partial or total resection of the kidneys in 60%, colon in 50%, and adrenal glands in 35%. Although nephrectomy was performed in 60% of cases, the kidney itself was rarely involved. Nevertheless, the encompassment of the kidney and the involvement of the hilar renal vasculature make the resection of the kidney often necessary.

In this study, 11 cases went in for adjacent organ resection. Nephrectomy was the most common procedure done along with the resection of the tumor. Left kidney removal (55.55%) was more common than right kidney removal (11.11%). Most common bowel loops to be resected was descending colon. In most of the cases, descending colon was completely removed and distal 2/3rd transverse colon was anastomosed to either sigmoid colon or rectum. The majority of cases which are inoperable are due to vascular involvement (19%). Vascular structures commonly involved are inferior vena cava and common iliac veins (Table 11).

In this study, no vascular resection and reconstruction was done (Table 12). Such cases were given postoperative radiotherapy, but overall survival was the same. Radiotherapy helped in local control only in few patients and tumor was progressive in some cases. Out of the 21 cases, 12 cases (57.14%) had previous biopsy done. Pre-operative biopsy is required in case of high suspicion when there is chance of the retroperitoneal tumor being a lymphoma or primitive neuroectodermal tumor (PNET) where such radical resection is not at all required as they are curable by chemotherapy alone. In this study, high-grade tumors accounted for 66.66% (Table 13).

Table 11: Adjacent organs involvement

Adjacent organs removed	Frequency (%)
Kidney	6 (66.66)
Bowel loops	5 (55.55)
Total	11 (100)

Table 12: Vascular resections

Blood vessels	Frequency (%)
SVC	2 (50)
Common iliac vein	2 (50)
Total	4 (100)

SVC: Superior vena cava

Table 13: Grade of the tumour

Grade	Frequency (%)
High	14 (66.66)
Low	7 (33.33)
Total	21 (100)

According to literature, most of the retroperitoneal sarcoma was low-grade, well-differentiated tumors unlike the reports of this study. In this study, 5 cases had adjuvant and neoadjuvant chemotherapy. Especially, PNET, MPNST, and rhabdomyosarcoma responded well to chemotherapy.

DISCUSSION

Retroperitoneal sarcoma forms 10-20% of the soft tissue sarcomas according to Mettlin *et al.*, but in this study, it accounts for 9.85% of soft tissue sarcomas, most of it occurs in males than in females in a ratio of 2.5:1.[5] As with other series, the age incidence is mainly in the 5th decade. The major histopathology of the retroperitoneal sarcoma is liposarcoma followed by leiomyosarcoma.[6] Majority of the cases at the time of presentation were about 15 cm in diameter and most belonged to Stage III (62%). Abdominal pain, discomfort, and neurologic pain were the most common presenting complaints. The majority were primary tumors and only 9.52% being recurrent tumors. About 35% of tumors were completely resectable and 28.5% were inoperable due to involvement or proximity to vascular structures. Only one case of metastasis was reported. Nearly 62.5% of operated cases were margin negative. Most of the tumors were high-grade tumors (66.66%).[7] Most of the resections involved adjacent organ removal with bowel loops and kidney being the common adjacent organs removed. Kidney (66.66%) removal was slightly more common than bowel removal (55.55%). Vascular involvement was seen in 19% of the cases. Only primitive neuroectodermal tumor and rhabdomyosarcoma showed good results with chemotherapy. Radiotherapy had no significant role in controlling local spread as well as on survival benefits.[8-10]

CONCLUSION

Most of the tumor occurs in the 5th decade and surgery was the mainstay of treatment. Only primitive neuroectodermal and rhabdomyosarcoma showed good results with chemotherapy.

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