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Clinical Presentation and Surgical Treatment Outcome of Retroperitoneal Sarcoma: A study in Department of Surgical Oncology, National Institute of Cancer Research Hospital, Mohakhali, Dhaka, Bangladesh

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Abstract

Background: Retroperitoneal sarcoma is a giant tumor usually develops in retroperitoneal space of abdomen which is a mysterious timorous lesion. It has versatile dimension of clinical presentations and surgical outcome. It contains, embedded in a meshwork of loose connective tissue, the adrenal glands, kidneys and ureters, aorta and its branches, inferior vena cava and its tributaries and numerous lymph nodes.[1] Retroperitoneal sarcoma (RPS) is a rare tumor accounting for approximately 10-15 percent of all soft tissue tumors.[2] **Objectives:** Toassess the clinical presentation and surgical outcome of retroperitoneal sarcoma. **Methods:** This prospective observational was conducted in the General Surgery Department in a tertiary care hospital, Dhaka, Bangladesh. The patients were enrolled by purposive sampling. All the patients underwent definitive surgery. A pre formed structured, peer reviewed data collection sheet was prepared which was used to collect data. Data were complied, edited, managed and analyzed by SPSS version 20.0. The result was tabulated and presented kin figure form. Data was done by Pearson's chi square test and student's t test. For significant calculation, p value considered at <0.05. **Result:** Out of 30 patients', maximum 14(46.6%) patients belongs to 51-60 years' age group, which was subsequently followed by 7(23.33%) in >60 years' age group. 5(16.67%), 3(10%) and 1(3.33%) patients belonged to 41-50 years, 31-40years and ≤ 30 years' age group respectively. Out of 30 patients, 25(83.33%) and 5(16.67%) were male and female respectively. The male and female ratio was 5:1. Out of 30 patients 25(83.33%) and 5 (16.67%) were male and female respectively. The male to female ratio was 5:1. Out of 30 patients 22(73.33%) and 8(26.67%) were primary and recurrent retroperitoneal sarcoma respectively. Out of 30 patients all 22 (100%) patients in primary and 8 (100%) patients in recurrent retroperitoneal sarcoma presented with abdominal mass. Butonly11(50%) in primary cases had pain or discomfort in comparison to 3(37.5%) out of 8 in recurrent cases (p=0.4). The median radiological tumor size in primary and recurrent cases were 15cm and 12cm respectively (p=0.003). Out of22 patients and 8 patients in primary and recurrent retroperitoneal sarcomere'spectively;focality and invasiveness showed statistically significant differences as well as number of resected organs /structured (p=0.006, 0.001 and 0.09respectively). On the contrary tumor resection margins, grade, histology and resection of adjacent visceral structures showednostatistically significant differences between the groups (p=>0.05). **Conclusion:** Retroperitoneal sarcoma is a giant abdominal tumor that takes it huge size silently. The surgical outcome of primary retroperitoneal sarcoma is relatively better than recurrent retroperitoneal Sarcoma.

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Keywords: Retroperitoneal Sarcoma, Abdominal Tumor, Surgical Treatment

INTRODUCTION

Soft Tissue Sarcomas (STS) account for 2% of all adult cancers. With an estimated incidence of 59 per millionand per year, approximately 30,000 new cases are diagnosed yearly in Europe, 11,900 in the United States and2000inJapan. Fifteen of all STS are located in the retro-peritoneal space.[3] The incidence of retroperitoneal sarcoma (RPS) is less than 1/100,000people per year and accounts for approximately one-third of all retroperitoneal masses. Currently more than 50 histological types of soft tissues sarcoma have been identified, but the most common adult types are liposarcoma (17%), leiomyosarcoma (16%), pleomorphic sarcoma/malignant fibrous histiocytoma (3.5%), myxofibrosarcoma (3%) and synovial sarcoma (2%).[4]Bothprimary and metastatic tumors in the retroperitoneum grow silently for a considerable period of time beforeclinical signs and symptoms appear. This is due to the availability of potential large space with abundant loose connective tissues and relative paucity of vital structures. In general, the clinical presentation is vague and related to the compression/invasion of neighboring structures and obstructive phenomena.[1]Thediagnosisofretroperitoneal soft tissue tumors is generally based on clinical, radiological and histological features. However, in certain tumors with overlapping histological features and in high grade sarcomas with poor phenotypic differentiation accurate diagnosis is a challenge, where in, immune his to chemistry for specific antigens or cytogenetic analysis to detect tumor specific genetic alterations contribute to the definitive diagnosis. Its clinical presentation is not apparently noticeable to the patient all the time. It constitutes a therapeutic challenge because of relatively late presentation and anatomical location, often close to vital structures in the retroperitoneal space. This close relationship to vital structures impacts on the ability to perform a radical wider section. It is often not possible to obtain a margin of normal tissue around the tumor. Local recurrence is the main causes of failure, ranging from 40 to 80 percent.[5] Seventy-five percent of sarcoma related deaths involve uncontrolled local recurrence.[6] Surgery plays a principal role in the management of retroperitoneal sarcoma (RPS) and provides the only opportunity for cure. No effective vechemo therapy exists influences survival in patients with retroperitoneal sarcoma.[7] Given that local failure remain causes of death after surgery in the patients, there is great interest in strategies that might improve local control. The role of radiotherapy in helping to achieve local control remains undefined with no prospective randomized controlled trials available to define indications, dose, route of administrative or impact on overall survival.[8]An initial attempt by the American College of Surgeon Oncology Group to investigate the role of prospective radiotherapy in retroperitoneal sarcoma in a randomized setting was closed early because of preoperative radiotherapy in retroperitoneal sarcoma in a randomized setting was close dearly because of poor patient accrual.[9]

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OBJECTIVES:

General Objectives:
To assess the clinical statusand surgical treatment outcome of retroperitoneal sarcoma.

Specific Objectives:
To assess the clinical status of retroperitoneal sarcoma
To determine the level of resection in retroperitoneal sarcoma surgery

METHODS AND MATERIALS

Study Design: This was a prospective observational study.
Place of Study: Department of General Surgery in a Tertiary Care Hospital, Dhaka, Bangladesh.
Study Population: All admitted to oncology surgical cases in the General Surgery Department of the Tertiary Care Hospital.
Study Period: From July 2017 to June 2018.
Sampling Methods: An Purposive sampling methods was followed according to the availability of the assigned patients.
Research Instruments: A semi structured questionnaire was prepared for the purpose of data captured. Which included all the variables of interest. The hospital's records also had been analyzed.
Study Procedures: The study was under taken on the patients diagnosed with retroperitoneal sarcoma. Diagnosis was done after proper history taking, imaging and FAC investigation. Patients were selected from the Department of General Surgery of a Tertiary Care Hospital, Dhaka, Bangladesh, which has patient's health records and all the surgical facilities. The clinical course of all patients with loco regional disease (without distant metastasis) at presentation, treated from July 2017 to June 2018 for soft tissue sarcomas of the retro peritoneum at our institute was reviewed both prospectively. The purpose and procedure of the study was discussed with the patients. Written consent was taken from those patients who agreed to participate in the study. The primary RPS was defined as a tumor which is untreated before definitive surgical intervention. Local vs distant recurrent was separated as the sample size is small. Surgical restriction was classified into complete (R_0) or incomplete (R_1 and R_2). The patients were followed up 1 and 3 monthly.

Data Processing: Collecteddata was checked and edited first. Data were than processed by SPSS software version 23.0. All the data were complied, edited, managed and in tabulator and figure form.

DataAnalysis: Statistical analysis was performed using the Statistical Package for Social Sciences (SPSS, Chicago, IL) version23.0 software for Windows. Descriptive statistics was performed, and all data was expressed as mean ± SD andpercentageratio.

Ethical Issues: Ethical clearance for the study was taken from the department of general surgery and the concerned authority NICRH. The entire study population was thoroughly appraised about the nature, purpose and implication of the study, as well as entire spectrum of benefits and risks of the study. There was physical, social and legal risk during collection of blood and physical examination and surgery; proper consent was taken. Interest of study population was being compromised to safeguard their rights and health. For safeguarding confidentiality protecting anonymity each of the patients was given special ID no, which was followed

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in sample collection, transport to lab and reporting, in each and every steps of the procedure. A signed informed consent was taken from the patients convincing that privacy of the patients was maintained and patient would become compensated for loss of work time if he wants. A data sheet (enclosed) was prepared for which a short interview of 10-15 minutes was required. No drug was used for this study. No experimental new drug was administered. No placebo was used here.

RESULTS

Age group in years	Frequency(%)
≤30	1(3.33%)
31 –40	3(10%)
41 –50	5(16.67%)
51 –60	14(46.67%)
>60	7(23.33%)
Median	55
Age range	29-68

Table 1 showed, out of total 30 patients, 14(46.67%) patients belonged to 51-60years age group which was subsequently followed by 7(23.33%) in >60 years' age group. 5(16.67%), 3(10%) and finally 1(3.33%) patients belonged to 41-50 years, 31-40 years and ≤ 30 years' age group respectively.

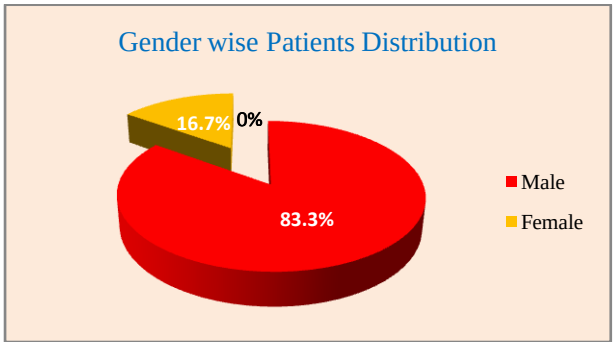

Figure 1: Patients Gender Wise Distribution
Figure I showed, out of 30 patients 25(85%) was male and 5(15%) was female patients.

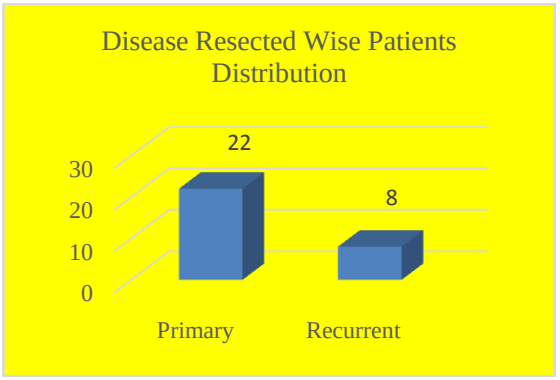

Figure-2: Distribution of patients according to diseaseresected (N=30)
Figure 2: showed, out of 30 patients, 22(73.3%) patients was primary and rest 8(26.7%) was recurrent patients.

Table-2: Clinical status of patients(N=30)

Clinical Status	Primary (n=22)		Recurrent(n =8)		p-Value
Abdominal mass					
Present	22	100 %	8	100%	0.54ns
Absent	0	0%	0	0%	
Pain/discomfort					
Present	11	50%	3	37.5%	0.003s
Absent	11	50%	5	62.5%	
Radiological Tumor Size (in Inch)	15		8		

P value was calculated by chi square test. ns= Not significant, s=Significant, Significant of P vale is <0.05

Table-2 shows that out of 30 patients all 22 (100%) patients in primary and 8(100%) patients in recurrent retroperitoneal sarcoma presented with abdominal mass. But only 11(50%)in primary cases had pain or discomfort in comparison to 3(37.5%) out of 8 in recurrent cases (p=0.4). The median radiological tumor size in primary and recurrent cases were 15cm and 12cm respectively(p=0.003).

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Table -3: Distribution of patients according to pathological status (N=30)

Pathological Status	Primary (n=22)		Recurrent(n=8)		p-Value
Tumor resection margins	n	%	n	%	
R_0	6	27.27%	2	25%	0.98^{ns}
R_1	13	59.09%	5	62.5%	
R_2	3	13.63%	1	4%	
FNCLLCC tumor grade					
G_1	12	54.54%	6	75%	0.45^{ns}
G_2	2	9.09%	1	12.5%	
G_3	8	36.36%	1	12.5%	
Histology					
Well differentiated	11	50%	4	50%	0.88^{ns}
Liposarcoma Dedifferentiated	7	31.81%	2	25%	
Liposarcimal/Leiomysarcoma	3	13.63%	1	12.5%	
Others	1	4.54%	1	12.5%	
Focality					
Unfocal	15	68.18%	1	12.5%	0.006^{s}
Multifocal	7	31.82%	7	87.5%	
Invasive					
Yes	3	13.63%	6	75%	0.001^{s}
No	19	86.36%	2	25%	
Number of resected organs(median)	2		2		
IQR	1-3		1-2		
Resection of adjacent viscera/structures					
Not performed	12	54.54%	2	25%	0.35^{ns}
Performed	10	45.45%	6	75%	

FNCLCC: Federation Nationaledes Centresde Lutte Contrele Cancer

Table 3 showed, out of 22 patients 8 patients in primary and recurrent retroperitoneal sarcoma respectively; focality and invasiveness showed statistically significant differences (p=0.006 & 0.0001). On the contrary tumor resection margins, histology and resection of adjacent visceral structures showed no significantly differences between the groups.

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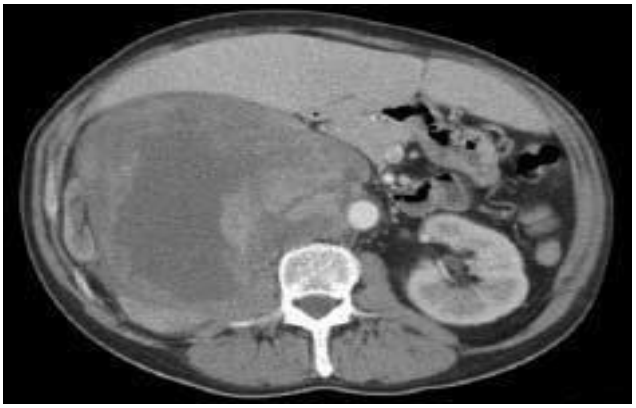

Figure-3: CT scan of a large, high graderetroperitonealleiomyosarcoma with lateral displacement and compression of the right kidney.

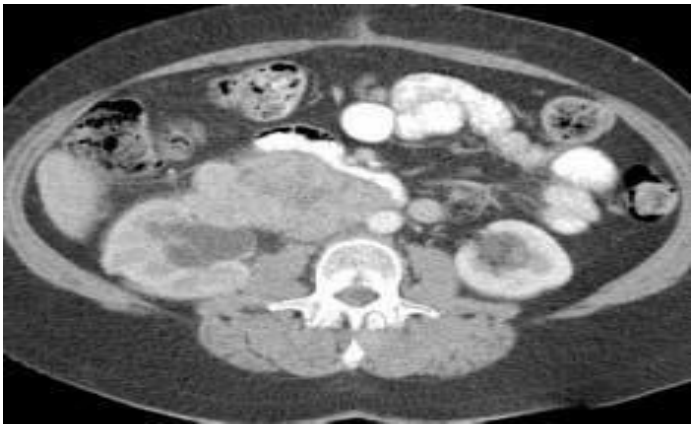

Figure-4:CT scan of a leiomyosarcomaarisingbetweentheinferiorvenacavaandduodenum.

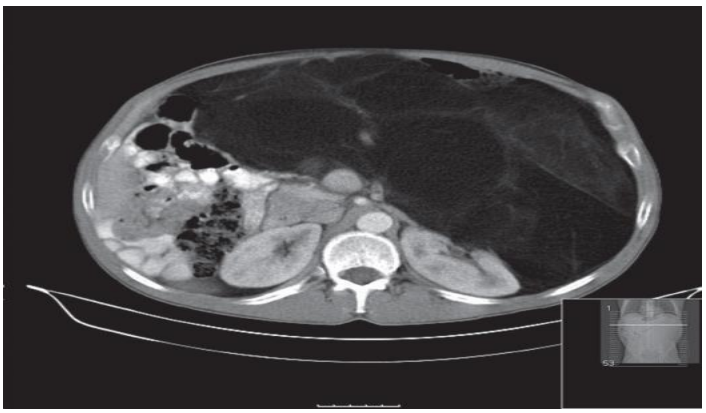

Figure-4: CT of a 42-year-old male demonstrating a huge, well differentiated liposarcoma arising from the leftretroperitoneumandanteriorlytotheleftkidney,andextendingintothepelvis.Thetumorextendssuperiorlytothe left hemidiaphragm, where it passes the midline with displacement of small and large bowel loops into theright flank. The CT attenuation reflects the histological subtype, specifically the amount of fat in the mass, withlow-grade,well differentiatedliposarcoma entirelyor

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predominantlyfatty.

DISCUSSION

RPS usually arise from the connective tissues posterior to the posterior peritoneum and uncommonly from specific retroperitoneal tissues such as the kidney, inferior vena cava, spinal nerve roots or the aorta.[10]RFP often grow silently to a very large size before diagnosis. Patients typically present with chronic non-specific complaints related to tumor compression rather than infiltration,[11] including abdominal distention and pressure, early satiety and anorexia, changes in bowel or bladder habit and peripheral oedema. Not in frequently diagnosis made on an incidentally found asymptomatic mass.[12] The guideline for the treatment of RS recommended complete surgery consisting of the resection of the localized tumor mass with clinically negative excision margins (NCCN; 2012). As histopathological margin status is recognized as being the most important prognostics factor contributing to long-term local disease free survival,[13]aggressive surgeries consisting of the organs and viscera adjacent to the tumor mass although clinically uninvolved has been proposed to improve local tumor control in patients with primary RS.[14] Complete and aggressive surgeries have been directly compared in retrospective series,[14] suggesting possible improvement in tumor control after aggressive surgical specially in patients with low-grade tumors.[15] Several concerns exist regarding the retrospective design of such studies (including limited length of follow-up) as well as a lack of standardization of the aggressive surgical technique, and the absence of prospective studies designed to compare complete and aggressive surgery.[16]We have evaluated 30 patients in department of Surgical Oncology where it was evident that 22(73.33%) cases were primary RPS and rest 8(26.67%) were secondary RPS. Statistically significant difference in clinical presentation (0.003). The similar result was observed in the study by Carlo Ricardo[17] where they showed the median tumor size were 15cm and 12cm respectively. The highest resection margin category in primary RPS was R_1(59.09%) and same (62.5%) in current category (p=0.98)[18] in their study revealed R_0/R_1 as the highest to overall rexction margin category (88.9%). Comparing in our study it was a little lower (86.67%) than study. From the point of view of FNCLCC: tumor grade 12(54.54%) and 6(75%) were the highest the primary and recurrent RPS respectively. Carlo Ricardo Rossi's results agreed with our findings were differentiated iposarcoma was the highest histologically category (50% each) in both the groups. This was also similar like previous study.[18] Focality, invasiveness, and number of rejected organs revealed statistically significant differences between primary and re current RPS groups(p=<0.05) that was also agreed by a previous study.[18] Recent publications have described the invasive behavior of RPS, helping to explain the propensity to local recurrence. Previously, grade RPS was thought to be invasive but Mussiental found invasive behavior in 25% and 33% respectively of the well differentiated liposacoma (WDLS) cases they

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reported.[19]Half (50%) of the tumors resected in our series demonstrated invasive behavior on histopathological examination but the proportion was reduced (35%) when considering WDLS patients only. The difficulty of microscopically examining the surface of a 20 cm tumor completely is noted. In the extensively histologically sampled prospective series. Mussietal described in filtration of at least one organ in 80% of their patients.[19] Wound infection (p=0.77) peritoneal hemorrhage (p=0.67) and septic complications (p=0.43) were the frequent complications through none of them showed significant difference between primary and recurrent RPS category. Out of 22 patients, 13(59.09%) in primary RPS showed uneventful outcome. On the contrary 3(37.5%) out of 8 patients in recurrent RPS revealed uneventful outcome. To the best of our knowledge, this study in Bangladesh regarding clinical status and outcome of RPS. As it was the short term cross sectional study, to evaluate the DFS and OS were beyond the scope the study.

LIMITATION OF THE STUDY:

This was a prospective observation study. The sample size was too small.Thestudyperiodwas small. This study wasasingle centered,singleblindedstudy.

CONCLUSION

Retroperitoneal sarcoma is a giant abdominal tumor that takes it huge size silently. The surgical outcome ofprimary retroperitoneal sarcoma is relatively better than recurrent retroperitoneal sarcoma. A case control studyis recommended. A multi-centered, double blinded study is recommended. A large sample size as well as longtermstudyisadvocated.

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