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Retroperitoneal Soft Tissue Sarcoma: Current Scenario in our Stateregional Cancer Hospital (AHRCC) & Medical College (SCB): A Retrospective Study

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ABSTRACT

Background: Retroperitoneal sarcomas are very rare neoplasms accounting for 1-2% of all solid malignancies &10-20% of all sarcomas. Peak incident is in the 5th decade of life. Liposarcoma & liomyosarcomas are the most common varieties presenting with abdominal pain or fullness. They usually attain a very large size at the time of Presentation. CT & MRI play a major role in the diagnosis & histological grading being the most important prognostic factor. Complete Surgical excision is the only hope of treatment with chance of recurrence. In our study, we focus on various clinical presentations& management of patients attending our medical college (SCB Medical College) & Regional cancer hospital (AHRCC). **Methods:** This retrospective study was based on study of total no of 20 patients getting admitted either to our regional cancer hospital (Acharya harihara cancer hospital) or department of general surgery S.C.B medical college during the period Aug 2014 to Aug 2016 according to the inclusion criteria. Cases were studied according to the age &sex, radiological imaging, staging & histological grading. Resectability was assessed & underwent radical resection whenever possible. Patients were followed up till their last clinical follow up or death. **Results:** In our retrospective study of 20 patients in our regional hospitals showed that most of the patients were male (n=14, 70%) with mean age of 55yrs.Most common organ resected was kidney (3), colon (2), pancreas (1).Almost 90%(n=18) patients underwent complete surgical resection except 10%(n=2) patients undergoing partial resection were due to involvement of major vessels (inferior vena cava & abdominal aorta). Most of the patients presented with high grade tumours (n=10, 50%) and 50% were presented with T4 stages. At the end of follow up 3 patients died of recurrence and another 2 underwent re-excision. **Conclusion:** Complete surgical resection is the goal of treatment which is the most effective treatment for primary or recurrent retroperitoneal sarcoma, but frequently not possible. Complete surgical resection is possible in 40-60% cases. Incomplete surgical excision increases chances of local recurrence. Each recurrence decreases chances of cure. Multiorgan resection often required to attain negative margins. Proper pre-op radiological assessment is very important.

Keywords: Retroperitoneal sarcoma, surgical excision, Recurrence, CT-imaging.

INTRODUCTION

Retroperitoneal sarcoma account for only 10 to 20 % of all sarcomas with a Peak incidence is in the 5th decade of life. The most common histologic subtypes are liposarcoma and leiomyosarcoma, MFH (malignant fibrous histiocytoma) With the use of immunohistochemistry many of MFH are now diagnosed as lipo or leiomyosarcoma.[2-4] Most of the patients present as abdominal pain or fullness. Histological grade is the most important prognostic factor.[6] Pulmonary metastasis remains the major site of dissemination, except in cases of leiomyosarcoma. CECT is imaging modality of choice. Surgery offers only hope of cure but complete excision is possible in only 40-60% of cases. Recurrences are common following surgery, complete resection possible in 80% cases in first recurrence and 60 to 70% in subsequent recurrence. GIST (gastrointestinal stromal tumor), lymphoma, GCT (germ cell tumour) are most common differential diagnosis.

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MATERIALS AND METHODS

This retrospective study was based on study of total no of 20 patients getting admitted either to our regional cancer hospital (Acharya Harihara Cancer Hospital) or department of general surgery S.C.B. Medical College during the period Aug 2014 to Aug 2016 according to the inclusion criteria.
Inclusion criteria: Cases of retroperitoneal sarcoma treated between Aug 2014 to Aug 2016.

Exclusion criteria:
Patients who refused surgery or inoperable cases based on imaging.

Cases were studied according to the epidemiological parameters (age & sex), [Table 1] radiological imaging based staging [Table 3] & histological grading [Table 2]. Resectability [Table 4] was assessed & underwent radical resection whenever possible. Patients were followed up till their last clinical follow up or death.

RESULTS

Results of our study were arranged in tabular form as given below-

Table 1: Age and sex distribution.

Sex	No. of cases	%
Male	14	70
Female	6	30
Mean Age	55(21-70)yrs	

Table 2: Grading of Tumour.

GRADE	N	%
G1	6	30
G2	4	20
G3	10	50

Table 3: (TNM Status)

T status	N status	M status
T1	N0 = 18	M0 = 20
T2 = 6	N1 = 2	M1 = 0
T3 = 4		
T4 = 10		

In our retrospective study of 20 patients in our regional hospitals showed that most of the patients were male (n=14,70%) [Table 1] with mean age of 55yrs. [Table 5] shows most common organ resected was kidney (3), colon (2) [Figure 2], pancreas (1). Almost 90% (n=18) patients underwent complete surgical resection [Table 4] except 10% (n=2) patients underwent partial resection were due to involvement of major vessels (IVC, AA). Most of the patients presented with high grade tumours (n=10,50%) a [Table 2] and 50% were presented with T4 stages [Table 3]. Most of the tumours were liposarcoma (45%) followed by leiomyoma (25%) [Table 6] At the end of follow up 3 patients died of recurrence and another 2 underwent re-excision.

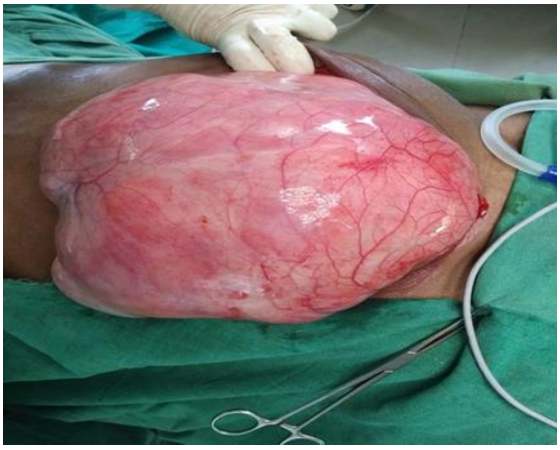

Figure 1: Retroperitoneal Tumor

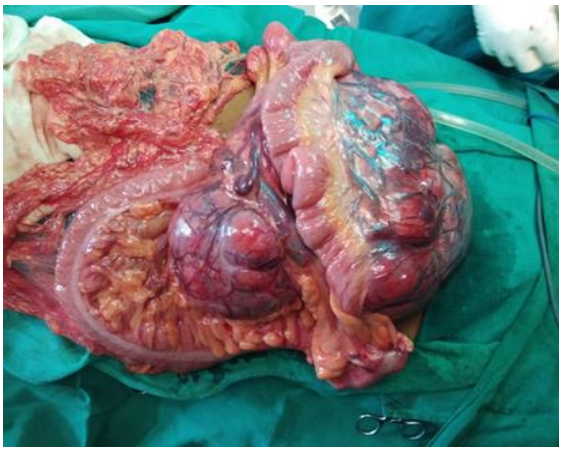

Figure 2: Enbloc resection of the tumour with Transverse colon

Table 4: (Resectability According to Grading of Tumour)

Resectability	Complete resection (No.)	Partial resection	Biopsy	Total
Grade 1	6	0	0	6
Grade 2	4	0	0	4
Grade 3	8	1	1	10
Total	18	1	1	20

Table 5: (No. of Organs resected during enblock resection)

Organ resected	number
Kidney	3
Colon	2
Pancreas	1

Table 6: (Histological Subtypes)

Histological Subtypes	n	%
Liposarcoma	9	45
Leiomyosarcoma	5	25
MFH	4	20
Lymphangioma	1	5
Malignant paraganglioma	1	5

DISCUSSION

Retroperitoneal tumours are very rare tumours affecting usually in the 5th decade. Usually spreads through blood stream .lymph node metastasis is very rare (<5%) except in certain histological types like myxofibrosarcoma, angiosarcoma, rhabdomyosarcoma, clear cell sarcoma, epitheloid sarcoma. Usually enlarging masses, masses>5 cm, deep to fascia need to be clinically evaluated by history, imaging & biopsy. MRI is the preferred technique for the extremity tumours whereas CT is the better modality for retroperitoneal & intra-abdominal sarcomas.[5,8,13] PET FDG imaging usually reserved for recurrences. Usually the 6prognosis depends upon the staging [Table 7a & 7b] & histological grading (G1, G2, G3 the most important factor). As the tumour is locally aggressive resection of the tumour along with adjacent organs need to be excised enbloc [Table 5 & Figure 2] like kidney, adrenal, small or large intestine.[17,18] Pre or post op radiation may be helpful in certain circumstances. In spite of complete resection 5 year survival is still poor (51%) & re resection may be required for recurrences which are

usually local & occur usually within 1st 2 years.[5,8] Follow up usually recommended by CT OR MRI every 3 to 4 months for 2 years & then 6 monthly for next 3 to 5 years.

Table 7 (a): ?

Histological grade (G)	
G1	Well differentiated
G2	Moderately well differentiated
G3	Poorly or very poorly differentiated
Primary site (T)	
T1	Tumor less than 5 cm in diameter
T1a	Superficial tumor
T1b	Deep tumor
T2	Tumor 5 cm or more in diameter
T2a	Superficial tumor
T2b	Deep tumor
N.B. Retroperitoneal and pelvic sarcomas are classified as deep tumors	
Nodal involvement (N)	
N0	No histologically verified metastases to lymph nodes
N1	Histologically verified regional lymph nodes

Table 7 (b): American Joint Committee staging of soft tissue sarcomas

Stage	Classification	Description
IA	GI, T1, N0, M0	Grade 1 tumor, <5 cm in diameter no regional lymph nodes and/or distant metastases
IB	GI, T2, N0, M0	Grade 1 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIA	G2, T1, N0, M0	Grade 2 tumor, <5 cm in diameter, no nodes and/or metastases
IIB	G2, T2, N0, M0	Grade 2 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIA	G3, T1, N0, M0	Grade 3 tumor, <5 cm in diameter, no nodes and/or metastases
IIIB	G3, T2, N0, M0	Grade 3 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIC	G1–3, T1, 2, N1, M0	Tumor of any grade and/or size, with regional involved nodes, but no metastases
IVA	G1–3, T3, N0, N1, M0	Tumor of any grade invading bone vessels/nerves, with/without nodes, no metastases
IVB	G1–3, T1–3, N0, N1, M1	Tumor with distant metastases

This analysis showed that liposarcoma (n=9,45%) was the most common encountered Retroperitoneal sarcoma followed by leiomyosarcoma (n=5,25%) Rate of resectability was 90% due to proper case selection and better imaging studies. 2 year survival was 85%. Major problem was recurrence. Grade was inversly associated with prognosis.

CONCLUSION

Complete surgical resection is the goal of treatmentmost effective treatment for primary/ recurrent RPS, but frequently not possible. Complete surgical resection ispossible in 40-60% cases. Incomplete surgical excision increases chances of local recurrence. Each recurrence decreases chances of cure. Multiorgan resection often required to attain negative margins.

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