



## A STUDY OF VARIABLES IN PRIMARY RETROPERITONEAL SARCOMAS : SINGLE INSTITUTE EXPERIENCE

### General Surgery

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### ABSTRACT

**BACKGROUND:** Retroperitoneal soft tissue sarcomas (RPSs) are rare and arise from connective tissue. They grow insidiously and present late with large tumour size. Wide surgical resection with negative margins is the best mode of management. In this research article we share our experience of managing RPSs in our institute. Material and methods : A retrospective study was undertaken in the Department of Surgery, GCS Hospital. Ten cases of operated RPS who underwent all the pre-operative work up followed by wide excision in our institute from January 2018 to June 2021 were included. Histopathology report was studied and values correlated. Results: The mean age of presentation was 43. 40% patients presented with abdominal pain, 70% had a palpable lump, 60% tumours were malignant and liposarcoma was the most common malignant tumour. R<sub>0</sub> resection was obtained in all benign tumours and 50% of malignant tumours. Local recurrence occurred in 30% of the patients all of which were malignant. Conclusions: RPSs pose a challenge to surgeons due to their insidious onset, large tumour size and involvement of vital structures. Complete surgical excision (R<sub>0</sub>/R<sub>i</sub>) is the best mode of treatment to obtain optimum overall survival. Close follow up is recommended due to recurrence.

### KEYWORDS

primary retroperitoneal tumours, soft tissue sarcoma, liposarcoma

### INTRODUCTION

Retroperitoneum soft tissue sarcomas (RPSs) are rare. They can be benign or malignant. They originally develop from soft tissues and may sometimes involve the organs as well as vital structures.<sup>[1]</sup> Approximately 70–80% of primary retroperitoneal soft tissue tumours are malignant; however, these account for just 0.1–0.2% of all malignancies.<sup>[2]</sup> The most frequent sarcomas are liposarcoma, malignant fibrous histiocytoma, and leiomyosarcoma, respectively.<sup>[3]</sup> Benign paraganglioma, neurofibroma, lipoma, angiofibroma, and schwannoma can be listed among the benign tumors.<sup>[3]</sup> Due to the inaccessibility of the region and chronic and vague symptoms they are usually large at presentation.<sup>[4]</sup> They are commonly seen in age group 50 to 70 years and have a 5-year survival rate of 40% to 50%.<sup>[5,5]</sup> Imaging carries a great importance in the diagnosis of RPSs. A study of the tumour and its proximity and involvement of surrounding vital structures and metastasis is done using CECT or MRI of abdomen and pelvis. FNAC or core needle biopsy is needed for accurate pathological diagnosis.<sup>[6]</sup>

Complete surgical resection of the primary tumour with optimal negative surgical margins is the first and the most effective line of management. Presence of a positive margin decreases the 5-year survival rate to 28%.<sup>[3]</sup> Further, AJCC 8th edition<sup>[7]</sup> mentions that prognosis can be determined by the Fédération Nationale des Centres de Lutte Contre Le Cancer (FNCLCC) criteria as used in extremity soft tissue tumours.<sup>[8]</sup>

This study evaluates the epidemiology, intra operative challenges and surgical outcomes and recurrence of RPSs in a single institution.

### MATERIALS AND METHODS

A retrospective study was carried out in which patients admitted in the Department of Surgery, GCS Hospital, Ahmedabad between January 2018 - June 2021 with radiological investigation suggesting operable primary retroperitoneal sarcomas were selected. Medical records were studied in detail and only those patients with complete records and follow up data were taken up.

**Inclusion Criteria:** All operated and histologically proven primary RPSs were included.

**Exclusion Criteria:** Inoperable and recurrent tumours were excluded.

Patients with radiologically confirmed RPSs were subjected for core needle biopsy. After pre-operative consent and optimisation, they underwent exploratory laparotomy. Histopathological parameters were evaluated. Follow up was noted and oncophysician opinion was also taken for adjuvant therapy. Data was compiled and studied in

using Microsoft excel software.

### RESULTS

A total of 10 patients were operated during the course of study, out of which 6 were males and 4 were females. The age group ranged from 16 to 70 years with an average age of 43.2 and median age being 39 years. (Table 1)

**Table 1 : Age and gender distribution**

Type	Benign	Malignant
R0	4	3
R1	-	3
R2	-	-
Total	4	6

The most common symptom was chronic abdominal pain in 50% followed by abdominal distention in 30% of the patients. There was a palpable lump in 70% of the patients.

All patients had undergone exploratory laparotomy for excision of tumour. 2 out of 10 patients underwent concurrent resection of surrounding structure i.e. right hemicolectomy and right nephrectomy respectively. 60% of the tumours turned out to be malignant and rest were benign. R<sub>0</sub> resection was obtained in all benign tumours. The patients with benign tumours did not report recurrence in the follow up period till June 2021. In cases of malignant tumour, recurrence was seen in 3 out of 6 cases. Out of the six, three cases had R<sub>i</sub> resection margin while the rest had R<sub>0</sub> margins. (Table 2)

**Table 2 : Resection margin for the tumours**

Type	Benign	Malignant
R0	4	3
R1	-	3
R2	-	-
Total	4	6

40% of the tumours belonged to T4 category. Further, out of those, 33.33% of the tumours were more than 20 cm in size. (Table 3)

**Table 3 : Tumour size for T staging**

Tumour size of RPT	Total	T category
≤5 cm	2	T1
>5 cm and ≤10 cm	2	T2
>10 cm and ≤15 cm	2	T3
>15 cm	4	T4

Liposarcoma was the most common malignant sarcoma (50%). The

malignant RPTs were graded according to the FNCLCC to obtain the prognosis. 3 out of 6 were grade 1 and the other 50% were grade 2. Local recurrence was reported in 3 out of 10 cases. Out of those three, two were liposarcomas. All patients survived during the entire study period.

## DISCUSSION

RPS is a challenging entity for a surgeon due to inaccessibility, its enormity and difficulty in achieving a complete resection. They are most commonly seen in 5th to 7th decade of life.<sup>[3]</sup> Toulmonde et al reported a median age of 57 with age range from 18-89 years<sup>[9]</sup> while Garcia-Ortega et al reported a median age of 47 with a range of 18-82 years.<sup>[10]</sup> In our study, the age group ranged from 16-70 with a median age of 39 years.

Joual A. Et al mentions abdominal pain and lump as being the most common symptoms.<sup>[11]</sup> Findings seen by Gemici et al are along the same lines with abdominal pain being a common symptom in 64.28% of the patients.<sup>[3]</sup> Our study also had chronic abdominal pain as the chief complain in most patients (50%). 20% of the patients had complain of a palpable lump. Retroperitoneal soft tissue sarcomas usually attain large sizes and the symptomatology is presented chronically at a later stage.

70% patients had a clinically palpable lump. Lump was most commonly palpable at right lower quadrant (30%). However, in a study by Gemici et al, lump was most commonly palpable at left upper and lower quadrants respectively (21.42%).<sup>[3]</sup>

Approximately 70–80% of primary retroperitoneal soft-tissue tumors are malignant.<sup>[2]</sup> Gemici et al revealed 75% of RPSs to be malignant.<sup>[3]</sup> Our study revealed 60% of the tumours to be malignant.

Out of this, liposarcoma was the most common malignant RPS (50%). Studies by Gemici et al (38.1%), Toulmonde M et al (64.7%) and Garcia-Ortega et al (61.1%) also show liposarcoma to be the most common malignant RPT.<sup>[3,9,10]</sup>

Studies conducted at John S Hopkins University School of Medicine showed that a complete surgical resection was the mainstay treatment.<sup>[12]</sup> Achieving R<sub>0</sub>/R<sub>1</sub> surgical resection for RPS increased the cure rate and prevented locoregional recurrence.<sup>[3]</sup> The difference of surgical outcomes between R<sub>0</sub> and R<sub>1</sub> resection have not been confirmed in the studies however, R2 resections have shown poor outcomes.<sup>[13]</sup> We were able to get an R<sub>0</sub> resection in 100% of benign RPSs and 50% of malignant RPSs. R<sub>1</sub> resection was achieved in 50% of the remaining malignant RPS. Additional organ resection was necessary in 17.85% in the Turkish review<sup>[3]</sup> while it was 20% in our study i.e. right hemicolectomy and right nephrectomy respectively.

Tumour size is an important prognostic and staging factor for RPSs. Tumours greater than 20 cm tends to have poorer prognosis.<sup>[14]</sup> In our study, 40% of the tumours were over 15 cm in size, similar to results obtained by other studies.<sup>[3]</sup> Further, 33.33% of malignant RPSs were over 20 cm in size.

For malignant RPSs, grade of tumour is one of the negative prognostic factors.<sup>[15]</sup> Our study revealed 50% of the tumour to be grade II and rest to grade I. This is in contrast to study by Patkar S et al who had maximum patients of grade III tumour.<sup>[15]</sup>

In our study, 30% of the tumours showed recurrence while a study by Virseda Rodríguez JA et al showed a recurrence rate of 45%.<sup>[16]</sup> Further, liposarcoma was the most commonly recurrent tumour in our study. Similar results were obtained in by Parkar S et al.<sup>[15]</sup>

## CONCLUSION

Primary retroperitoneal sarcomas are a rare entity. Owing to its late presentation as well as proximity and involvement of vital structures, one needs an experienced surgical team to manage it. Complete surgical resection with negative margins is the best modality of treatment. Grading of tumour and its size gives an idea regarding the prognosis. Due to high recurrence, regular monitoring is recommended.

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