

# A Study of Clinicopathological Spectrum and Surgical Procedural Diversity of Retroperitoneal Tumors and Tumors Originating from Retroperitoneal Solid Organs Over a Two-Year Period

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## Abstract

**Background:** The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms that can be either primary or metastatic lesions. Retroperitoneal tumours can cause a diagnostic dilemma and present several therapeutic challenges because of their rarity, relative late presentation and anatomical location. **Material and Methods:** To compare Retroperitoneal tumors and tumors arising from retroperitoneal organs on the basis of clinic-pathological spectrum and surgical management. **Results:** Surgery is the cornerstone of treatment in both groups compared in our study. For primary disease, this is frequently carried out upfront once the diagnosis is established and staging has been completed, verifying the absence of metastatic disease. Surgery is challenging given the frequently large size of the tumor and close proximity to critical organs and great vessels within the limited space of the retroperitoneum and intraabdominal cavity. **Conclusion:** Though by location both groups of tumors are same but they vary in variety of other aspects which are discussed in details in the study.

**Keywords:** Retroperitoneal tumors, Clinico-pathological spectrum, Surgical resection, Complications.

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## INTRODUCTION

The retroperitoneum hiding behind the peritoneum is full of diversity. Retroperitoneum is bounded anteriorly by posterior parietal peritoneum, posteriorly by transversalis fascia, superiorly by the diaphragm and inferiorly by the pelvic brim. The surgical approach to retroperitoneum is technically demanding. Primary retroperitoneal tumors are defined as a growth that arises from the connective tissues, nerves, blood vessels, or vestigial structures within the retroperitoneal space. Approximately 70-80% of retroperitoneal tumors are malignant and retroperitoneal malignancy accounts for 0.1-0.2% of all malignancies. The major retroperitoneal solid organs are kidney, adrenal and pancreas. The primary retroperitoneal tumors and organ based retroperitoneal malignancies presents in different ways. Usually, the patients with retroperitoneal tumors presents to a clinician when significant in size -average presenting size 11-20 cm, with vague nonspecific symptoms like dragging sensation, back pain, abdominal swelling or discomfort, palpable mass, early satiety, lower limb swelling and less commonly with gastrointestinal obstruction and urinary obstruction. However, these symptoms do not appear until tumor has grown to some extent. So, the tumors are often quite large in size when discovered. Nearly all the primary retroperitoneal pathologies behave in the same way. Most of the tumors originating from

retroperitoneal solid organs may present with organ specific symptoms or systemic symptoms depending upon the organ of origin. They may present with symptoms even when they are very small in size. This study will deal with the signs and symptoms, pathological diagnosis, operation performed, operative difficulties during the procedure, multi-visceral resection needed or not.

**Aims and objectives:** Tumors of the retroperitoneal space can be divided into two groups like primary retroperitoneal tumor (not arising from any organ) and tumours arising from retroperitoneal solid organ. There are several studies in literature that described the different perspectives of retroperitoneal tumors. But no such study is there to compare the primary retroperitoneal tumor and tumor arising from retroperitoneal solid organs. The distinction between primary

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retroperitoneal tumor and retroperitoneal tumor arising from named organ may have a unique clinical perspective in both diagnosis and treatment. In this study, the following aspects will be described in details for both the primary retroperitoneal tumors and tumor arising from retroperitoneal organs and comparisons will be done

1. Gender wise distribution.
2. Age wise distribution
3. Pathological spectrum
4. Operative planning and complications.

## MATERIALS AND METHODS

Study is based on the patients attending Department of Surgical Oncology IPGMER AND SSKM HOSPITAL, Kolkata, from 1st January 2024-31st December 2025.

### Primary Retroperitoneal Tumors [Not Arising from Any Organ]

Retroperitoneal tumors are classified based on the type of tissue of origin.<sup>[1]</sup> More than 90% of retroperitoneal tumors are of mesodermal origin, mostly liposarcoma, leiomyosarcoma, and undifferentiated pleomorphic sarcoma.<sup>[2]</sup> The characteristics of each tumor are different, which makes it difficult to determine the optimal treatment strategy for retroperitoneal tumor.<sup>[3]</sup> Primary retroperitoneal tumors are rare group of neoplasms with an incidence of 0.3% to 3%. Retroperitoneal sarcoma is a rare tumor that accounts for 0.15% of all malignant tumors and approximately 15% of all sarcomas.<sup>[4]</sup> Retroperitoneal tumor is rarely found when the tumor is small, as symptoms are less common in the early stages than in later ones.<sup>[5]</sup> Thus, the tumor is typically found after it has grown to a considerable size and often requires extensive resection of the surrounding organs. For this reason, the prognosis of Retroperitoneal sarcoma is worse than that of other sarcomas.<sup>[6]</sup>

The pathological diagnosis of tumor is difficult for several reasons, as follows: (1) the histological diagnosis of sarcoma itself has changed significantly with time (e.g., malignant fibrous histiocytoma was renamed undifferentiated pleomorphic sarcoma in the 4th edition of the WHO classification in 2013, and 13 diseases not included in the 4th edition were added to the 5th edition in 2020); (2) approximately 5% of sarcomas are unclassifiable; and (3) with the introduction of genetic testing, many tumors with new fusion genes have been identified, and tumors that were previously thought to be the same tumor are now known to follow different clinical courses (e.g., Ewing sarcoma and CIC sarcoma).<sup>[7-10]</sup> The French Federation of Cancer Centers (FNCLCC) system is the standard for grading histological malignancies of retroperitoneal tumors.<sup>[11]</sup> In this system, tumor differentiation, mitotic count, and tumor necrosis are scored, and the histological grade of the tumor is evaluated in three stages. The evaluation of tumor malignancy according to

the FNCLCC system is used for tumors in the TNM classification of the Union for International Cancer Control (UICC) and American Joint Committee on Cancer (AJCC). However, in the latest UICC/AJCC TNM 8th edition, the distinction between G2 and G3 is no longer reflected in the tumor staging.

### Primary tumors arising from retroperitoneal organ

- **Tumors arising from kidney:** Among the tumors arising from retroperitoneal organs, kidney neoplasms are not very un-common. Most common renal tumor is renal cell carcinoma. Median age of diagnosis is 65 yrs. Approximately 60% of kidney cancers are incidentally detected. In CECT, an enhancement of >20 HU measured in the CMP compared with the unenhanced phase marks a tumor as indeterminate and suspicious for RCC. Optimal sensitivity and specificity for RCC detection are reported when a cut off value of 84 HU in the CMP and 44 in the NP are used for the tumor.
- **Tumors Arising From Adrenals:** ACCs account for only 0.05–0.2% of all cancers,<sup>[12,13]</sup> or 1–2 patients per 1 million population per year.<sup>[14]</sup> The age distribution of the affected population is bimodal, with an increased incidence in infants and children younger than 5 years old and in individuals in their fourth and fifth decades of life.<sup>[15,16]</sup> A female preponderance has been noted,<sup>[13,14]</sup> and women are more likely than men to present with more well-differentiated tumors that tend to be functional.<sup>[17]</sup>
- **Tumors arising from pancreas:** Pancreatic cancer refers to the carcinoma arising from the pancreatic duct cells, pancreatic ductal carcinoma, and is the fourth leading cause of cancer deaths in the United States. The 5-year survival rate in the United States ranges from 5% to 15%. Surgical resection is the only current option for a cure, but only 20% of pancreatic cancer is surgically resectable at the time of diagnosis.<sup>[18]</sup>

## RESULTS

Total no of patients of my study is 34 who were divided on the basis of their origin. So, my result, analysis, discussion and conclusions are based on these 34 reported patients.

In our study out of total 34 patients, 18 of them were diagnosed with tumors arising from solid organ of retroperitoneum which include kidney, pancreas and adrenal. The rest 16 cases were diagnosed with tumors arising from epithelial tissues and non-epithelial tissues but not from any specific solid organ. These include Liposarcoma, Leiomyosarcoma, solitary fibrous tumor, GIST, Dermatofibrosarcoma protuberance [DFSP], Castleman's disease, Schwannoma, Smooth muscle tumor of uncertain malignant potential [STUMP], Synovial sarcoma. In our study 5 patients expired during post-operative period.

Non-Organ Based Retroperitoneal Tumors

### Age wise distribution of non-organ based tumors

**Table 1: table depicting age wise distribution of non-organ based tumors.**

| Pathology of tumor     | 12-20 yrs | 21-30 yrs | 31-40 yrs | 41-50 yrs | 51-60 yrs | 61-70 yrs | 71-80 yrs |
|------------------------|-----------|-----------|-----------|-----------|-----------|-----------|-----------|
| Liposarcoma            |           |           | 1         | 1         | 2         |           |           |
| Leiomyosarcoma         |           |           |           | 1         |           |           |           |
| solitary fibrous tumor | 1         |           |           |           |           |           |           |

|                     |   |   |   |   |   |   |   |
|---------------------|---|---|---|---|---|---|---|
| GIST                |   |   |   | 1 |   |   |   |
| DFSP                |   |   |   | 1 | 1 |   |   |
| Castleman's disease |   |   | 1 | 1 |   |   |   |
| Schwanoma           |   |   | 1 |   |   |   |   |
| STUMP               |   |   |   | 2 |   |   |   |
| Synovial sarcoma.   |   |   |   | 1 |   |   |   |
| Paraganglioma       |   | 1 |   |   |   |   |   |
| Total               | 1 | 1 | 3 | 8 | 3 | 0 | 0 |

In the following study we can observe that incidence of non-organ-based tumors of retroperitoneum occur in between 40-50 yrs of age. Our study does not include patients of pediatric age group.

Gender wise distribution of non-organ based tumors

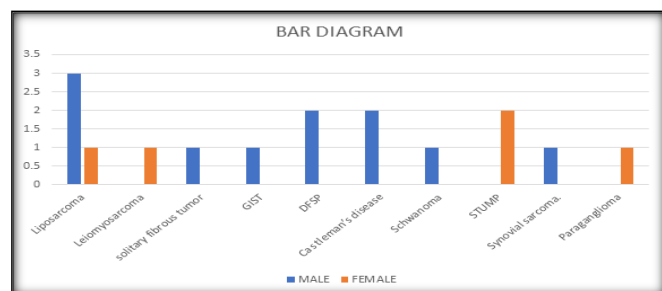


Figure 1: Bar diagram depicting gender wise distribution of non-organ based tumors.

In the following study we can observe that incidence of non-organ-based tumors of retroperitoneum were seen mostly in male population attending Department of Surgical Oncology, SSKM.

Pathological spectrum of non-organ based tumors

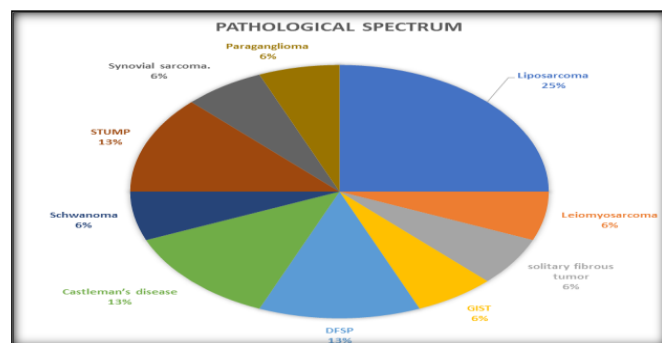


Figure 2: Pie chart depicting pathology wise distribution of non-organ based tumors.



Figure 3: Primary retroperitoneal tumor resected En-bloc with transverse and descending colon.

In our study we found out that out of 16 cases of tumors which have non-organ-based origin, 22% [highest] were diagnosed to be Liposarcoma and pathological types with highest malignant potential include Liomyosarcoma and `Liposarcoma.



Figure 4: Primary (Non-organ-based) huge retroperitoneal tumor insinuated into left inguinal canal.

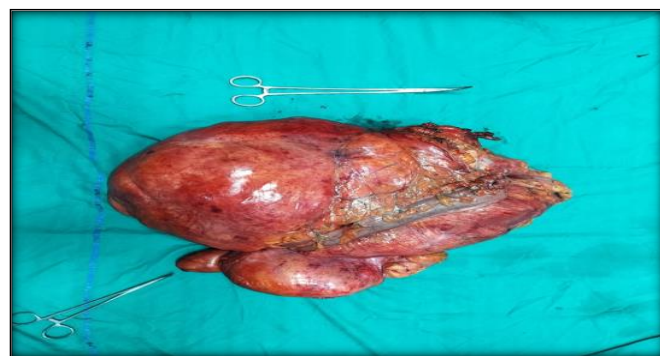


Figure 5: Specimen of Non-organ-based huge retroperitoneal tumor weighing approximately 15 kgs.

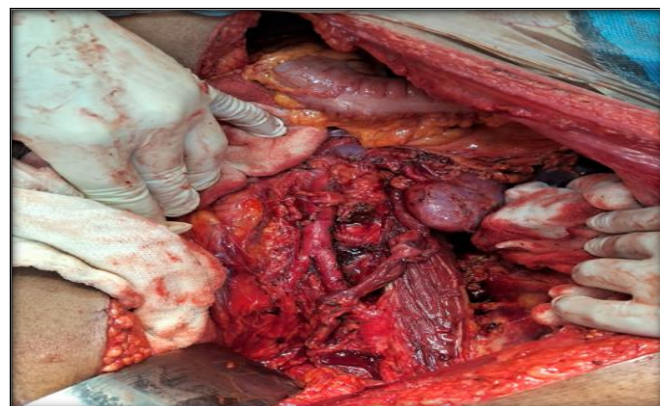


Figure 6: The tumor bed after resection of the huge retroperitoneal tumors.

Operative planning and complications of non-organ based tumors.

**Table 2: table depicting operative complications and planning of non-organ based tumors**

| Types of tumors        | Type of incision                                         | Intra-op complications                                                         | Adherence to surrounding structure                    | Post-op mortality     | Deviation from initial pre-op planning                                       |
|------------------------|----------------------------------------------------------|--------------------------------------------------------------------------------|-------------------------------------------------------|-----------------------|------------------------------------------------------------------------------|
| Liposarcoma            | midline in three cases and T shaped incision in one case | Excessive blood loss [>700ml] in intra-op period noted in 2 out of three cases | Intimate adhesion to Lt kidney was noted in one case  | One out of four cases | Lt sided nephrectomy was performed in one case along with tumor excision     |
| Leiomyosarcoma         | midline                                                  | Incomplete excision                                                            | IVC and Iliac vessels were completely encased         | nil                   | Incomplete resection had to be done                                          |
| solitary fibrous tumor | T shaped incision                                        | Diaphragmatic injury                                                           | Close relation with Lt kidney and spleen              | nil                   | Diaphragmatic repair had to be done in one case along with tumor excision    |
| GIST                   | midline                                                  | uneventful                                                                     | Colonic wall was inseparable from tumor               | nil                   | Rt Hemicolectomy was performed along with tumor removal                      |
| DFSP                   | Both cases midline incision                              | Diaphragmatic injury in one case                                               | Tumor invaded 9th and 10th rib in one case            | One out of two cases  | Diaphragmatic repair and excision of 9th and 10th ribs were done in one case |
| Castleman's disease    | One case midline other case T shaped incision            | Both cases were uneventful                                                     | Close relation to Rt renal vein and IVC in both cases | nil                   | No deviation from pre-operative planning                                     |
| Schwannoma             | midline                                                  | Excessive blood loss [> 1000ml] and incomplete resection                       | Tumor originating from neural foramina of sacrum      | nil                   | No deviation from pre-operative planning                                     |
| STUMP                  | One case midline other case T shaped incision            | Excessive intra-op blood loss [>1500ml] in one case                            | nil                                                   | One out of two cases  | No deviation from pre-operative planning                                     |
| Synovial sarcoma.      | midline                                                  | uneventful                                                                     | nil                                                   | nil                   | No deviation from pre-operative planning                                     |
| Paraganglioma          | midline                                                  | uneventful                                                                     | nil                                                   | nil                   | No deviation from pre-op plan.                                               |

**Organ Based Retroperitoneal Tumors:** In our study out of total 34 patients ,18 of them were diagnosed with tumors arising from solid organ of retroperitoneum which include

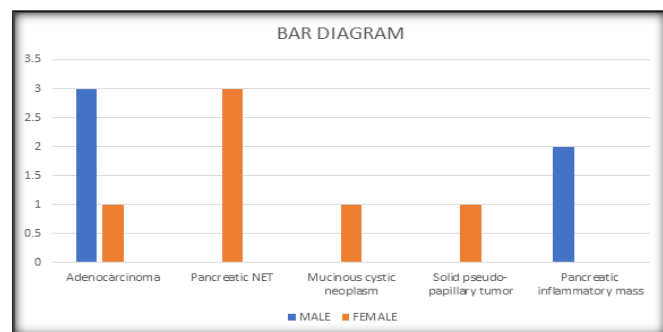
kidney, pancreas and adrenal.

**Pancreas**

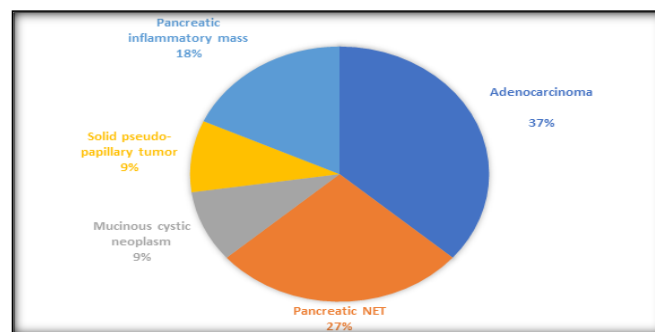
**Age wise distribution.**

**Table 3: depicting age wise distribution of Pancreatic tumors.**

| Pathology of tumor           | 12-20 yrs | 21-30 yrs | 31-40 yrs | 41-50 yrs | 51-60 yrs | 61-70 yrs | 71-80 yrs |
|------------------------------|-----------|-----------|-----------|-----------|-----------|-----------|-----------|
| Adenocarcinoma               |           |           |           | 2         | 2         |           |           |
| Pancreatic NET               |           |           | 2         | 1         |           |           |           |
| Mucinous cystic neoplasm     |           |           |           | 1         |           |           |           |
| Solid pseudo-papillary tumor | 1         |           |           |           |           |           |           |
| Pancreatic inflammatory mass |           |           |           |           |           | 2         |           |
| TOTAL                        | 1         |           | 2         | 4         | 2         | 2         | 0         |



**Figure 7: Bar diagram depicting gender wise distribution of Pancreatic tumors.**



**Figure 8: Pie chart depicting pathology wise distribution of Pancreatic tumors.**

In our study total 11 cases were found to be originating from Pancreas. Highest incidence of pancreatic tumors is noted to be in 40-50 years age group in our study. This group also includes inflammatory mass lesion of pancreas which are sequelae of Chronic Pancreatitis where the patients present with obstructive jaundice and pre-op biopsy reported as high-grade dysplasia in both cases.

**Gender Wise Distribution:** In the following study we can

observe that incidence of Pancreatic tumors was seen mostly in female population attending Department of Surgical Oncology, SSKM.

**Pathological Spectrum:** In our study we found out that out of 11 cases of tumors which have Pancreatic origin, 37% [highest] were diagnosed to be Adenocarcinoma.

Operative planning and complications

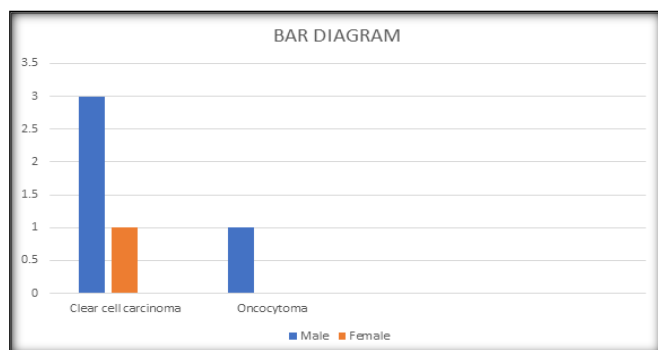
**Table 4: depicting operative complications and planning of Pancreatic tumors**

| Types of tumors              | Type of incision                                                | Complications                            | Post-op mortality     | Deviation from initial pre-op planning                                                                       |
|------------------------------|-----------------------------------------------------------------|------------------------------------------|-----------------------|--------------------------------------------------------------------------------------------------------------|
| Adenocarcinoma               | Roof top incision given in all 4 cases                          | Pancreatico-jejunostomy leak in one case | One out of four cases | One out of four cases were inoperable so palliative bypass surgery was done                                  |
| Pancreatic NET               | Makuchi Incision in 2 cases and Roof-top incision in one case.  | Pancreatico-jejunostomy leak in one case | nil                   | Colonic resection was required in one out of three cases where lesion was in pancreatic tail invading colon. |
| Mucinous cystic neoplasm     | Midline incision                                                | Uneventful                               | nil                   | No deviation from pre-operative planning                                                                     |
| Solid pseudo-papillary tumor | Midline                                                         | Uneventful                               | nil                   | No deviation from pre-operative planning                                                                     |
| Pancreatic inflammatory mass | Makuchi Incision in one case and Roof-top incision in one case. | Uneventful                               | One out of two cases  | No deviation from pre-operative planning                                                                     |

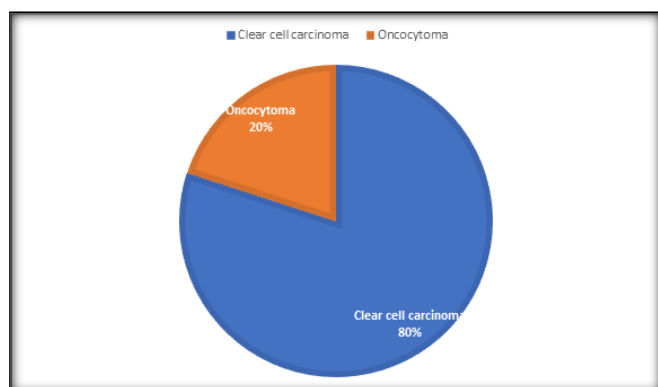
**Kidney: Age wise distribution**

**Table 5: depicting age wise distribution of Kidney tumors.**

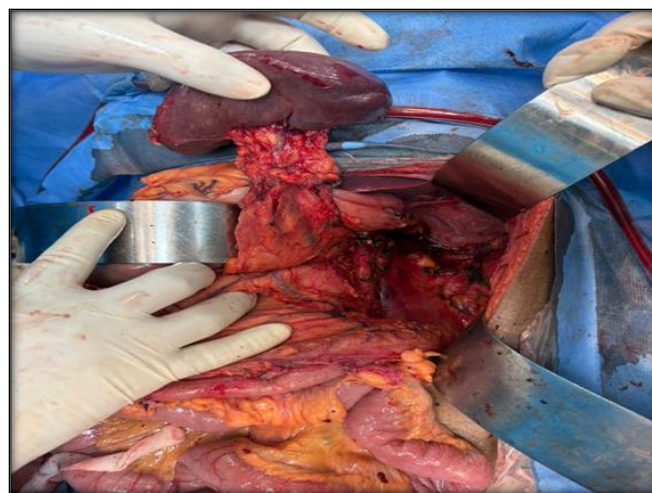
| Pathology of tumor   | 12-20 yrs | 21-30 yrs | 31-40 yrs | 41-50 yrs | 51-60 yrs | 61-70 yrs | 71-80 yrs |
|----------------------|-----------|-----------|-----------|-----------|-----------|-----------|-----------|
| Clear cell carcinoma |           |           |           |           | 2         |           | 2         |
| Oncocytoma           |           |           |           |           | 1         |           |           |
| TOTAL                | 0         | 0         | 0         | 0         | 3         | 0         | 2         |



**Figure 9: Bar diagram depicting gender wise distribution of Kidney tumors.**



**Figure 10: Pie chart depicting pathology wise distribution of Kidney tumors.**



**Figure 11: Spleen and Pancreas has been lifted up from retroperitoneum to remove left sided renal tumor.**

In our study total 05 cases were found to be originating from Kidney. Upon post-operative biopsy 4 cases were found to be Clear cell Renal Carcinoma, two of them were in the age group of 50-60 years and the rest two were in the age group of 70-80 years. Only one case was reported to be Renal Oncocytoma which was in the age group of 50-60 yrs, thus showing higher incidence of renal tumors in the 50-60 years of age group in our study.

**Gender Wise Distribution:** In the following study we can

observe that incidence of Renal tumor was seen mostly in male population attending Department of Surgical Oncology, SSKM.

**Pathological Spectrum:** In our study we found out that out

of 5 cases of tumors which have Renal origin, four cases were diagnosed to be Clear cell renal carcinoma and only one case was reported to be Oncocytoma.

**Operative planning and complications**

**Table 6: depicting operative complications and planning of Kidney tumors**

| Types of tumors      | Type of incision                                                 | Complications | Post-op mortality | Deviation from initial pre-op planning                                                                |
|----------------------|------------------------------------------------------------------|---------------|-------------------|-------------------------------------------------------------------------------------------------------|
| Clear cell carcinoma | Sub-costal incision in two cases and Flank incision in two cases | Uneventful    | nil               | Pancreas and spleen mobilization was performed in one out of four case, rest three cases no deviation |
| Oncocytoma           | Flank incision was taken                                         | Uneventful    | nil               | No deviation from pre-operative planning                                                              |

**Adrenal:** In our study only one case of Adrenal gland tumor was reported, a female patient of age 32 years attended Surgical Oncology Department of SSKM with a pre-op FNAC report showing poorly differentiated adrenal carcinoma. Intra-operative findings showed left lobe of liver metastasis and the adrenal mass was infiltrating Inferior Venacava and was extending till diaphragm behind the liver, so the tumor was deemed inoperable and only biopsy was taken which was same as pre-op FNAC report. In the second case laparoscopic left sided adrenalectomy was done for an 18-year old female patient and post operative biopsy was reported as Ganglioneuroma.

## DISCUSSION

Retroperitoneal tumors sometimes possess challenges to Surgical Oncologists. These are often massive in size and in close relation with major vessels and have propensity for multi-visceral resection. These make any surgery daunting, especially more so when the tumors are large occupying most of the abdomen. Both organ-based and non-organ-based tumors of retroperitoneum are same as per location but there is a great variation in clinico-pathological characteristics. Based on the study done in our centre, we can compare them as follows:

**Table 7: depicting comparison between primary and organ based retroperitoneal tumors.**

| Sl no. | Characteristics                                       | Organ-based tumors                                                                                                                                                                                                                                                 | Non-organ-based tumors                                                                                                                                                                                                                     |
|--------|-------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1      | Age                                                   | For pancreatic based tumors most common age group is 40-50 years and for kidney tumors it is 50-60 years                                                                                                                                                           | Most common age group for non-organ-based tumors is 40-50 yrs.                                                                                                                                                                             |
| 2      | Gender                                                | Pancreatic and adrenal tumors are most common in female patients and kidney tumors in male patients                                                                                                                                                                | Most commonly seen in male patients                                                                                                                                                                                                        |
| 3      | Mean size of detection of neoplastic lesion           | 1.Pancreatic tumors-4cm.<br>2.Kidney tumor: 8.8 cm.<br>Adrenal tumors have not taken due to small sample size.                                                                                                                                                     | All the primary retroperitoneal tumors had a mean detection size of 21 cm.                                                                                                                                                                 |
| 4      | Pathological diversity                                | Most common tumor arising from pancreas is Adenocarcinoma and from kidney is clear cell carcinoma. Only one adrenal tumor was reported which was found to be poorly differentiated carcinoma                                                                       | Diversity is seen more in non-organ-based tumors, most common tumor in our study is liposarcoma                                                                                                                                            |
| 5      | Pre-Operative planning and intra-operative deviations | There is not much deviation in pre-operative planning during intra-op period. Only one case out of all kidney tumors in our study needed special intervention apart from convention. Two cases out of all pancreatic tumors needed deviation from pre-op planning. | In non-organ-based tumors, nearly all cases showed deviation from pre-op planning with high incidence of multi-visceral resection, hemorrhage, injury to surrounding structure leading to prolonged operative time and operative morbidity |
| 6      | Mortality                                             | Mortality is less according to our study-11.11%                                                                                                                                                                                                                    | Mortality is more according to our study-18.75%                                                                                                                                                                                            |
| 7      | Relationship between tumor size and symptoms          | Even a smaller sized tumor can be symptomatic so they are detected early [particularly in case of pancreatic tumors]                                                                                                                                               | Symptoms arise after the size of tumor becomes considerably large so they are detected late [often presents with abdominal lump]                                                                                                           |

**Pitfalls:** The following study is a study done in the department of surgical oncology, IPGMER, Kolkata, India, which is a tertiary referral center. So, drawing comparison with world literature may not be accurate. In our study we have not included pediatric oncology as there is separate department for such cases. There is also a separate Urology department thus limiting exposure to Uro-oncological cases.

## CONCLUSION

Retroperitoneal tumors encompass a wide variety of lesions, making them challenging to diagnose due to their nonspecific and overlapping clinical and imaging characteristics. A

thorough understanding of retroperitoneal anatomy, key histological features, and a systematic approach to evaluating these lesions is essential for narrowing the differential diagnosis.<sup>[19]</sup>

During the diagnostic workup of a retroperitoneal mass, a large variety of histological findings in this location must be kept in mind; approximately half of these retroperitoneal masses are mesenchymal tumours.<sup>[20]</sup>

Tumors arising from retroperitoneal organs like kidney, pancreas and adrenals are relatively common. Specific blood investigations and urine analysis play a major role in their diagnosis. Surgical resection with negative margins is the mainstay of treatment in both types of tumours for better

outcome.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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