Original Article

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A Nine Years Histopathological Retrospective Study of Retroperitoneal Masses

Pathology Section

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ABSTRACT

Introduction: Retroperitoneum is potentially large space that allows both primary and metastatic tumours to grow silently before clinical signs and symptoms appear. However, retroperitoneal masses due to their uninhibited growth and with no facial boundaries they are often attain large size before the development of symptoms and the establishment of a diagnosis. There are a vast number of neoplasms occurring in this region. Understanding the pathology and lesions is important, for early diagnosis and for treatment of these neoplasms.

Aim: To study the overall prevalence of primary and secondary retroperitoneal neoplasms and the varied histomorphological spectrum of retroperitoneal masses. Also to categorise the lesions as benign and malignant tumors of retroperitoneum.

Materials and Methods: This retrospective study was conducted in the Department of Pathology, Karnataka Institute of Medical Sciences Hubballi, Karnataka, India from 2011 to 2019. A total of 39,757 specimens were recieved to histopathology section during this period. Among them, 53 (0.13%) were retroperitoneal tumours. Haematoxylin and Eosin (H&E) stained slides were thoroughly analysed and categorised into non neoplastic, benign and malignant tumours. Further, based on the finer microscopic features, the accurate subtype of the tumour type was made. The descriptive statistics like mean, range and percentage are used.

Results: Total 53 cases of retroperitoneal tumours were studied of which 10 (18.9%) cases were primary retroperitoneal masses and 43 (81.1%) cases were secondary retroperitoneal masses. Mean age was 40.7 years with male to female ratio 1.5:1. Malignant lesions were more common accounting to 50.94% cases, followed by benign 22.64% cases, the rest were intermediate grade and inflammatory. Kidney lesions were most common secondary retroperitoneal masses, accounting for 41.5% cases, followed by pancreas 22.64% cases, adrenals 13.20% cases and Lymphnodes 3.7% cases. Primary retroperitoneal masses which accounted for 18.9% cases were categorised histomorphologically as spindle cell tumours, adipocytic, small round cell tumours, cartilaginous origin and cystic lesions.

Conclusion: Diagnosis of retroperitoneal tumours is challenging due to their varied histomorphology and histopathology forms the gold standard for diagnosis.

Keywords: Retroperitoneum, Secondary neoplasms, Spindle cell tumours

INTRODUCTION

The retroperitoneal space is the portion of the lumboiliac region limited anteriorly by the peritoneal covering, posteriorly by the posterior abdominal wall, superiorly by the twelfth rib and vertebra, inferiorly by the base of the sacrum and iliac crest, and laterally by the side borders of the quadratus lumborum muscles. Its contents, embedded in a meshwork of loose connective tissue are the adrenal glands, kidneys and ureters, aorta and its branches, inferior vena cava and its tributaries, and numerous lymphnodes [1].

This potentially large space allows both primary and metastatic tumours to grow silently before clinical signs and symptoms appear. The problem dealing with retroperitoneal masses is primarily due to their uninhibited growth with no facial boundaries often leading to the large size attained before the development of symptoms and the establishment of a diagnosis. The proximity to vital vascular and neural structures as well as intra abdominal organs adds to the problem [2].

There is very few research articles published in the literature, which have dealt with the retroperitoneal tumours in a nutshell. This study is undertaken to understand the spectrum of lesions occurring in this region. Further, histopathology is still the gold standard method to study and analyse these tumours. Ancillary techniques like immunohistochemistry, aid in confirming the diagnosis. Hence, histomorphological study of the retroperitoneal masses is critical for diagnosis.

Present study aimed to analyse the overall prevalence of primary and secondary retroperitoneal neoplasms and the varied histomorphological spectrum of retroperitoneal masses. Also to categorise the lesions as benign and malignant tumors of retroperitoneum.

MATERIALS AND METHODS

This retrospective study was conducted in the Department of Pathology at Karnataka Institute of Medical Sciences, Hubballi, Karnataka, India, from 2011 to 2019. A total of 39,757 specimens were received to histopathology section during this period. Among them, 53 (0.13%) were retroperitoneal tumours. All procedures performed in the current study were approved by Institutional Ethical Committee in accordance with the 1964 Helsinki declaration and its later amendments.

Inclusion and Exclusion criteria: All the neoplastic and non neoplastic lesions arising in the retroperitoneal space were included in the study. Lesions arising in the intra peritoneum and the autolysed tissues were excluded from the study.

Procedure

The formalin fixed tissue specimens of retroperitoneal masses were received from the Surgical Departments and the tissue sections were properly processed and stained with Haematoxylin and Eosin (H&E). The authors analysed the H&E stained slides which were retrieved from the Department Records. The data regarding the age, sex and the clinical findings were taken from the clinical database. The H&E stained slides were categorised into non neoplastic, benign and malignant tumours. Further, based on the finer microscopic features, the accurate subtype of the tumour type was made. In cases of soft tissue tumours, immunohistochemistry was performed, to ascertain the findings and final diagnosis was made.

STATISTICAL ANALYSIS

The descriptive statistics like mean, range and percentage are used. They are calculated and tabulated using Microsoft Excel 2017.

RESULTS

Total 53 cases of retroperitoneal tumours were studied of which 10 (18.9%) cases were primary retroperitoneal masses and 43 (81.1%) cases were secondary retroperitoneal masses. The age ranged from 9 months to 72 years, with mean of 40.7 years. The study comprised of 32 (60.37%) males and 21 (39.63%) females. Male to female ratio is 1.5:1.

The majority of the cases were in the age group of 61-70 years comprising of 13 (24.52%) cases. The youngest case was, 9 months of age diagnosed as nephroblastoma and the eldest was 72 years diagnosed as pheochromocytoma [Table/Fig-1].

Age group (years)	n, %
0-10	6 (11.33%)
11-20	1 (1.88%)
21-30	11 (20.75%)
31-40	6 (11.33%)
41-50	9 (16.98%)
51-60	6 (11.33%)
61-70	13 (24.52%)
71-80	1 (1.88%)
Total	53 (100%)

[Table/Fig-1]: Age distribution of retroperitoneal tumours in the present study.

The kidney was the most commonly involved retroperitoneal organ in our study accounting to 22 (41.51%) cases, followed by pancreas 12 (22.64%) cases, primary retroperitoneal masses 10 (18.87%) cases, adrenals 7 (13.20%) cases and lymphnodes 2 (3.78%) cases as shown in [Table/Fig-2].

Organs involved	No. of cases	Percentage (%)
Kidney	22	41.51
Pancreas	12	22.64
Primary retroperitoneal masses	10	18.87
Adrenal	7	13.20
Lymphnodes	2	3.78
Total	53	100

[Table/Fig-2]: Organ wise distribution of tumours in the retroperitoneum.

In the present study, malignant lesions were more common accounting for 27 (50.94%) cases, followed by benign 12 (22.64%) cases, 1 (1.89%) case of intermediate grade and 13 (24.53%) cases were inflammatory lesions. The distribution of non neoplastic and neoplastic lesions are shown in the [Table/Fig-3]. Malignant and intermediate grade comprised a majority of the tumours, 28 (52.8%) and non neoplastic and benign were 25 (47.2%).

Organs involved	Benign	Malignant	Inflammatory	Intermediate grade	Total
Kidney	2 (9.09%)	16 (72.72%)	4 (18.19%)	-	22
Pancreas	-	3 (25%)	9 (75%)	-	12
Primary retroperitoneal masses	4 (40%)	5 (50%)	-	1 (10%)	10
Adrenal	6 (85.71%)	1 (14.29%)	-	-	7
Lymphnodes	-	2 (100%)	-	-	2
Total	12 (22.64%)	27 (50.94%)	13 (24.53%)	1 (1.89%)	53

[Table/Fig-3]: Spectrum of non neoplastic and neoplastic tumours of the retroperitoneum.

The retroperitoneal tumours are divided into primary and secondary tumours. The primary tumours include the tumours arising from the soft tissue of the retroperitoneum while the secondary tumours are the tumours arising from the solid organs in the retroperitoneum like kidney, pancreas, adrenals and the lymphnodes. The results are compiled based on the frequency of occurrence.

1) Kidney: Nephrectomy was the predominant type of specimen authors received of the total 22 cases. Among them, 16 (72.7%) were malignant tumours, 2 (9.09%) were benign and 4 (18.19%) cases were inflammatory. Renal Cell Carcinoma (RCC) was the most common malignant tumour. Ten cases (45.5%) of renal cell carcinoma was encountered, in the 5th and 6th decade, followed by 4 (18.18%) cases of Wilm's tumour in younger age group, with age of presentation being 1st and 2nd decade of life and 2 (9.09%) cases of urothelial carcinoma of the renal pelvis in 7th decade of life. Two (9.09%) cases of benign tumours were encountered in 2nd and 6th decade. Four (18.18%) cases of chronic pyelonephritis presented in the 6th and 7th decade of life. In RCC, the clear cell variant was the most common histomorphological variant followed by the spindle cell variant of RCC [Table/Fig-4].

Of the two benign lesions, one case each of angiomyolipoma and medullary cystic kidney were encountered at 65 years and 21 years respectively [Table/Fig-5].

Four cases of non neoplastic lesions of kidney were studied, all were cases of chronic pyelonephritis. One was a case of tuberculosis pyelonephritis presenting at an age of 66 years [Table/Fig-6] and the rest three were cases of non specific chronic pyelonephritis presenting at 6th and 7th decade. One case showed hydrouretric changes.

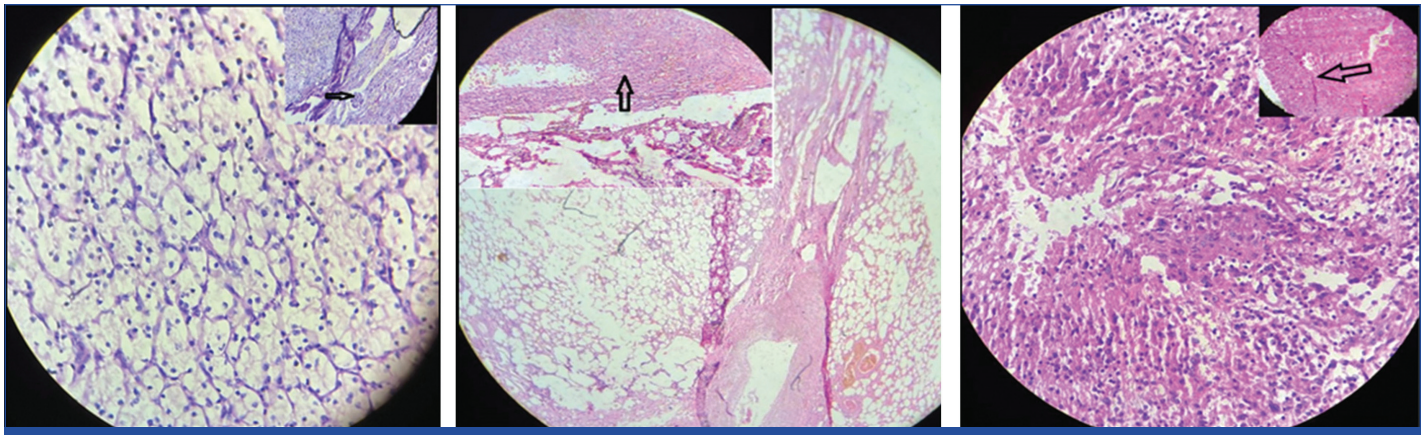
2) Pancreas: Pancreatic tumours were the second most common secondary tumours in the retroperitoneum, in the present study comprising 12 (22.64%) cases. Among them, 9 (75%) of them were pseudocyst of pancreas, presenting at 4th decade of life and 3 (25%) cases were malignant in 5th-6th decade. Three (25%) specimens of the whipples procedure were received, of which two were pancreatic adenocarcinoma presenting at 5th decade of life. The other case was a neuroendocrine carcinoma at 60 years of age. One of the pancreatic adenocarcinomas showed two distinctive components, a spindle cell component with osteoclastic giant cells and a adenocarcinoma component. This case was subjected to immunohistochemistry to rule out Gastrointestinal Stromal Tumour (GIST). Adenocarcinoma component was immunopositive for pancytokeratin, Cytokeratin 7 (CK7), SMAD4 and vimentin [Table/Fig-7]. Spindle cell component was positive for vimentin. CD117 and CK20 was negative. A final diagnosis of anaplastic carcinoma of pancreas with giant cells was reported.

3) Primary retroperitoneal masses: Authors encountered various mesenchymal lesions in primary retroperitoneal masses which were categorised histomorphologically as spindle cell tumours, adipocytic, small round cell tumours, cartilaginous origin and cystic lesions. Among them 5 (50%) cases were malignant, 1 (10%) was intermediate and 4 (40%) were benign.

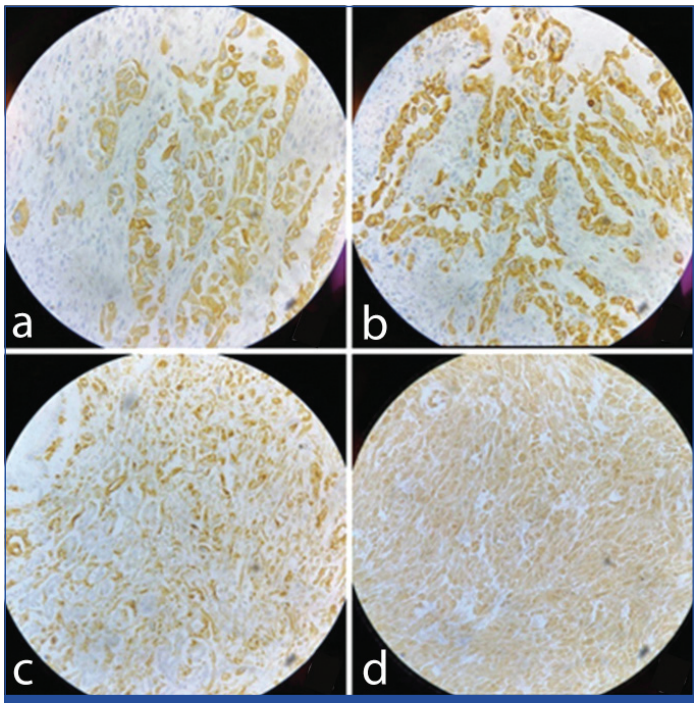
- **Spindle cell tumours:** Spindle cell pattern was the most commonly encountered histomorphological pattern among the primary peritoneal masses comprising of 4 (40%) cases. Among them, 2 (50%) were malignant,1 (25%) was intermediate grade and 1 (25%) was benign.

Among malignant spindle cell lesions, one case each of malignant peripheral nerve sheath tumour and malignant spindle cell tumour at 45 years and 65 years of age, respectively [Table/Fig-8]. The benign lesion was a case of cellular leiomyoma at 45 years of age. One case of the intermediate grade of spindle cell tumour at an age of 40 years was studied. The intermediate grade tumour was an inflammatory pseudotumour.

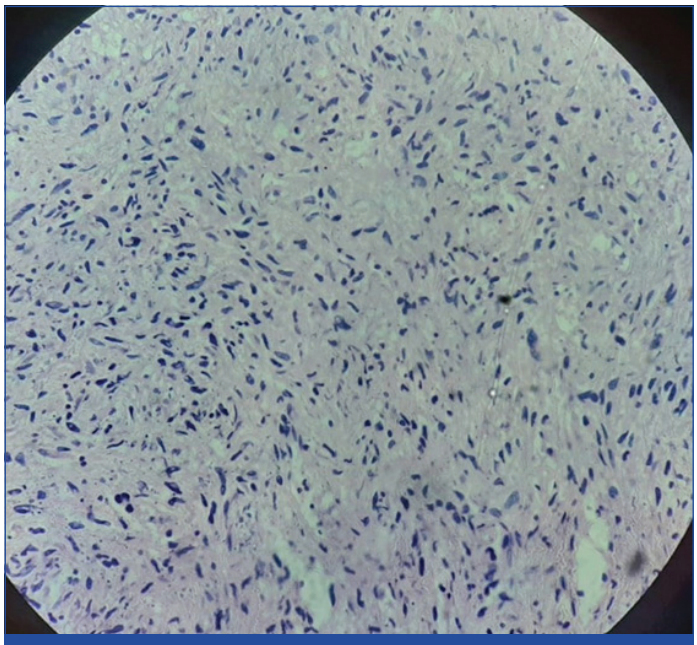
- **Small round cell tumours:** Two (20%) cases of small round cell tumours, were studied which was the second most common



[Table/Fig-4]: Renal Cell Carcinoma: Nests of clear cells separated by thin fibrovascular stroma (H&E,400X). Inset image: Normal Kidney (arrow) with adjacent tumour (H&E,100X). **[Table/Fig-5]:** Angiomyolipoma (H&E,100X). Inset: Normal Kidney proximal tubules with areas of haemorrhage (arrow) with adjascent tumour (H&E,100X). **[Table/Fig-6]:** TB Pyelonephritis, Granuloma with caeseous necrosis (H&E,400X). Inset: Normal Kidney (arrow) with adjascent caeseous necrosis (H&E,100X). (Images from left to right)



[Table/Fig-7]: Immunohistochemistry of anaplastic carcinoma pancreas: a) Pancytokeratin CK-Positive; b) CK 7- Positive; c) Vimentin- Positive. d) SMAD4-Positive.



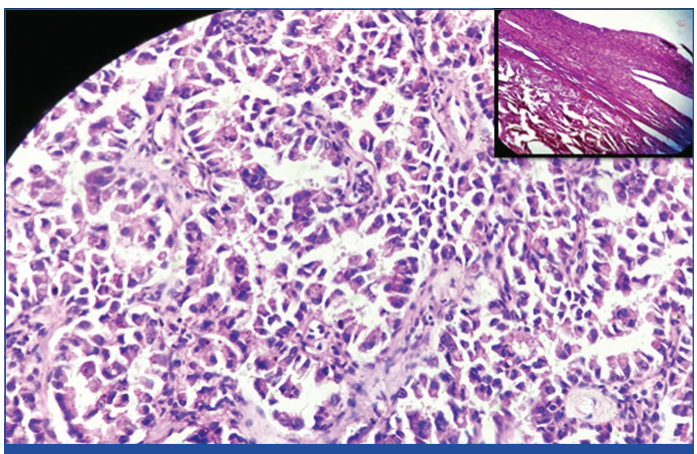
[Table/Fig-8]: Malignant peripheral nerve sheath tumour: Malignant spindled cells having pointed ends and are wavy in nature (H&E, 400X).

among the primary peritoneal masses encountered. Age of presentation was first decade of life. One case was subjected to immunohistochemistry and was positive for Desmin. A final diagnosis of, Embryonal Rhadomyosarcoma, the specific type of SRCT was made.

- **Adipocytic tumours:** Two (20%) cases of benign adipocytic lesions i.e Lipoma were studied. The age of presentation was 50 years and 63 years.
- **Cartilaginous tumours:** There was 1 (10%) case of chondrosarcoma- grade II, who presented at an age of 55 years.
- **Hamartomatous cyst:** One (10%) case of hamartomatous cyst was encountered which had both epithelial and mesenchymal component. The cyst was lined by squamous and columnar epithelium.

4) Adrenals: Seven (12.96%) cases of adrenal lesions were encountered. Among them, 6 (85.71%) were benign and 1 (14.29%) was malignant. All six benign lesions were of pheochromocytoma of which, one presented as bilateral pheochromocytoma [Table/Fig-9]. The most common age of presentation of these lesions was in the 4th decade of life. One malignant case was of Neuroblastoma at the age of 1 year.

5) Lymphnode: There were 2 (3.71%) cases who presented as retroperitoneal lymphnodes. Both the cases were of Non Hodgkins Lymphoma at 52 years and 40 years of age.



[Table/Fig-9]: Pheochromocytoma (H&E,400X). Inset: Adrenal Cortex (H&E,100x).

DISCUSSION

Retroperitoneum is an area of curiosity and interest since long. Considering the numerous and heterogeneous contents of this region which includes adrenal, pancreas, kidney, rertroperitoneal lymphnode, gastrointestinal tract (second part of duodenum, ascending and descending colon) and retroperitoneum proper

lesions can be encountered here [3]. In the present study, 53 cases of retroperitoneal masses were encountered during the period of 2011 to 2019. The mean age of presentation was 40.7 years with age range being 9 months to 72 years which is in accordance with study by Gangopadhyay M et al., while study by Mangal N et al., had the majority of cases in the 6th decade [4,5].

Malignant lesions were more common among the retroperitoneal masses in the present study accounting for 27 (50.94%) and benign and non neoplastic lesions were 25 (47.2%). This was comparable with study by Mangal N et al., who also reported malignant masses as more common accounting for 47 (55.38%) and benign and non neoplastic masses as 38 (44.7%) cases [5].

Kidney was the most common retroperitoneal organ to be involved accounting for 22 (41.51%) cases, followed by pancreas 12 (22.64%) cases which is similar to the study by Pant H et al., who found 17 (34%) cases of renal origin followed by 14 (28%) cases as pancreatic origin [6]. Studies by Mangal N et al., [5], Mehdi G et al., [7], Das A et al., [8] and Ahmad SS et al., [9] also found kidney was the most common retroperitoneal organ to be involved.

Kidney: In the present study, a total of 22 (41.51%) cases of kidney were studied and nephrectomy was the predominant type of specimen received. The malignant neoplasm was commonly observed in kidney accounting for the majority, 16 (72.2%) of the cases. Renal cell carcinoma was the most common malignant tumour encountered in the present study in the 5th and 6th decade, which is similar to the findings by Rafique M [10] and Popat VC et al., [11]. Clear cell RCC was the common histological pattern observed which was comparable with Agarwal D et al., Hashmi AA et al., and Latif F et al., [12-14]. Wilms tumour was the next common malignant tumour observed in the 1st and 2nd decades of life. Agarwal D et al., found the incidence of Wilms tumour in the 1st decade of life [12].

Chronic pyelonephritis was the non neoplastic lesion encountered among which most were non specific while one case was TB pyelonephritis. However, Mangal N et al., found pyonephrosis as the most common non neoplastic lesion [5].

Pancreas: In the present study, 12 (22.64%) were pancreatic lesions. The majority of the lesions were non neoplastic accounting for 9 (75%) while 3 (25%) were malignant. Pancreatic lesions were more common in males than females. The non neoplastic lesion is the pseudocyst. However, Basina G et al., 27 (69.2%) were malignant and 11 (28.2%) were non neoplastic. But the sex distribution was similar [15].

Primary retroperitoneal masses: In the present study, 10 (18.87%) cases were primary tumours of retroperitoneum. They had varied morphology. Among them 5 (50%) cases were malignant, 1 (10%) case was intermediate and 4 (40%) were benign. Most of the lesions were in the 5th and 6th decade. Small round cell tumours were in the 1st decade. Spindle cell sarcomas and small round cell sarcoma were common malignant lesions and adipocytic lesions were the common benign neoplasm.

The incidence of benign neoplasms is less common than the malignant neoplasms. This finding is similar to Das A et al., and Rekhi B et al., [8,16].

Adrenals: Pheochromocytoma is the most common adrenal lesion encountered in the present study in the 4th decade of life, while neuroblastoma was found in an infant. Kumari NS et al., also found pheochromocytoma as the common adrenal neoplasm [17]. While Mangal N et al., found Neuroblastoma as common tumour [5].

Lymphnode: In the present study, Non Hodgkin's Lymphoma was the only lesion encountered in the retroperitoneal lymphnodes. Studies by Chakrabarti I et al., and Das A et al., found Non Hodgkin's Lymphoma as the most common lesion [8,18]. They encountered metastatic adenocarcinoma as the next common lesion in lymphnodes.

Limitation(s)

The sample size was small as the cases were compiled and studied only upto 2019 due to the COVID-19 pandemic. The number of surgeries was less during 2019 to 2021, only emergency surgeries were conducted during this period.

CONCLUSION(S)

The diagnosis of retroperitoneal lesions is challenging. The retroperitoneal space allows the growth of both primary and secondary tumours silently before the appearance of clinical signs and symptoms. Most of the retroperitoneal masses are malignant. Accurate diagnosis of these neoplasms is critical for treatment. The sensitivity and specificity of Fine Needle Aspiration (FNA) smears is high but, histopathological pattern is of prime importance for further management. Hence, histopathology still holds the gold standard for diagnosis and further management. Ancillary tests aid in confirmation of the findings and also in making a final diagnosis.

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