



Original Research Article

Rare tumors of retroperitoneum-A diagnostic challenge

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ABSTRACT

Introduction: Retroperitoneum can show a wide spectrum of diseases, including a variety of rare benign and malignant neoplasms that can be either primary or metastatic. Most retroperitoneal tumors are malignant, and soft tissue sarcomas account for one third of the cases. The most common sarcomas occurring in the retroperitoneum are Liposarcomas, Malignant Fibrous Histiocytomas, and Leiomyosarcomas. Subtyping of these tumors is important for the different targeted therapeutic options available.

Aims and Objectives: To study the distribution of benign and malignant retroperitoneal tumors.

Materials and Methods: Study design – Prospective study done in the department of Pathology from January 2013 to December 2017. Immunohistochemistry (IHC) was done wherever necessary.

Results: A total of 62 retroperitoneal tumours were studied of which 19 were benign & 43 were malignant. Rare benign tumors encountered were Adrenal Myelolipoma, Benign Fibrous Histiocytoma, Fibrolipomatous Hamartoma and multiple Paragangliomas. Rare malignant tumors were adult onset Wilms' tumor, De-differentiated Liposarcoma, Epithelioid GIST, malignant GIST of the mesentery and Carcinosarcoma of Kidney.

Conclusion: Amongst the retroperitoneal tumors received, malignant tumors were higher in distribution than benign tumors. The commonest epithelial tumor being Renal Cell Carcinoma and mesenchymal tumor being Liposarcoma. The study of rare malignant tumors was done with the help of IHC markers.

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1. Introduction

Primary retroperitoneal neoplasms are rare comprising 0.1-0.2% of all malignancies.¹ In retroperitoneum malignant tumours are more frequent than benign lesions but elsewhere in the body benign diseases predominate. Soft tissue (mesenchymal) tumours of retroperitoneum are less common, but constitute 15% of all primary sarcomas.² Retroperitoneal tumours can cause a diagnostic dilemma and present several therapeutic challenges because of their rarity, relative late presentation and anatomical location often in close relationship with several vital structures in the retroperitoneal space.³ We aim to study the distribution of both benign & malignant tumors of the retroperitoneum with emphasis on the clinicopathological picture of the rare

tumors we encountered.

2. Materials and Methods

Biopsies of 62 retroperitoneal tumors from patients who were admitted to the hospital from January 2013 to December 2017 were studied. Complete clinical details including radiographic findings were procured. All specimens were fixed in 10% Formalin and then subjected to macroscopic examination. Representative samples were taken, processed and stained with routine Hematoxylin & Eosin stains. Special stains and IHC were used where ever necessary.

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3. Results

A total of 62 retroperitoneal tumours were studied. Amongst the 62 cases, 5 cases were under the age of 14 years and 57 cases were above 14 years. Males were 33 cases & Females were 29 cases with a male to female ratio being 1.1:1. Number of benign tumors in children were 1 case & in adults 18 cases. Total malignant tumors were 4 cases in children and in adults 39 cases. Table 1 shows the age wise distribution of all the neoplastic lesions.

Table 2 shows the various histological types of tumors in our study including rare types.

We attempted to classify retroperitoneal tumors encountered in pediatric age group and Table 3 illustrates the histological diagnosis of these tumors and the age group in which they were seen.

4. Discussion

Malignant tumors of retroperitoneum are four times more frequent than benign tumors. In our study also malignant tumors outnumbered benign tumors with a ratio of 2:1. This ratio is in sharp contrast to neoplastic disease occurring elsewhere in the body, where benign disease predominates.⁴

In the current study tumors occurred predominantly in the 5th and 6th decades with a male, female ratio of 1.13:1. Tumor size in most cases was >15cm with an average size of 12cm. Liposarcoma constituted the common histological type followed by Renal Cell Carcinoma and Lymph node metastases. Routine histological examination with clinical correlation aided in diagnosis of most of the cases, some requiring special stains and IHC. The age and sex incidence in our study was concordant with other studies in literature and are depicted in Table 4.

We encountered many rare tumors in the retroperitoneum, both benign and malignant. Adrenal Myelolipomas are extremely rare tumors, benign in nature and composed of a variable mixture of mature adipose tissue and hematopoietic elements.⁹ We diagnosed a case of Adrenal Myelolipoma in a 60-year-old male who presented with pain in upper abdomen and had the characteristic histological picture.

We reported a rare case of multiple Paragangliomas in a 40-year-old woman who presented with a history of throbbing headache and fever. On examination she was found to be hypertensive with ultrasound of abdomen showing two tumors, one in the left suprarenal region and the other at the region of the aortic bifurcation. Both the specimens received were 5x4.5x4cm with cut section showing encapsulation, grey tan and grey yellow areas (Figure 1A,B). Microscopically the tumors were similar with neoplastic cells arranged in the characteristic Zellballen patterns and also showed lymphovascular emboli in the pericapsular area (Figure 1C,D). IHC by Chromogranin and Synaptophysin for tumor cells, S-100 for the

sustentacular cells. Santosh R et al reported multiple Paragangliomas in a 14-year-old girl with similar locations as in our study.¹⁰

Only 3% of Wilms' tumors are reported in adults and most are diagnosed un-expectedly following nephrectomy for presumed Renal Cell Carcinoma.¹¹ A 53-year-old male presented with left flank pain and fever for 2 months. On examination a huge tender mass was palpable in the left side of abdomen with CT scan showing a large heterogeneous mass in the left kidney. Nephrectomy was performed and the specimen received showed a tumor almost replacing the entire kidney. The tumor was solid, partially lobulated with a variegated appearance (Figure 2A). Microscopy revealed Wilms' tumor with biphasic histology and epithelial predominance. Majority of the tumor showed primitive tubule and glomeruli formation admixed with a minimal mesenchymal component (Figure 2B,C). Varma et al described a Triphasic Wilms' tumor in a 48-year-old male with similar clinical presentation.¹¹

De-differentiation in a Liposarcoma can be an Undifferentiated Pleomorphic Sarcoma or resemble a carcinoma, melanoma, meningioma or a nerve sheath tumor. Chondrosarcomatous and Osteosarcomatous elements may also be seen.¹² A 58-year-old male patient presented with a mass in retroperitoneal region with loss of weight. An abdominal contrast-enhanced computerized tomography (CECT) revealed a large encapsulated mass. Surgery was done and specimen received was of size 18x15x12cm with cut section showing yellow-tan areas admixed with firm tan-gray areas (Figure 3A). Microscopically the tumor displayed areas of Well Differentiated Liposarcoma and non-lipogenic components of Mesenchymal Chondrosarcoma and Osteosarcoma. The interphase between the two zones was typically abrupt.

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms of the gastrointestinal tract. Rarely they occur outside the gastrointestinal tract (omentum, mesentery and retroperitoneum) and are called Extra-gastrointestinal stromal tumors (EGISTs).¹³ We reported a case of EGIST in retroperitoneum of a 64-year-old male patient. He presented with back pain since 4 months and CT scan showed a retroperitoneal solid mass in left paravertebral region. Microscopically the tumor was cellular and composed of sheets of round to ovoid cells with scant eosinophilic cytoplasm & dense nuclear chromatin (Figure 4A). The mitotic count was <5/50 High power fields (HPF) (Low mitotic activity). IHC showed positivity for CD117 (Figure 4B).

We also encountered a case of Malignant GIST in a 50-year-old male who presented with complaints of progressive pain abdomen for 4 months. CECT scan revealed a lobulated solid mass lesion in the retroperitoneum with necrotic areas. Ultrasound guided needle core biopsy showed a malignant spindle cell neoplasm with

Table 1: Age wise distribution in this study

Age	Number of patients
0-10 years	4
11-20 years	2
21-30 years	11
31-40 years	6
41-50 years	9
51-60 years	13
61-70 years	14
>70 years	3

Table 2: Distribution of various histological types of tumors

Most common benign lesions	Renal Oncocytoma
Most common malignant lesions	Wilms' tumor Clear Cell Renal Cell Carcinoma Liposarcoma Lymphnode metastasis
Rare benign lesions	Adrenal Myelolipoma Multiple Paragangliomas Benign Fibrous Histiocytoma Fibrolipomatous Hamartoma
Rare malignant lesions	De-differentiated Liposarcoma Clear Cell Sarcoma of Kidney Carcinosarcoma of Kidney Malignant GIST Adult onset Wilms' tumor

Table 3: Classification of pediatric cases

S.No	Age/sex	Diagnosis
1.	15y/F	Metastatic Dysgerminoma
2.	8y/F	Malignant Mixed Germ Cell Tumor (Embryonal carcinoma + Yolk sac tumor)
3.	2y/F	Classic triphasic Wilms' tumor
4.	3y/M	Clear Cell Sarcoma of Kidney
5.	15 days/M	Fibrolipomatous Hamartoma

Table 4: Comparison of Age & Sex distribution with other studies

Parameters	Stoeckle et al, 2001 ⁵	Van Dalen et al, 2001 ⁶	Lewis et al, 1998 ⁷	Gronchi et al, 2004 ⁸	Present study, 2018
Number of patients	165	142	500	165	62
Male : Female ratio	1.0:1.2	1.01:1.22	1.34:1.0	1.2:1.0	1.13:1
Age (years)					
Median	54	60	58	53	60
Range	16-82	18-88	16-88	15-82	15 days-74

high cellularity, fascicular growth pattern and large areas of necrosis (Figure 5A). Tumor showed high grade cellular features with mitotic activity of >10 per 50 HPF (Figure 5B). The tumor demonstrated classical CD117 positive features on IHC.

We reported a rare case of Carcinosarcoma of the kidney in a 56-year-old male patient who presented with right flank pain and hematuria. Surgery was done after radiological confirmation. We received a nephrectomy specimen with a tumor almost occupying the entire kidney (Figure 6A). Microscopically the tumor was composed of malignant glandular & sarcomatoid components with a Ki-67 index of 40% (Figure 6B). Xiu-Peng Zhang et al reported a similar case of Carcinosarcoma occurring in the renal pelvis in a 56-year-old male patient. In their case the tumor also showed chondroid and osteoid components which were not observed

in our case.¹⁴

For all our cases of soft tissue sarcomas we used the FNCLCC grading system developed by The French Federation of Cancer Centers Sarcoma Group which is based on the three histological parameters including tumor differentiation, mitotic rate and tumor necrosis.

5. Conclusion

To conclude, in our study malignant tumours were higher in distribution than benign tumours in both pediatric and adult age groups. The commonest epithelial tumour being Renal Cell Carcinoma and mesenchymal tumour being Well Differentiated Liposarcoma. Immunohistochemistry using an appropriate panel of antibodies was done to arrive at a definitive diagnosis. Rare tumours of epithelial &

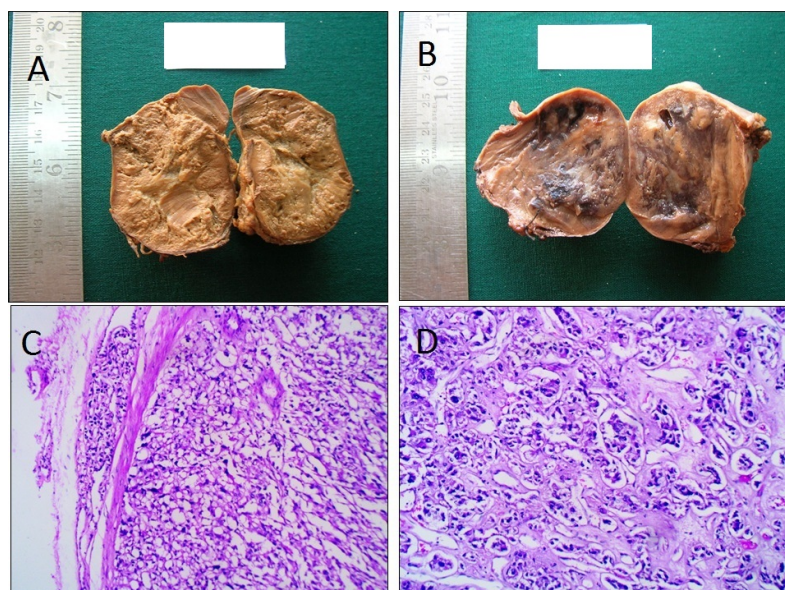


Fig. 1: Multiple Paragangliomas; **A:** Left suprarenal mass: C/S – encapsulated, grey tan & grey yellow; **B:** Mass at Aortic bifurcation: C/S – encapsulated, grey brown; **C:** Tumor with lymphovascular embolization (H&E x100); **D:** Tumor with characteristic Zell-ballen pattern and hyalinized stroma (H&E x100).

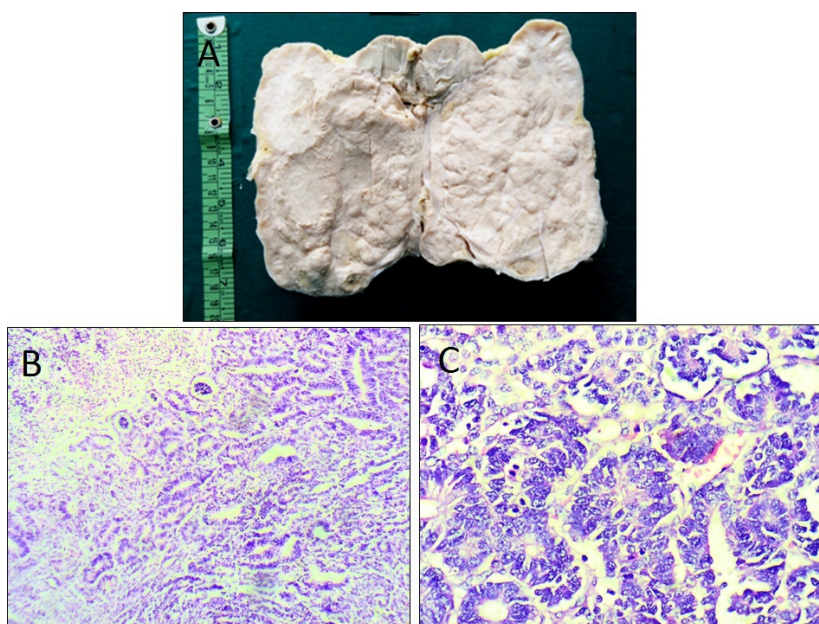


Fig. 2: Adult Wilms Tumor; **A:** Left radical nephrectomy specimen with tumor involving almost entire kidney; **B:** Epithelial predominant Wilms tumor (H&E x100); **C:** Tumor with attempted tubular & glomeruli formation (H&E x400).

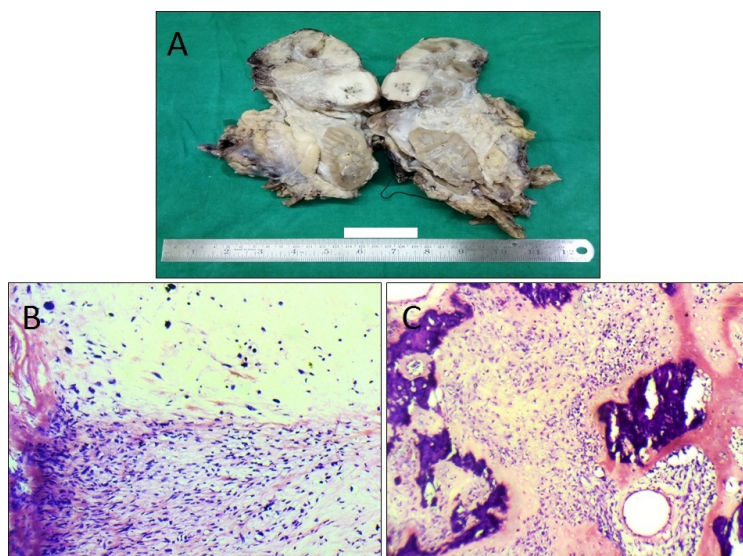


Fig. 3: De-differentiated Liposarcoma; **A:** 6x6cm mass, C/S: showing yellow-tan areas admixed with firm tan-gray areas; **B:** Well differentiated Liposarcoma with abrupt transition to mesenchymal Chondrosarcoma (H&E x100); **C:** Tumor with areas of Osteosarcoma(H&E x100).

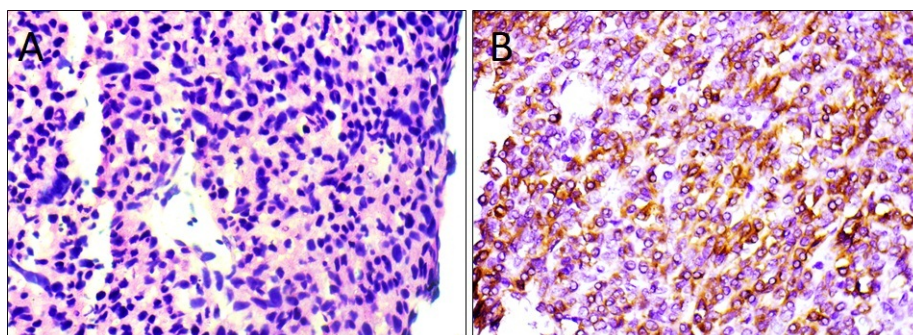


Fig. 4: Epithelioid GIST; **A:** Cellular tumor with sheets of round to ovoid cells, scant eosinophilic cytoplasm & dense nuclear chromatin (H&E x100); **B:** Tumor cells showing diffuse CD117 Positivity (IHCx100).

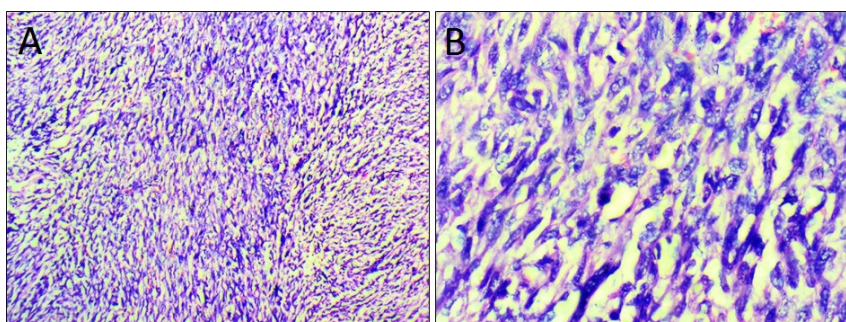


Fig. 5: Malignant GIST; **A:** Malignant spindle cell neoplasm showing high cellularity & fascicular growth pattern (H&E x100); **B:** Tumor showing high grade cellular features & high mitotic activity (H&E x400).

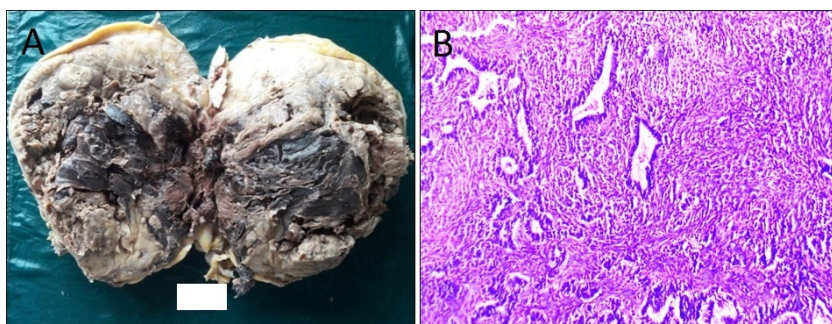


Fig. 6: Carcinosarcoma kidney; **A:** Nephrectomy specimen with tumor occupying entire kidney; **B:** Tumor with malignant glandular & sarcomatoid components (H&E x100).

mesenchymal origin were encountered both in adult & childhood age group.

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8. Competing interests:

Nil

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