ORIGINAL RESEARCH PAPER

Pathology

KEY WORDS: Immunohistochemistry, Retroperitoneum, Soft tissue sarcoma

A CLINICO PATHOLOGICAL STUDY OF RETROPERITONEAL TUMORS IN A TERTIARY CARE HOSPITAL - A 3 YEAR STUDY.

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ABSTRACT

BACKGROUND: Retroperitoneal tumors are rare tumors and approximately 1/3 are soft tissue sarcomas. This is a retrospective study to review the tumors which were reported in our institution and to highlight on rare tumors occurring in this site.

MATERIALS AND METHODS: Totally 72 cases of retroperitoneal tumors were analyzed retrospectively from August 2015 to July 2018 for 3 years in Government Mohankumaramangalam Medical College Hospital, Salem ,Tamilnadu.

RESULTS: In this study of 72 cases, Soft tissue tumors constituted 35%, which were mostly malignant (68%), Sympathetic nervous tissue tumors and extra adrenal tumors constituted 21% ,Small round cell tumours -14%, Germ cell tumors- 5%, Gastrointestinal stromal tumors(GIST)- 4 %. , Pancreatic tumors accounted for 14% and tumors of lymphnode -7%.

CONCLUSION: Sarcomas are the most common retroperitoneal tumors, liposarcoma is the most common and fibrosarcoma is the least common tumour . Myelolipoma and Neuroblastoma are rare tumors and should be considered in the differential diagnosis of fat containing tumors and Ewings sarcoma respectively .

INTRODUCTION:

Retroperitoneum is the second most common site of origin of primary soft tissue tumors, after the deep soft tissues of the lower extremities[1]. Liposarcoma, Pleomorphic sarcoma NOS and Leiomyosarcoma are accounting for more than 80% of primary retroperitoneal sarcomas[2]. Among these, Liposarcoma is the most common sarcoma in this site accounting for 10 to 15% of all liposarcomas and about 35% of retorperitoneal sarcomas[3]. Fibrosarcoma is the rarest retroperitoneal tumor. Neurogenic tumors constituted about 20% of retroperitoneal tumors including both benign and malignant[4]. Majority of the Neurogenic tumors are benign, seen in young population and includes Neurofibroma and Schwnnoma[5,6]. Paraganglioma and Ganglioneuroma are the tumors from sympathetic nervous tissue occurring in retroperitoneum. It is important to recognize Neuroblastoma in this site and to distinguish it from Ewings sarcoma. Symptoms secondary to retroperitoneal neoplasms are vague and appear late in the course of the disease and are related to displacement of organs and obstructive phenomena[7]. Retroperitoneal sarcomas in general have a worse prognosis than those with extremity sarcomas due to their large size before they become clinically apparent and inadequate surgical margins because of anatomic constraints[8]. Hence in this study an attempt has been made to study about the spectrum of retroperitoneal sarcomas.

AIM:

To find the distribution of various histological types, proportion and clinical presentation of retroperitoneal tumors in a tertiary care centre.

MATERIALS AND METHODS:

This is a retrospective study conducted in Department of Pathology, GMKMCH, Salem,Tamil Nadu. All patients with provisional diagnosis of retroperitoneal tumors based on clinical and radiological features registered from August 2015 to July 2018 for a period of 3 years were considered for this study. Immunohistochemistry was done wherever necessary for confirmation of diagnosis. A total of 72 histologically diagnosed cases of retroperitoneal tumors including soft tissue sarcomas, Germ cell tumors, Gastro intestinal stromal tumors (GIST), Pancreatic, Neural and lymphoid origin were included.

OBSERVATION AND RESULTS:

Out of 72 cases of retroperitoneal tumors, malignancy constituted about 58 cases (87%), benign accounted for 12 cases (13%) with age ranging from 4 years to 66 years and peak age group was 5th decade. Male:Female ratio was 4:1. The most common clinical presentation was palpable mass abdomen . Tumor size ranged from 5cm to 15cms, the most common histopathological diagnosis was sarcomas which constituted about 35%, of which liposarcoma is the predominant sarcoma reported. 2nd most common tumors were sympathetic nervous tissue origin (21%), of which benign and malignancy were 66% and 34% respectively. Small round cell tumors constituted (14%), Germ cell tumors - 5%, GIST -4%, Pancreatic tumors 14% and lymphomas constituted 7%.

Figure 1: Distribution of Retroperitoneal Tumors

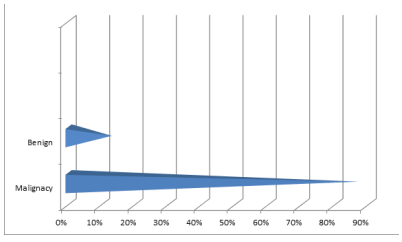

Table 1: DISTRIBUTION OF HISTOPATHOLOGICAL TYPES

HISTOPATHOLOGICAL DIAGNOSIS	NUMBER OF CASES	PERCENTAGE
SARCOMAS	25	35%
SYMPATHETIC NERVOUS TISSUE TUMORS	15	21%
SMALL ROUND CELL TUMOURS	10	14%
PANCREATIC TUMORS	10	14%
LYMPHOMAS	5	7%
GERM CELL TUMORS	4	5%
GIST	3	4%

DISTRIBUTION OF SARCOMAS

Liposarcoma was the most common sarcoma in retroperitoneum accounting for 36% , pleomorphic sarcoma NOS - 27%, MPNST- 18%, Leiomyosarcoma -16%, solitary fibrous tumor -2% and fibrosarcoma -1%. Among liposarcomas, myxoid variant constituted 67% and pleomorphic variant accounted for 33%.

Small round cell tumors constituted 14% including Embryonal Rabdomyosarcoma (Embryonal RMS) 3 cases (4.6%), Ewings Sarcoma - 6 cases (8.5%) and Desmoplastic small round cell tumor 1 case (1.4%) Most of these tumors were presented in children with palpable abdominal mass. Embryonal RMS and Ewings sarcoma were confirmed by IHC markers Myogenin and CD 99. Solitary fibrous tumor was confirmed with IHC marker CD 34.

GERM CELL TUMOURS

These tumors constituted 5% of retroperitoneal tumors, which included 2 cases of mature teratoma, 1 case of immature teratoma and 1 case of Yolk sac tumor.

SYMPATHETIC NERVOUS TISSUE TUMORS.

These tumors constituted 21%, of which benign and malignant tumors were 66% and 34% respectively. Paragangliomas were the most common benign tumor with male predominance and age ranging from 32 to 40 years. Malignant tumors reported were 4 cases of Neuroblastoma and 1 case of Ganglioneuroblastoma. Most of the neuroblastomas presented in the paediatric age group ranging from 4 to 5 years and size ranging from 3.5 to 5.5 cms , one patient was presented with hypertension.

EXTRA ADRENAL MYELOLIPOMAS were also reported in retroperitoneum. It is a rare benign tumor and clinically presented as asymptomatic mass and cut surface showed bright yellow with areas of tan brown discoloratlon.

PANCREATIC TUMOURS

14 % of Pancreatic tumors including 5 cases of ductal adenocarcinoma,2 cases of mucinous cyst adeno carcinoma, one case of inflammatory pseudo tumor, Pancreatoblastoma and solid pseudopapillary tumor each. The overall tumor size ranged from 6 cm to 15cm. Inflammatory pseudotumor was presented in 66years male with palpable mass and headache.

Figure 2: HPE & IHC of Retroperitoneal Tumors

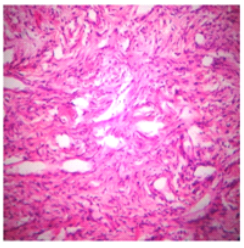

Figure 2A: HPE -Solitary Fibrous Tumor

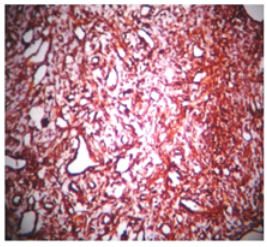

Figure 2B: IHC- CD34- Solitary Fibrous tumor

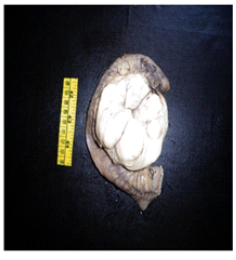

Figure 2C: Gross-Leiomyosarcoma

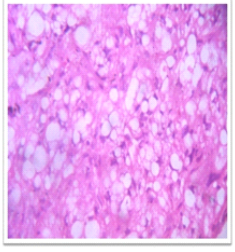

Figure 2D: HPE- Liposarcoma

DISCUSSION

Sarcomas were the most common tumors among retroperitoneal tumors in our study, with peak incidence in the age group of 50 to 60 years. These findings are correlating with following studies.

Table 2: Age Distribution of Retroperitoneal Tumors

STUDY	NO OF CASES	MEDIAN AGE IN YEARS
Stockle et al 2001[9]	165	54
Lewis et al 1998[10]	500	58
Present study	72	52

The histopathological analysis of specimen revealed that Liposarcoma was the most common sarcoma in retroperitoneal region constituting about 36% which correlated with most of other studies in the literature. Perter GA et al study showed the similar findings.[11]

Solitary fibrous tumors and fibrosarcomas were the least common sarcoma in retroperitoneum constituting 2% and 1% respectively.

Among sympathetic nervous tissue tumors, benign (66%) were more common than malignant tumors (34%), of which Paragangliomas were the predominant one.

Myelolipomas can occur in this region similar to those of adrenal glands and is a benign tumour in this extra adrenal site and are ususally asymptomatic which is correlating with literature [12] and it is important to differentiate from other tumors containing mature adipocytes and hematopoietic elements.

CONCLUSION:

This study highlighted the spectrum of soft tissue tumors presented in retroperitoneum. Malignant tumors were common than benign tumors and sarcomas were the most common tumor in retroperitoneum , of which liposarcoma was the predominant one and fibrosarcoma was the rarest tumor. Extra adrenal myelolipoma and neuroblastoma were rare in this site. Myeloipoma should be differentiated from tumors containing mature adipocytes and hematopoietic elements and neuroblastoma from Ewings sarcoma. Retroperitoneal tumors can cause therapeutic challenges due to their rarity and late presentation. Improved diagnostic accuracy will enable potential curative treatment, there by reducing mortality.

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