Retroperitoneal Tumors: Types and Presentations

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Summary:

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Background *:The retroperitoneal tumors is usually confined to lesions arising from tissues (muscles, fat, lymph nodes, nerves, and, developmental remnant) of this compartment but excluding origin from the retroperitoneal organs (panaceas, kidney, ureters and adrenals). The aim of the study is to focus a light on the types of retroperitoneal tumors, ways of the investigations and the presentations.*
Methods *:A prospective review study of 25 patients with retroperitoneal tumors has been collected in the Medical City Teaching Hospitals During the period between Jan 2001 to Mar 2004. The data included age, gender, risks factors, clinical presentations, diagnostic modalities and results of histopathology.*
Results *:14 males (56%) and 11 females (44%_ patients. The highest incidence of patients were in the (51-60 years) age group (10/25, 40%). The most common presenting feature was abdominal mass (20/25, 80%). u/s and CT scan were the most reliable investigations in the diagnosis. The most common tumors were sarcomas (15/25, 60%) and lymphomas (5/25, 20%).*
Conclusion *:Although retroperitoneal neoplasms account for 0.1 - 0.2% of all malignancies [5], it seems that from our findings most of the tumors were malignant, where the sarcoma represents the common tumor among them, the presentations and the diagnostic modalities especially u/s and CT have a similarity with most of the studies reviewed.*
Key words *:Retroperitoneal tumors, types, presentations.*

Introduction:

The retroperitoneum consist of that portion of the body which is bounded anteriorly by the peritoneum. Posteriorly by the spine, psoas and quadrtus lumborum muscles. Superiorly by the twelfth ribs and attachment of the diaphragm. Inferiorly by the brim of the pelvis. The lateral margins of the space correspond to the lateral borders of qudratus lumborum muscle(1). The retroperitoneum contains a number of vascular and visceral structures in the form of gastrointestinal, genitourinary, musculoskeletal, nervous and lymphatic tissues (2). The multiplicity of structures within the retroperitoneum gives rise to a variety of pathological conditions such as primary tumor of the retroperitoneal space and other diseases (non tumor) e.g. retroperitoneal hematomas & abscesses(3)

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The term retroperitoneal tumor is usually confined to lesions arising from tissues (muscles, fat, fiubrous tissue, lymph nodes, nerves and developmental remnants) of this compartment but excluding origin from the retroperitoneal organs (pancreas, kidneys, ureters and adrenals)(4). Patients may present initially with history of enlarging mass in the abdomen, vague abdominal discomfort or sense of heaviness or fullness. Pain may become severe if compression of adjacent nerves or nerve plexuses occurs(5). Ultrasound is the first noninvasive method for diagnosis; it shows the mass and its location. It was not reliable in identifying the origin and the anatomical relation of the mass in which CT scan excelled (6,7).

Primary retroperitoneal neoplasms account for 0.1 - 0.2% of all malignancies (8), and 10% Of all soft tissue sarcomas (9), in another study in USA 17%(5).

Retroperitoneal sarcomas are the most common malignant lesions of the retroperitoneum, among these liposarcomas is the most frequent (10). Benign retroperitoneal masses can be removed and cured by simple excision. Malignant lesions such as sarcomas, require more extensive dissection (11,12).

Next to sarcomas, lymphoma is the second most common malignant retroperitoneal neoplasm. Although primary retroperitoneal lymphoma is rare, if it occurs it will mostly of non-Hodgkin's lymphoma. Diagnosis is confirmed preferably by open biopsy after which chemotherapy can be given (13).

Regarding retroperitoneal cyst, true retroperitoneal cyst are rare (14). retroperitoneal pancreatic and renal cyst are much more common but they do not fall within the defmition of a true retroperitoneal cyst which does not arise from or in relation to a major retroperitoneal organ (15). True cysts include mesothelial, dermoid and hydatid cysts(15,16). There treatment varied from total excision if technically feasible to marsupialization in difficult cases with intimate contact to vital organs(17).

Patients and Methods:

This is a prospective review study of 25 patients with retroperitoneal masses carried out at Medical City Teaching Hospitals during the period between January 2001 and March 2004. the gathered information included: age, sex, risk factors, clinical presentations, diagnostic modalities, , results of histopathology, adjuvant therapy , These data were collected and analyzed and the results were shown in the form of figures and tables.

Diagnosis was achieved by complete medical evaluation with history and physical examination, ultrasound and CT scan with oral and intravenous contrast to determine the exact nature of the mass and its relation to the near by organs, to exclude primary condition of the gastrointestinal and urinary tracts and to assess the function of the kidneys in case ipsilateral nephrectomy will be needed to achieve complete clearance. Among patients with retroperitoneal tumors we exclude the masses which are related to the kidneys, adrenals, pancreas, bowel and urinary tract. retroperitoneal abscesses and hematomas were also excluded. Concentration was made only on malignant retroperitoneal tumors namely nonmetastatic sarcomas and lymphomas and the true retroperitoneal cyst.

Results:

Regarding the age of our patients' sample, Table 1 shows that the cases were organized in 10 years cohort groups. The majority of cases were found in the (51 - 60 years) age group in which we have 10 patients (10/25, 40%) with solid tumors all of them had retroperitoneal sarcomas proved later on by histopathological study.

Benign cystic lesions pathology was found in patients whom their age is less than 50 years with a mean age of 28 years. Lymphomas were scattered from 40 years to more than 70 years age groups, with a mean age of 60 years.

Concerning the sex distribution of our patients sample, figure 1 shows that we had 14 male patients (14/25, 56%) while the remaining 11 patients (11/25, 44%) were females.

In respect to the association of certain types of retroperitoneal tumors namely sarcomas with specific risk factors, figure 2 clarified that we found history of ionizing radiation in one of the patients with retroperitoneal sarcomas (1/15,6.7%) and the presence of neurofibromatosis in another one (1/15,6.7%).

Table 1 : Age distribution

	Lipoma (No.)	%	Cyst (No.)	%	Lymphoma (No.)	%	Sarcoma (No.)	Age (years)
4%	1					4%	1	<21
		4%	1					21-30
		8%	2	4%	1			31-40
		4%	1	4%	1	8%	2	41-50
						40	10	51-60
				4%	2	4%	1	61-70
				4%	1	4%	1	>70

Table 2 verifies the different clinical features by which our patients were presented. It is shown that the majority of cases (20/25,80%) were presented with abdominal mass which was felt anteriorly, in either flanks or could be felt per rectal examination in pelvic masses (7/25,28%). In addition, some of the cases were presented with the pressure effect of the masses causing gastrointestinal obstructive symptoms in (10/25,40%) of cases in the form of vomiting and constipation, urinary obstructive symptoms in (6/25,24%) in term of recurrent urinary colic and infection, and vascular obstructive symptoms in (8/25,32%) of cases in the form of leg edema because of inferior venal caval obstruction. Features of anemia, weight loss, fever and night sweat was noted in(5/25,.20%) of cases proved later on to have lymphoma.

Our patients were investigated by full blood count, plain abdominal X-ray, abdominal and pelvic ultrasound and CT scan.

Table 2 Clinical resentations

	No.	Clinical features
80%	20	Abdominal mass
40%	10	Gastrointestinal obstructive symptoms
24%	6	Urinary obstructive symptoms
32%	8	Vascular obstructive symptoms
20%	5	Fever, weight loss, sweat and pallor
28%	7	The per rectal examination

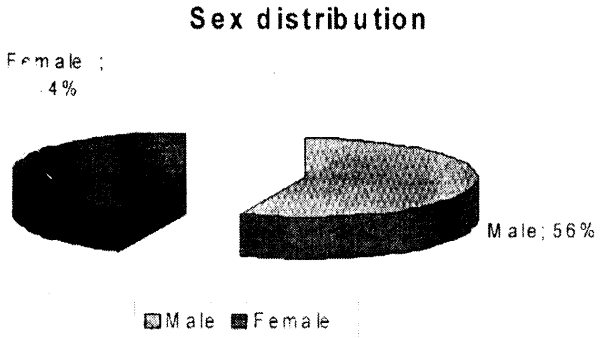
Table 3 shows that hemoglobin level that 10 gm/dl together with leukocytosis more than 20xlO9/L were found in 4 of our cases (4/25,16%) proved later to have lymphoma.

Plain abdominal X-ray showed soft tissue shadow in (8/25,32%) of cases, and calcification in two cases (2/25,8%) proved later on to have sarcoma. Ultrasound examination diagnosed (22/25,88%) of cases as having solid

retroperitoneal masses, the remaining (3/25,12%) were diagnosed with retroperitoneal cystic lesions. Hydronephrosis was in (6/25, 24%) of cases.

CT scan with oral and intravenous contrast showed more detailed informations and more diagnostic accuracy in form of diagnosing the exact nature of the retroperitoneal masses in our patients sample, being solid in (15/25,60%), cystic in (4/25,16%) and solid with necrotic areas in (6/25,24%) proved later on by histopathological study as a high grade sarcomas.

Fig. 1 Sex distribution Male : 56% 14/25 Female : 44% 11/25



In addition, CT scan gave an idea about the structures which were composed by the tumor, being gastrointestinal in 10 cases (10/25, 40%), urinary tract in 6 cases (6/25,24%) and inferior vena cava compression in 8 cases (8/25,32%).
Fine needle aspiration was used in two cases suspected to have lymphoma and proved to be inconclusive, a situation which necessitated open surgical biopsy which proved lymphoma. In the rest of suspected lymphoma cases fine needle aspiration was not done.

Table 3 Diagnostic Modalities

%	No.	Diagnostic Methods and its results
		Haematological
16%	4	Hb < 10 gm/dl
16%	4	WBC > 20 x 109
		Plain abdominal X- Ray
32%	8	Soft tissue shadow
8%	2	Calcification
		ultrasound
8	22	Solid lesion
12%	3	Cystic lesion
24%	6	Hydronephrosis
		CT scan
60%	15	Solid lesion
16%	4	Cystic lesion
24%	6	Solid with necrotic areas
32%	8	Gastrointestinal compression
24%	6	Urinary tract compression
32%	8	IVC compression
8%	2	FNAC (Inconclusive findings)

Table 4 classified the patients sample according to the postoperative histopathological results of the resected samples. Thus it was found that sarcomas one (1/5,20%) had Hodgkin lymphoma. The above were present . in (15/25,60%), most of them results concern the malignant lesions, regarding (8/15,58%) were liposarcomas. Lymphomas were benign lesions they were in the form of found in (5/25,20%), four of them (4/5,80%) were retroperitoneal benign cystic mass in (4/25,16%) having non-Hodgkin lymphomas and the remaining and retroperitoneal lipoma in (1/25,4%).

Table 4: Postoperative histopathological diagnosis

%	No.	Diagnosis
32%	8	Liposarcoma
16%	4	NH Lymphoma
4%	1	H L
12%	3	Fibro sarcoma
8%	2	Rhabdomyosarcoma
8%	2	Malignant fibrous histiocytoma
12%	3	Dermoid cyst
4%	1	Hydatid cyst
4%	1	Lipoma
10%	25	Total

Discussion

The data regarding 25 cases presenting with retroperitoneal masses were gathered, reviewed and analyzed and it was found that: Regarding the age of the patients and its correlation to the postoperative histopathological results, most of the retroperitoneal sarcomas (10/15,66.7%) were found in the (51 - 60 years) age group, benign cystic lesions were discovered below the age of 50 years with mean age of 28 years. Lymphomas were scattered from 40 to more that 70 years with a mean age of 60 years. These results are in accord with what was found by:

1 Roberto I. et al(18) who stated that retroperitoneal sarcomas are most prevalent from the fifth to the seventh decade of life.

2 El-Bahar TM et al(17) who found that patients with retroperitoneal cystic swelling have a mean age of 30 years.

3 Robert LH et al(13) who documented that patients with retroperitoneal lymphomas have a mean age of 61 years.

In respect to the sex of the patients (14/25,56%) were males and (11/25,44%) were females with no significant preponderance of the male population. Concerning the risk factors for the development of retroperitoneal masses history of ionizing irradiation was present in one patient proved later on to have retroperitoneal liposarcoma, another patient had a history of

neurofibromatosis and he suffered from neurofbrosarcoma. These results goes with that of Davidson T et al(19) and Lt Col G Rajgopal(9) who identified ionizing irradiation and genetic transmitted diseases such as neurofibromatosis as risk factors for the development of retroperitoneal sarcomas. In the clinical presentation of our patients, the mass attained considerable size before being discovered, and this can be attributed to the fact that the great dimensions, loose boundaries and flimsy areolar tissue of the retroperitoneal space in addition to the indolent nature of the tumors, all of these factors will enable tumors in this location to attain a very large size before any symptoms occur and if they do so they will be non-specific or too late. The most prevalent symptoms was abdominal mass in (20/25,80%), gastrointestinal and urinary obstructive symptoms were found in (10/25,40%) and (6/25,24%) of patients respectively. These findings concede with:

1 John M et a1(5) who concluded that late presentation as a feature of retroperitoneal tumors is due to their presence in a potentially growing space.

2 Cuschieri A.(4) Who concluded that the majority of retroperitoneal tumors are presented as abdominal mass.

3 Lt Col G Rajgopal(9) who verified that retroperitoneal masses often present as asymptomatic abdominal mass, generally they do not cause any until they are quite large and produce pressure or traction on adjacent nerves and muscles. 4 El Bahar TM et al(17) who found that retroperitoneal masses can be presented with gastrointestinal obstructive symptoms in 42.8% or urinary obstructive symptoms in 28.57%. Concerning the diagnostic modalities by which our patients were investigated, full blood count showing hemoglobin <10gm/dl and leukocytosis >20xI09/L was found in (4/25,16%) of cases proved later on to have lymphoma. These results with those Robert LH et al(13) who found that some cases with hematological malignancy may have primary retroperitoneal presentation.

In the plain abdominal X-ray finding, one peculiar finding was calcification within the soft tissue mass noted in (2/25,8%) of cases proved later on to have sarcoma. These results are in accord with that of Ferrero DR et al(20) who demonstrated calcification in retroperitoneal liposarcoma.

In the abdominal ultrasound findings cystic lesions were differentiated from solid lesions and were found in 3 patients out of the 4 with retroperitoneal cyst (75%), a percentage which escalates to reach 100% in the diagnosis of retroperitoneal cystic lesion by CT scan. These results goes with that of El-Bahar TM et al(17) who found that ultrasound was diagnostic -in 85% of retroperitoneal cystic lesion while CT scan was diagnostic in 100%. In addition CT scan added more diagnostic accuracy to the solid lesions in form of providing informations about its exact nature and whether it contains necrotic soft tissue, this was found in (6/25,24%) of cases proved later on to have high grade sarcomas, these results are in agreement with that of Suzuki S et al (21) who stated that the composition of malignant mesenchymal tumor whether or not it contain necrotic soft tissue seems to determine its exact nature.

Also CT scan with oral and intravenous enhancement provided informations about the involvement of the adjacent organs and the function of both kidneys so as to put in mind the surgical plan needed to achieve complete clearance as there is a high chance of contiguous organ resection to achieve that goal. This results goes with what was stated by John MD et a$5) "intravenous pylogram to check the kidneys and bowel preparation is mandatory before surgery for retroperitoneal neoplasm as dissection of these organs with the tumor may be needed for complete clearance". In respect to fine needle aspiration, it was not used as diagnostic modalities in cystic lesions, as it will not provide informations about the exact nature of the cyst and it could cause hazardous infra-cystic bleeding (17).

Regarding solid lesions, fine needle aspiration was only used in suspected cases of lymphoma because, its used in sarcomatous lesions will not provide enough informations abut degree of differentiation, cellularity, necrosis and number of mitotic figures per high power field which need open surgical biopsy and are very important for the discrimination of high and low grade lesions and thus for determining the prognosis of the disease and planning for further postoperative adjuvant treatment(9).

In the the suspected cases of lymphoma fine needle aspiration was feasible in 2 cases only (2/25,8%), in the other cases it was not technically possible because of the fear of jeopardizing nearby vital organs with injury rendering the procedure hazardous to be undertaken. The results of fine needle aspiration as they are not coupled with other advanced accessory diagnostic modalities such as flow cytometry and immune histochemistry were inconclusive in both cases mandating open surgical biopsy. These are results are in accord with that of Robert LH et al(13) who found that "although open lymph node biopsy is preferred method for diagnosis of retroperitoneal lymphoma, fine needle aspiration plays a

significant role in this setting and coupled with other informations such as flow cytometry and immune histochemistry, it is considered a practical and reliable method ". Although the (FNA) has not achieved its full potential as a diagnostic modality in the diagnosis of retroperitoneal tumors, it has been concluded in certain study that FNA is the procedure of choice for the detecting most malignancies in this location(22) .

As there are no means in the time being that are more reliable and less invasive than surgery that can definitely distinguish benign from malignant soft tissue lesions, all of our patients were submitted to surgery by which the lesion was totally excised in (13/25,52%) of cases. These results concede with that of Lt Col G Rajgopal(9) who found that ""As there are no reliable signs and symptoms that distinguish benign from malignant soft tissue tumors, it is imperative to biopsy all soft tissue lesions".

In respect to the results of postoperative histopathological study, it was demonstrated that liposarcoma and lymphoma constituted most of the malignant of retroperitoneal lesions (13/20,65%). These results concede with that of John MD et al(5) who stated that "retroperitoneal sarcomas including mainly liposarcoma, and lymphoma are the most common malignant retroperitoneal soft tissue tumors". Non-Hodgkin lymphoma was found in 4 of the five lymphoma case (4/5,80%), and the last one was having Hodgkin lymphoma (1/5,20%). These results are close to those of Robert LH et ah13) who demonstrated non-Hodgkin lymphoma in 90% and Hodgkin lymphoma in 10% of cases with retroperitoneal presentation of hematological malignancy.

Retroperitoneal cysts were found in (4/25,16%) of cases and this concedes with what was found by Walker AR and Putnam TC(14) who conducted in Weir siuuy 01 .3.3 cases inat uue retroperlioneai cysts are rare.In our study one of four cysts was hydatid cyst and it is well known that Iraq is an endemic area so one might expect to find hydatid cyst anywhere in the body including the retroperitoneal space although it is one of the uncommon site of the hydatid disease

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