Primary Retroperitoneal Mass--- A CT Overview with FNAC /Histopathological Correlation

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Abstract
Describe and characterize specific CT imaging finding in varieties of Primary Retroperitoneal masses. To confirm the diagnosis of CT by FNAC / histopathological correlation.

Material & Method: A hospital based cross sectional study was conducted over a period of 1 year between 1st July 2012 to June 0th 2013 on 67 patients in the deptt. of Radio-Diagnosis, Assam Medical College and Hospital, Dibrugarh. Approved was obtained from the Institutional ethics review committees. All the patients CT on of abdomen are done both initial unenhanced CT followed by contrast CT scan by giving parenterily iodinated low osmolar non ionic contrast media. Delayed scan were also obtain wherever necessary. Based on the findings observed on the CT Scan, a probable diagnosis was given, which was confirmed by Fine Needle Aspiration Cytology (FNAC) or by Histopathological examination (HPE) of the operative specimens.

Result: Primary Retroperitoneal tumours accounted for 40.3% of the masses and secondary retroperitoneal masses (arising from retroperitoneal organs) accounted for 59.7% in this studies. Most of the primary retroperitoneal neoplasm were malignant accounting 78%. 22% of the masses were benign. Among the primary retroperitoneal masses lymphoma accounted 38% followed by liposarcoma 19% and leiomyosarcoma 14%. nodes (100%). Among the retroperitoneal soft tissue sarcomas, liposarcoma 33.4% was commonest followed by leiomyosarcoma 25% and malignant fibrous Histiocytoan (16.7%).

Conclusion: CT with its fast scanning times and good spatial resolution and multiplanar reconstruction capability is a better imaging modality for characterizing retroperitoneal masses and detecting the organ of origin. Most of the CT diagnoses were correlated with FNAC/HPE diagnosis.

Key words: CT, Retroperitoneal masses, FNAC, HPE.

INTRODUCTION
The retroperitoneon is the compartmentalized space located external to and predominantly posterior to the posterior parietal peritoneum. About half a century ago, the retroperitoneon was described as a “veritable jungle of strange things” and as a hinterland of straggling mesenchyme, with vascular and nervous plexus, weird embryonic rests and shadowy fascial boundaries.¹ The mass lesions in retroperitoneal space may present with clinical features like vague pain in the lumbar region, abdominal lump and deep vein thrombosis of lower limbs. Of the primary retroperitoneal neoplasms, 70-80% are malignant.
in nature, and these account for 0.1-0.02% of all malignancies in the body. Because the treatment options vary, it is useful to be able to noninvasively distinguish these masses.3

Because of its usual abundant fat relatively uncomplicated cross-sectional anatomy, the retroperitoneal space is especially suitable for examinations by Computed Tomography (CT).4 Contrast Enhanced Computed Tomography (CECT) is considered the best imaging modality for the retroperitoneal masses. CT is the most widely used cross-sectional imaging modality to assess the retroperitoneum.1 Although CT findings will not always yield a definitive diagnosis attention to these finding provides a road map to guide image interpretation.6 CT-guided Fine Needle Aspiration Cytology (FNAC) allows visualization of entire abdomen and retroperitoneum, and allows accurate planning of a biopsy for deep seated lesions such as retroperitoneal lymphadenopathy and it avoids damage to important organs and major vessels.7

MATERIALS AND METHODS

THE STUDY GROUP

A hospital based cross sectional study was conducted over a period of 1 year between July 1st 2012 to June 30th 2013 on 67 patients in the department of Radiodiagnosis, Assam Medical College and Hospital, Dibrugarh, Approval was obtained from the Institution ethics review committee.

INCLUSION CRITERIA

1. Clinically suspected patients presenting with symptoms of involvement of retroperitoneal structures, in all age groups.
2. Involvement of retroperitoneal organs detected incidentally by routine Ultrasonography of the abdomen / referred patients.

EXCLUSION CRITERIA

1. Patients with traumatic, congenital and inflammatory masses.
2. Retroperitoneal metastases from primary in another organ.
3. Primary bowel masses.
4. FNAC was not done in patients with history of bleeding diathesis and in those not giving consent.

PATIENT PREPARATION

Patients were kept nil per oral for 6-8 hours preceding the examination except plain water. Dilute oral contrast agent (20% Urograffin) was administered 45 minutes before the study. Risks of contrast administration were explained to the patient and informed consent was taken prior to the administration of i. v. contrast medium.

TECHNIQUE OF THE STUDY

The CT scanner used in this was SIEMENS Somatom Spirit Dual Slice Ct Scanner. Patients were scanned in a supine position. Initial unenhanced scans were obtained in all patients. Contrast studies were performed by manually injecting Iodinated low osmolar non ionic monomer given as a single bolus injection at a maximum dose of 2 ml/kg body weight. Scanning parameters used were spiral mode with slice thickness of 6mm and collimation 6 x 2.5 mm, pitch: 1.4; kVp: 130; mAs: 80. Field of view from top of diaphragm till symphysis pubis. Delayed scans were obtained with the same parameters wherever necessary.

Based on the findings observed on CT scan, probable diagnosis was given, which was confirmed either by Fine Needle Aspiration Cytology( FNAC) or by Histopathologic Examination (HPE) of the operative specimen, which was considered as the Gold standard. The areas of interest in the study were organ of origin, presence of fat, myxoid stroma, necrosis/cystic changes, calcification, post contrast enhancement, extension of mass and invasion into adjacent
structures, vessel infiltration, nodal status and distant metastases.

**FINDINGS ON CT**

Each patient was evaluated on CT as per the protocol mentioned below:

- **Location and organ of origin**: Primary retroperitoneal/ kidney/ Adrenal gland/ pancreas.
- **Appearance of the mass**: fat, myxoid, necrosis/ cystic changes, hemorrhage, calcification, etc.
- **Post contrast enhancement**: none/ mild/ moderate/ intense
- **Infiltration into vessels and adjacent structures**:
- **Nodal and distant metastases**:
- **Presence of Ascites**:
- **Other Routine Investigations**:
- **FNAC/HPE results**:

**TECHNIQUE OF FNAC**

After taking informed consent from the patient, following skin cleansing with Betadine (Povidone-Iodine) solution, USG/CT- guided FNAC was done using 20 G 88mm Lumbar-Puncture needle fitted to 20-mL disposable syringe. Aspiration was performed by back-and-forth movements of the needle under continuous negative pressure. Immediately after FNAC, the percutaneous site was sealed using Betadine swab. All patients were observed for at least thirty minutes. If no complications were observed, the patient was allowed to leave.

Cytologic stains used were May-Grunwald-Giemsa (MGG), Papanicolaou (PAP) stain and other stains, whichever applicable. And for histology, Hematoxylin and Eosin (H & E) stains were used. 4 to 8 smears including air dried and alcohol fixed smears were prepared in each case from the aspirated material. Alcohol fixed smears were stained with PAP stain, while air-dried smears were stained with MGG.

**Statistical Analysis**

The diagnosis accuracy was determined using the following formula:

\[
\text{Diagnostic Accuracy} = \frac{\text{True Positive} + \text{True Negative}}{\text{Total Number of Cases}} \times 100
\]

**Discussion**

Many comprehensive studies have been carried out regarding the imaging features of retroperitoneal masses. We evaluated 67 cases of Retroperitoneal masses for a period of one year using Computer Tomography, the characteristics of which are discussed below.

**PRIMARY RETROPERITONEAL TUMOURS**

In the present study, we had 27 patients with primary RTPs, of which 11 (41%) were males and 16 (59%) were females, with a Male: Female (M: F) ratio of 0.69, showing a female predominance. Age at presentation ranged from 18 days to 76 years with a mean age of 43.8 years. 42% of the Retroperitoneal Soft Tissue Sarcomas (RSTS) occurred in males and 58% in female with a M: F ratio of 0.72. Our study correlated well with the study conducted by Thijs van Dalen, et al (2001) which showed a M: F ratio of 0.73 for RSTS.

In the present study, 21 (78%) of the primary RPTs were malignant and 6 (22%) were benign. Lymphoma was the most common (38%; n=8) primary malignant retroperitoneal tumour, followed by other RSTS. Liposarcoma 4 (33.4%) was the most common RSTS, followed by Leiomyosarcoma 3 (25%), MFH 2 (16.7%), Angiosarcoma 1 (8.3%), Chondrosarcoma 1 (8.3%) and Synovial cell sarcoma 1 (8.3%).

It correlated well with the study conducted by Neville A, et al (2004) in which 70%-80% of RPTs were malignant in nature and Lymphoma was the most common (33%) Liposarcoma (33%) was the most common RSTS, followed by Leiomyosarcoma (28%) and MFH (19%).
Thijs van Dalen, et al (2001) studies 706 patients with primary RPTs of which 80% were malignant and 20% were benign. Our study also correlated well with the study conducted by Ingo Aldinger, et al (2006) in which they studies 117 patients with RSTS of which Liposarcoma (26.5%) was the most common, followed by Leiomyosarcoma (19.6%) and MFH (15.4%) other histological entities were rare. In the present study, we had 6 (22%) benign retroperitoneal tumours. 2 teratomas, 1 FIF, 1 schwannoma, 1 Ganglioneuroblastoma and 1 paraganglioma. Hayasaka K, et al (1994) studies 21 primary benign RPTs including 9 teratomas, 6 schwannomas, 3 leiomyoma, 1 lymphangioma and 1 neurofibroma. The present study comprised 14 paediatric RPTs, among which Neuroblastoma 5 (35.7%) was the most common, followed by Wilms’ tumour 4 (28.6%) and teratoma 2 (14.3%). We had 1 case each of Clear cell sarcoma, Ganglioneuroblastoma and FIF. However it was in sharp contrast to the study conducted by Muhammad Sharif, et al (2007) which showed Wilms’ tumours (50%) as the most common, followed by Neuroblastoma (26.6%) and Teratoma (16.6%).

Lymphoma
In the present study, we had 8 cases of Lymphoma, and all of them were Non Hodgkin’s type. CT could diagnose all the 8 cases with 100% accuracy. Lymphoma accounted for 38% of the primary malignant retroperitoneal tumours in our study. The age group ranged from 42 to 70 years with a mean age of 56.62 years. We saw a male predominance with a M: F ratio of 1: 3. Two of them were well differentiated (50%), 1 myxoid (25%) and 1 dedifferentiated (25%) type. On CT scan, the well differentiated types revealed large lobulated predominant fat tissue attenuation mass in the retroperitineum with very minimal soft tissue component, and showed minimal post contrast enhancement. The myxoid variety showed mixed areas of fat, soft tissue and myxoid attenuation with secondary infiltration of kidney. Our study correlated well with the study conducted by Dieckmann C, et al (1997) in which the highest percentage of fat and the biggest tumours were seen with well-differentiated liposarcomas, whereas myxoid ones showed the sharpest margins. Round-cell and pleomorphic types showed mainly soft-tissue attenuation. Sun Hwa Hong, et al (2010) retrospectively studied 15 patients with histological verified retroperitoneal dedifferentiated liposarcoma in associated vessel infiltration and moderate heterogeneous enhancement on contrast studies. Ascites was noted in 4 (50%) cases, necrosis in 3 (37%), liver involvement in 2 and bowel involvement in 1 patient. Our study correlated well with the study conducted by Joseph k. T. Lee, et al (1978) and Zhi-gang Yang, et al (1999) both of which reported a predominance of lower para-aortic group of lymphadenopathy involvement with bulky low attenuating homogenous enhancing pattern of lymph nodes. Joseph k. T. Lee, et al (1978) reported an accuracy of 90% of CT in staging lymphoma patients.

Liposarcoma
In the present study, we had 4 cases of liposarcoma accounting for 33.4% of all soft tissue sarcomas, and all 4 were diagnosed by CT with 100% accuracy. The age group ranged from 39 to 70 years with a mean age of 56.2 years. We saw a female predominance with a M: F ratio of 1: 3. Two of them were well differentiated (50%), 1 myxoid (25%) and 1 dedifferentiated (25%) type. On CT scan, the well differentiated types revealed large lobulated predominant fat tissue attenuation mass in the retroperitineum with very minimal soft tissue component, and showed minimal post contrast enhancement. The myxoid variety showed mixed areas of fat, soft tissue and myxoid components, with coarse calcifications. The dedifferentiated type showed areas fat attenuation and enhancing soft tissue components with secondary infiltration of kidney. Our study correlated well with the study conducted by Dieckmann C, et al (1997) in which the highest percentage of fat and the biggest tumours were seen with well-differentiated liposarcomas, whereas myxoid ones showed the sharpest margins. Round-cell and pleomorphic types showed mainly soft-tissue attenuation.
which CT showed well-circumscribed, large round, or lobulated retroperitoneal mass with various spectra of imaging findings.

**Leiomyosarcoma**
The present study included 3 cases of leiomyosarcoma, accounting for 25% of soft tissue sarcomas. Two of them were males aged 70 and 76 years and a female patient aged 45 years. Two of them were completely extravascular (66.6%) and one was both intra and extravascular (33.3%). On CT, they showed large lobulated retroperitoneal muscle density masses with large non enhancing areas of necrosis. They had ill defined margins with infiltration of the adjacent structures. None of them showed fat or calcifications, however myxoid components was seen in two of them. Distant metastases were noted to the liver in 1 case and to the lungs in another.

We had a case of inferior vena caval Leiomyosarcoma in a 76 year old male patient, which involve intra-hepatic and supra-renal IVC and had imaging pattern similar to the other cases of extra-vascular leiomyosarcoma.

Our study correlated well with David S. Hartmen, et al (1992), in which 62% cases were completely extravascular, 33% both extra- and intraluminal and 5% completely intraluminal. On CT, it is usually solid with large conspicuous cystic zones corresponding to areas of necrosis.

**Malignant Fibrous Histocytoma**
The present study included 2 cases of MFH, accounting to 16.7% of soft tissue sarcomas, one of them a 49 year old male and the other a 35 year female patient; the former was wrongly diagnosed as myxoid variety of Liposarcoma on CT scan, but on HPE turned out to be Pleomorphic Malignant Fibrous Histocytoma. CT scan revealed large soft tissue masses in the retroperitoneum, with predominat low attenuation myxoid components and showed moderate post contrast enhancement. Out of them contained areas of fat attenuation, but neither of them showed necrosis of calcifications. Our study correlated well with Bivek Karki, et al (2012). Primary retroperitoneal MFHs are generally hugs soft tissue masses containing areas of low attenuation and mild to moderate contrast enhancement.

**Sympathetic plexus Neuroblastoma**
The present study include a case of Sympathetic plexus neuroblastoma in a 2 year female child. CT scan revealed a large ill defined prevertebral low attenuation hypo-enhancing soft tissue mass with areas of coarse calcifications and necrosis, which encased the aorta, celiac, mesenteric and renal vessels without definite infiltration.

It correlated well with the study of Ruppert David, et al (1989). Neuroblastoma is a common tumour in childhood. It arises in the Adrenal gland or in various extra-adrenal primary sites of the sympathetic chain. Calcification is seen in 85% of cases. Most lesions are homogeneous on CT, with attenuation similar to or less than that of muscle.

**Ganglioneuroblastoma**
The present study included a case of Ganglioneuroblastoma in a 3 year male child. CT scan revealed an ill defined heterogeneously enhancing prevertebral soft tissue mass with areas of coarse calcifications within. Post operative HPE revealed Ganglioneuroblastoma.

It correlated well with Sung Eun Rha, et al (2003). Ganglioneuroblastoma is most often seen in patients 2-4 years old. It also correlated well with KK Sabharwal, et al (2006). CT examination revealed a large lobulated heterogeneously enhancing mass with smooth margins, Enhancing septae and amorphous calcific foci were also seen within the mass. Trucut biopsy revealed Ganglioneuroblastoma.
Retroperitoneal paraganglioma
The present study included a case of retroperitoneal paraganglioma in a 48 year female patient who presented with hypertension. CT showed a large is to slightly hyperdense mass lesion in the right upper retroperitoneal along the lower abdominal aorta with central cystic changes, displacing the IVC posteriorly. Moderate to intense enhancement was noted on post contrast study.
In correlated well with the study conducted by W S Hayes, et al (1990).47 Average age was 37 years. 80% had hypertension. Extra-adrenal retroperitoneal paragangliomas are functionally active more often and they are readily detected by CT as soft-tissue masses closely associated with the entire length of the abdominal aorta.

Retroperitoneal Schwannoma
A 35 year female patient presented with vague lump in right iliac fossa. CT scan revealed a large, well circumscribed hypodense mild heterogeneously enhancing SOL in the right lower retroperitoneum with a focus of calcification, causing antero-medial displacement of the ureter and common iliac vein. A diagnosis of benign peripheral nerve sheath tumour was given and post operative HPE revealed benign schwannoma. It correlated well with Ali Kamalati, et al (2013)48 who reported a case of benign schwannoma in a 26-year-old woman. On CT scan, it was hypodense and showed heterogeneous enhancement following contrast injection.
Li Qiang, et al (2007)49 analysed 82 patients with retroperitoneal schwannoma. 46% were men and 54% were women. Only in 13 patients (15.9%) a correct preoperative diagnosis was made either by USG, CT or MRI. Pathological result showed 98.8% were benign and 1.2% malignant.

Primary Germ Cell Tumours
The present study had 2 cases of mature teratomas. Both were female aged 6 years and 15 years. On CT, both cases showed huge retroperitoneal masses with large cystic spaces, areas of fat attenuation and coarse calcifications, one of them had multiple tooth within. Both of them did not show any enhancement on post contrast scans.

Retroperitoneal Angiosarcoma
The present study had 1 case of Retroperitoneal Angiosarcoma in a 45 year female patient. CT revealed a large mass in the right anterior pararenal space with variable soft tissue components, necrosis, hemorrhage and coarse calcifications. The lesion was infiltrating into the adjacent liver segments. On post contrast scans, it showed moderate to intense enhancement.
Our finding correlated with Changhoon Yoo, et al (2009).52

Retroperitoneal Chondrosarcoma:
The present study included a case of Retroperitoneal Chondrosarcoma in a 65 year old female patient. CT scan revealed a large relatively well defined soft tissue mass in the retroperitoneal space with extensive linear and arc-like streaks of calcifications. The mass showed only mild enhancement on post contrast study. FNAC suggested Extra skeletal Mesenchymal Chondrosarcoma.

Retroperitoneal Synovial cell sarcoma
A 50 year male patient presented with painless lump in the right lumbar region. CT scan revealed a large ill defined mass in the right side of retroperitoneal with infiltration of IVC and compression of right ureter. There were large areas of cystic changes, necrosis and few calcifications within, with mild to moderate heterogeneous enhancement on post contrast study. A CT diagnosis of Leiomyosarcoma was given. However, FNAC revealed it as synovial cell sarcoma.
A.R Alhazzani, et al (2010)54 described Primary retroperitoneal synovial sarcoma as extremely rare and has poor prognosis.
Retroperitoneal fetus-in-fetu:
An 18 day old girl child had one malformed fetus in her abdomen. A CT scan of the abdomen showed a large well-defined round retroperitoneal soft-tissue mass with relatively well-formed bony components resembling those of a small fetus with a well-defined peripheral cystic covering. Elective laparotomy anencephalic fetus was found. Akhtar Murtaza, et al (2010) described a case of FIF in a 30 year female. CT scan of the abdomen showed a cystic lesion with solid component of varying densities which was initially diagnosed as retroperitoneal dermoid.

Primary retroperitoneal masses are generally clinically occult until they assume a large size and invade surrounding structures. CT with its fast scanning times and good spatial resolution and multiplanar reconstruction capabilities is a better imaging modality for characterizing retroperitoneal masses and detecting the organ of origin. In this study, we came across a wide variety of retroperitoneal masses both benign and malignant with few very rare cases such as fetus-in-fetu, most of which were correctly diagnosed on CT. Most of the CT diagnoses were correlated with FNAC/HPE diagnosis. Thus this study helped in showing the importance of CT as a clinical and evaluatory tool in the characterization and diagnosis of retroperitoneal masses and helped in better management of the patients.
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