International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Case of Recurrent Retroperitoneal Liposarcoma

Dr. M. V. Krishna Raj M. S.[1], G. Gopu M. S.[2]

[1]General Surgery, Madras Medical College, Chennai - 600003, Tamil Nadu, India

[2]Professor, M. Ch., (Surgical Oncology), Madras Medical College, Chennai - 600003, Tamil Nadu, India

Abstract: ***Retroperitoneal sarcomas (RPS) are a rare group of cancers whose management can be challenging due to various presentation patterns, multiple organ involvement, and a high local and distant recurrence rate. Retroperitoneal sarcomas comprise approximately 10% of all soft tissue sarcomas, out of which around two - thirds of retroperitoneal sarcomas are high grade (either grade 2 or 3). Though any histologic type of soft tissue sarcoma may arise in the retroperitoneal space, liposarcoma and leiomyosarcoma do so most commonly. Retroperitoneal sarcomas are diagnostically challenging, owing to their diversity and morphological overlap with other tumors arising in the retroperitoneum. The main imaging methods used for staging and surgical planning, as well as for selecting the biopsy site and guiding the biopsy procedure, are computed tomography and magnetic resonance imaging. In most cases, the treatment is challenging, because of the size of the lesions (70% are >10 cm at diagnosis), vascular involvement, or involvement of adjacent organs. Histopathology and prognostic factors analysis are essential to predict the behaviour of the disease and plan the best therapeutic strategy.***

Keywords: retroperitoneal sarcoma, liposarcoma, leiomyosarcoma, computed tomography, magnetic resonance imaging

1. Introduction

Retroperitoneal masses constitute a heterogeneous group of lesions, originating in the retroperitoneal spaces, that pose a diagnostic challenge for radiologists. Though benign lesions typically predominate over malignant lesions elsewhere in the body, malignant lesions of the retroperitoneum are roughly four times more frequent than benign lesions. Approximately 75% are mesenchymal in origin. Although such tumors are more prevalent in adults, they can occur at any age. When they do not originate from organs such as the kidneys, adrenal glands, pancreas, or bowel loops, retroperitoneal masses are classified as primary and are categorized as solid or cystic, depending on their appearance on imaging. Solid lesions can be divided into four groups, by origin: mesenchymal, neural, germ - cell, and lymphoproliferative. Among the cystic lesions, the most common are lymphangioma and cystic mesothelioma. There are also non - neoplastic processes, primarily retroperitoneal fibrosis, non - Langerhans histiocytosis (Erdheim - Chester disease), and extramedullary hematopoiesis.

The clinical manifestations of retroperitoneal masses are nonspecific, depending on their location and relation with the adjacent structures. They typically do not produce symptoms until they grow large enough to compress or invade contiguous structures, although pain, early satiety, and obstructive gastrointestinal symptoms may occur early in the disease course in some patients.

Soft tissue tumors have been classified predominantly based on the line of differentiation. The 2020 World Health Organization (WHO) classifies soft tissue tumors into 12 subtypes according to their lineage: (1) adipocytic tumors, (2) fibroblastic and myofibroblastic tumors, (3) fibrohistiocytic tumors, (4) vascular tumors, (5) pericytic (perivascular) tumors, (6) smooth muscle tumors, (7) skeletal muscle tumors, (8) gastrointestinal stromal tumors, (9) chondro - osseous tumors, (10) peripheral nerve sheath tumors, (11) tumors of uncertain differentiation, and (12) undifferentiated small round cell sarcomas.

To categorize a retroperitoneal mass as a primary retroperitoneal lesion, its origin from a larger retroperitoneal organ should be excluded. It should then be classified as solid or cystic, its main imaging characteristics (macroscopic fat, calcifications, myxoid stroma, necrosis, and cystic areas of vascularization) should be evaluated, and its relationship with adjacent structures should be described. There are radiological signs (the crescent sign, embedded organ sign, and phantom organ sign) that aid in the diagnostic assessment; the absence of those signs can confirm the categorization of a mass as a primary retroperitoneal lesion.

The diagnosis of retroperitoneal tumors is complicated by (1) a large number of tumor types, (2) morphological overlap between various tumor types, and (3) the increasing use of minimally invasive biopsy techniques with very limited tissue. An accurate diagnosis is crucial for correct management and prediction of prognosis.

Classification of Retroperitoneal Lesions:

Table 1:

	Retroperitoneal lesions in adults	
Category	Solid	Cystic
Neoplastic	Lymphoma Liposarcoma Malignant fibrous histiocytoma Leiomyosarcoma Neurogenic tumors Germ-cell tumors	Lymphangioma Cystadenoma Cystadenocarcinoma Cystic mesothelioma Mature teratoma
Non neoplastic	Retroperitoneal fibrosis Extramedullary hematopoiesis Erdheim-Chester disease	Epidermoid cyst Epidermoid cyst Non-pancreatic pseudocyst Bronchogenic cyst

Source: Mota MMDS, Bezerra ROF, Garcia MRT. Practical approach to primary retroperitoneal masses in adults. Radiol Bras.2018 Nov - Dec; 51 (6): 391 - 400.

2. Case Details

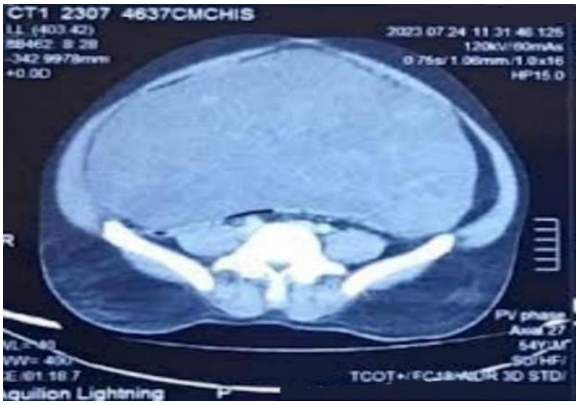
Chief Complaints and History of Presenting Illness:
This case report pertains to a 54 - year - old man, a known diabetic and hypertensive patient on regular medication, who was admitted with chief complaints of abdomen distention and pain for the last 3 months, with no other significant complaints pertaining to the abdomen or other systems. He had similar complaints 2 years prior, following which he was diagnosed with retroperitoneal liposarcoma, with involvement of right kidney, for which he had undergone Wide Local Excision (WLE) with Right Nephrectomy followed by adjuvant Radiotherapy (RT). He had normal bowel, bladder and sleep habits, and had negative history of consumption of alcohol, tobacco or other banned substances.

General and Systemic Examination:
The patient was conscious, oriented, afebrile and adequately hydrated. He was moderately built and well nourished. His vital signs were normal, with blood pressure coming under the pre - hypertension range (130/70 mm Hg). General examination findings were insignificant. Examination of the abdomen revealed a grossly distended abdomen with fullness of flanks. Palpation revealed an ill - defined mass palpable throughout the abdomen, which was not associated with warmth, tenderness and guarding of the abdominal wall. Dull note was present throughout the abdomen upon percussion, which was not associated with fluid thrills. No abnormal findings were found in the examination of the other systems.

3. Radiological Findings

CECT – Abdomen and KUB:
It showed a well - defined heterogeneously enhancing lobulated mass of size 27.4 X 28.6 X 16.4 cm within the peritoneal cavity, extending from L1 to sacral vertebral level, with internal areas of necrosis and calcification. Another similar lesion of size 11.2 X 13.5 X 8.9 cm was noted in the right renal fossa, posterolateral to the above - mentioned lesion, with internal areas of necrosis – which displaced the ascending colon anterolaterally and superior mesenteric artery & small bowel loops to the left. The lesion showed areas having contrast enhancement. The fat plane between the lesion & the head and body of pancreas was lost.

Impression: Large heterodense mass lesion in the retroperitoneum – indicating Liposarcoma.



CT Chest:
Visualized neck section showed heterogenous mass in the left lobe of the thyroid gland measuring about 2.3 X 2.7 cm.
Impression: No significant abnormality noted.

Histopathology Report (USG guided biopsy):
It showed fragments of neoplasm arranged in sheets, composed of elongated cells having moderate eosinophilic cytoplasm, dark staining pleomorphic spindle shaped nucleus. Stoma showed extensive areas of collagenization with focal areas of necrosis and hemorrhage. Tumour marker S100 tested negative, while testing for MDM2 showed scattered nuclear positivity.

Impression: Suggestive of Liposarcoma.

Surgical Removal of the Tumour:
The types of resections done are R0 (complete resection with negative microscopic margins), R1 (complete resection with positive microscopic margins) and R2 (incomplete resection). Under SAP, and with the patient under Endotracheal General Anaesthesia (ETGA) in the supine position, laparotomy was performed, where midline abdominal incision was made and abdomen was opened in layers. The mass was mobilised from the right colon, left colon (after opening the paracolic gutter), and the left ureter was identified and preserved. Adhesions from the duodenum and IVC were released. Wide excision of the retroperitoneal mass was done, following which complete hemostasis was obtained. Finally, the wound was closed in layers.

HPE of Excision Specimen

Gross Examination:
External surface of the soft tissue mass was multilobulated, with the capsule remaining intact. The cut surface revealed homogenous grey - white and yellowish glistening areas, predominantly soft to firm in consistency.

Microscopic Examination:
It revealed a neoplasm composed of lobules of mature adipocytes admixed with few lipoblasts (well - differentiated liposarcoma areas) with foci that consisted of sheets and clusters of round cells with vacuolated cytoplasm, vesicular nuclei and prominent nucleoli (round - cell liposarcoma areas). Also seen were areas showing sheets and clusters of spindle cells with bizarre nuclei and multinucleated tumour giant cells (undifferentiated pleomorphic sarcoma areas). Stroma showed areas of necrosis, hyalinization, calcification and congested blood vessels. Number of mitoses were 5 - 6/10 HPF, with presence of atypical mitoses. The tumour had a pseudo - capsule, which separates the tumor from adjacent soft tissue. Pseudo - capsule was not invaded by tumor. Circumferential margins were free from tumor infiltration. Lympho - vascular invasion was present. Regional lymph nodes were not found. Pathological stage: T4NxMx

Impression: De - Differentiated Liposarcoma (Grade 3)

4. Conclusion

Complete surgical resection is the most effective treatment for primary or recurrent retroperitoneal sarcoma. En bloc

resection often necessitates sacrificing contiguous structures such as the colon, kidney, spleen, pancreas, psoas muscle, small bowel, inferior vena cava, and aorta. A non - specialist may also perform lymph node dissection, which is completely unnecessary in RPS and can expose the patient to undue risk (e. g., chylous leak). Concomitant multiorgan resection occurs often in RPS surgery and the risk of complications can be additive with each component of the operation. For example, a patient undergoing concomitant distal pancreatectomy or splenectomy or left colectomy may develop a leak that could compromise an iliac artery reconstruction.

Pre - operatively, any pre - existing co - morbidities should undergo medical evaluation in anticipation of major surgery. Those with borderline or pre - existing renal insufficiency may need further investigation (e. g., split renal perfusion scan) to determine the tolerance of an ipsilateral nephrectomy, while those with significant cardiopulmonary disease should undergo updated evaluation (e. g., echocardiogram). Also, advanced discussion with the anaesthesiologist is often helpful to anticipate potential intraoperative needs (e. g., blood products) and the appropriate level of venous access and hemodynamic monitoring.

References

[1] Choi JH, Ro JY. Retroperitoneal Sarcomas: An Update on the Diagnostic Pathology Approach. Diagnostics (Basel).2020 Aug 27; 10 (9): 642. doi: 10.3390/diagnostics10090642. PMID: 32867125; PMCID: PMC7555595.
[2] Carbone F, Pizzolorusso A, Di Lorenzo G, Di Marzo M, Cannella L, Barretta ML, Delrio P, Tafuto S. Multidisciplinary Management of Retroperitoneal Sarcoma: Diagnosis, Prognostic Factors and Treatment. Cancers (Basel).2021 Aug 10; 13 (16): 4016. doi: 10.3390/cancers13164016. PMID: 34439171; PMCID: PMC8392612.
[3] Mota MMDS, Bezerra ROF, Garcia MRT. Practical approach to primary retroperitoneal masses in adults. Radiol Bras.2018 Nov - Dec; 51 (6): 391 - 400. doi: 10.1590/0100 - 3984.2017.0179. PMID: 30559557; PMCID: PMC6290739.
[4] Sassa N. Retroperitoneal tumors: Review of diagnosis and management. Int J Urol.2020 Dec; 27 (12): 1058 - 1070. doi: 10.1111/iju.14361. Epub 2020 Sep 10. PMID: 32914475.
[5] Dominguez DA, Sampath S, Agulnik M, Liang Y, Nguyen B, Trisal V, Melstrom LG, Lewis AG, Paz IB, Roberts RF, Tseng WW. Surgical Management of Retroperitoneal Sarcoma. Curr Oncol.2023 Apr 29; 30 (5): 4618 - 4631. doi: 10.3390/curroncol30050349. PMID: 37232807.