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Case Report

Retro Peritoneal Tumor - Neurofibroma

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ABSTRACT

Retroperitoneal Tumors (RTs) are important group of neoplasms but very rarely occur. RTs develop insidiously and are generally seen as large masses. Most of the times 50% of retroperitoneal masses are larger than 20 cm at the time of diagnosis. Cross-sectional imaging has shown revolutionary changes in diagnosing the patients with retroperitoneal neoplasms. Both computed tomography (CT) and magnetic resonance imaging (MRI) can contribute to the diagnosis of the tumor, though histological confirmation by FNAC or Truecut biopsy is often required because of the considerable overlap of imaging features. This article aims to share our experience in management of a 40 years old male with complaints of painless swelling in the abdomen since three years with rapid increase in size and shortness of breath for past three months. Clinically there was a mass measuring approximately 14x10 cms in left hypochondrium extending upto left flank. CECT suggested tumor mass entrapping the left kidney and spleen. Surgery was done and the mass along with spleen and left kidney were excised.

Key Words: Retro peritonium Neoplasm, Retroperitoneal masses, Truecut biopsy.

INTRODUCTION

The retroperitoneum is a complex potential space in the abdomen bounded anteriorly between the posterior parietal peritoneum and the transversal is fascia posteriorly. It extends from the diaphragm superiorly to continue into the extra peritoneal space in the pelvis inferiorly and is loosely divided into the anterior and posterior pararenal, perirenal and great vessel spaces. The retroperitoneum can hiber a wide varieties of pathologies, like benign tumors and malignant neoplasm's that can be either metastatic or primary lesions. Incidence of Benign lesions is four times less when compared to malignancies of retroperitoneum. RTs present several therapeutic challenges because of their relative late presentation and anatomical location.

Retroperitoneal tumors (RTs) usually present with abdominal distention and palpable mass. In many cases, they are accidental findings detected as a result of imaging techniques performed to investigate unrelated issues. It is not uncommon to find RTs in the gastrointestinal and urinary tracts but, patients rarely present with symptoms in these systems are involved. [1,2] Each year, approximately 250 to 300 new cases of retroperitoneal sarcoma are diagnosed in the United Kingdom. Approximately, one third of RT cases are sarcomas. The most common sarcomas are liposarcoma, malignant fibrous histiocytoma, and leiomyosarcoma, respectively. Other malignant RT types are lymphoma, epithelial tumors, malignant paraganglioma (which is considered to be benign when no metastasis occurs), and metastatic tumors. Fibromatosis, renal angiomyolipoma,

benign paraganglioma, neurofibroma, lipoma, angiofibroma, and schwannoma can be listed among the benign tumors. Surgery remains the most successful treatment method for RTs, significantly affecting post-operative survival. [3] RTs are frequently confused with lymphomas and develop without compressing the inner organs or causing significant lumen blockage. Liposarcomas are commoner in fifth to seventh decades of age, with a 5year survival rate between 40% and 50%. [4] The goal in surgical management of RTs is to achieve the optimal negative surgical border. In the presence of a positive surgical border, the 5-year survival rate decreases to 28%.High histological grade, inoperability due to invasion into vital organs, positive surgical border and Delayed diagnosis are considered as the most significant factors affecting survival. The average life expectancy for patients with high-grade RTs is 20 months, while for low-grade RTs, it is 80 months. Moreover, RTs larger than 10 cm generally have distant metastasis at the time of diagnosis. [5] Inspite of its rarity Primary retroperitoneal masses are important group of neoplasms. They account for only 0.1–0.2 % of all malignancies and arise outside the retroperitoneal organs. [6] Most primary retroperitoneal neoplasms arise from the mesodermal system with liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma together, accounting for greater than 80 % of primary retroperitoneal sarcomas. The remaining primary retroperitoneal masses arise predominantly from the nervous system. [7]

CASE REPORT

A 40 years old male who is moderately built and nourished presented to surgical OPD with complaints of painless swelling in the abdomen since three years (Fig-1) which showed rapid progression in size since three months and shortness of breath since two months. There is no history of decreased appetite and weight loss. No history of constipation, increased frequency of micturition. No evidence of pallor, lymphadenopathy, icterus with normal BP and PR. All routine hematological and biochemical tests were within normal limits. Normal chest x-ray. USG abdomen suggested a mass in the retroperitoneum. CECT abdomen showed a huge mass measuring approximately 14X10cms engulfing left kidney in the tumor capsule.(Fig-2)True cut biopsy was suggestive of ? neurofibroma. Patient was posted for surgery and a mass measuring 5.5kgs along with spleen and left kidney were excised.(Fig-3,4,5,6) Specimen was sent for Histological examination, which showed neuronal fibers wavy in nature with slender nuclei in lobular arrangement typically seen in Neurofibromas. (Fig-7)

Fig no-1

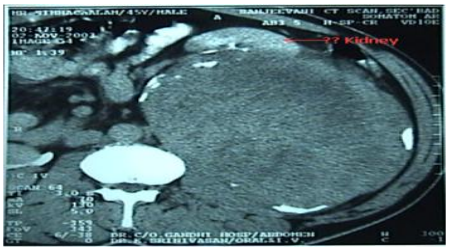

Fig no-2 CECT

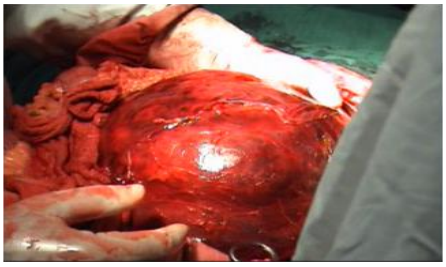

Fig no-3 Intra OP

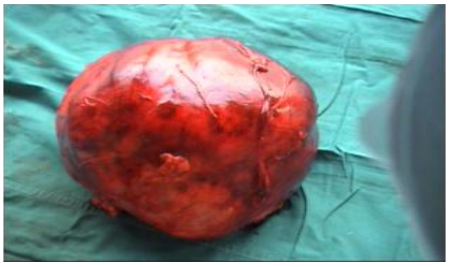

Fig no-4 5.5 Kgs mass

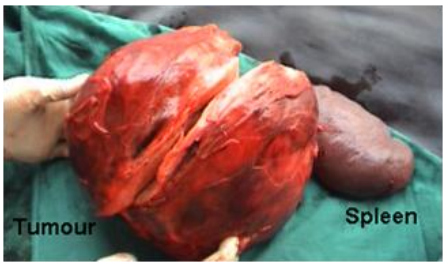

Fig no-5 mass with spleen

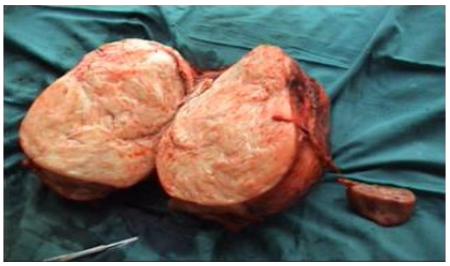

Fig no-6 mass with left kidney

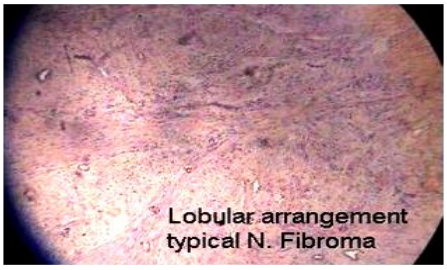

Fig no-7 histopathology

DISCUSSION

Patients with retroperitoneal tumors often present with huge tumors (larger than 10 cm in diameter) due to their insidious location and It is very challenging for surgeons to operate RTs due to their unpredictable clinical behavior, inaccessible location, and lack of successful treatments. A diagnosis can be reached through the evaluation of complaints, clinical findings, imaging methods, and trucut biopsy. The following are negative prognostic factors: poorly differentiated type, stage 2 to 3 tumor, tumor size larger than 20 cm, and a positive surgical border. [8] National Cancer Institute (NCI) and French Federation of Cancer Centers (FFCLCC) are currently used well known grading systems for sarcomas. [9] Studies have proved that the average life expectancy for patients who had complete resection was approximately 103 months, this decreased to 18 months for patients who had incomplete resection or whose tumors were inoperable. In 2000, Linehan et al. studied 159 patients diagnosed with RTs and reported that the increase in tumor size showed a significant role in the increase of local recurrence and in metastasis rates. [10] It is not uncommon for surgeons to be confused between Intra-abdominal sarcomas with retroperitoneal sarcomas, core needle biopsy should be done if imaging results cause practitioners to suspect lymphoma. Ultrasound role is very minimal except for USG guided biopsy. Before recognizing the symptoms the retroperitoneal tumors may reach sizable proportions. The results of any type of treatment for malignant retroperitoneal lesions are not particularly favorable, Since there is an extensive involvement of retroperitoneal tissues. Benign lesions in the retroperitoneal area usually can be surgically excised without undue difficulty. Pyelography and air contrast studies are other modalities in diagnosing the RTs

There are many primaries retroperitoneal masses and are classified as follows:

Liposarcoma: Most common sarcoma to occur in the retroperitoneum. Retroperitoneal liposarcomas account for 10–15 % of all liposarcomas and about 30 % of retroperitoneal sarcomas. [6]

Leiomyosarcoma: Second most common primary retroperitoneal tumour in adults. [6] Leiomyosarcomas can be divided into three main categories: extravascular (62 %), intravascular (5 %) and a combination of both (33 %). [11] They are believed to arise from blood vessels, spermatic cord or wolfian and mullerian duct remnants within the retroperitoneum.

Undifferentiated pleomorphic sarcoma (UPS): Previously called as malignant fibrous histiocytoma (MFH), is the third most common retroperitoneal sarcoma. [7] It is the most common soft tissue sarcoma in late adult life, occurring primarily in the 5th to 6th decades of life. Men are more commonly affected. [6]

Neurogenic tumors: Neurogenic tumours constitute up to 20 % of the primary retroperitoneal masses. [7] Majority of neurogenic tumours is benign and are seen in younger population. Neurogenic tumors

are classified according to their cells of origin. [12] Schwannomas and neurofibromas arise from the nerve sheath; ganglioneuromas, ganglioneuroblastomas and neuroblastomas from sympathetic nerves; and paragangliomas and pheochromocytomas from chromaffin cells.
Others: Solitary fibrous tumours (SFTs), Dermatofibrosarcoma protuberans (DFSP), Synovial sarcomas, Extra-adrenal myelolipoma, Sacrococcygeal teratoma, Aggressive angiomyxoma, Castleman disease (a benign giant lymph node hyperplasia, is an uncommon benign lymphoproliferative disorder), Lymphangioma and lymphangiomatosis, Paraganglioma.

Management of retroperitoneal tumors:

Complete surgical resection is the only potential curative treatment modality for RTs but local recurrence occurs in a large proportion of patients and is responsible for as many as of sarcoma related deaths. The likelihood of a complete margin negative surgical resection depends on tumor biology and invasion of adjacent visceral organs and vascular structures. [13,14] The most common organs requiring resection are the colon, kidney, pancreas and spleen. Local recurrence is common for RTs and remains the major cause of death. There is no strong evidence for the utility of radiotherapy or chemotherapy for treatment of RTs. [15] The dose and duration of radiotherapy are limited because of its toxic effect on the gastrointestinal tract. Radiotherapy is especially recommended for patients with high-grade tumors and those for whom complete resection is impossible. [16,17] In some tumors, such as the Ewing tumors, chemotherapy forms a significant part of the treatment, while for some particular histological types, special chemotherapeutic agents are sometimes used. These include agents such as doxorubicin and ifosfamide for the palliation of sarcomas, taxanes for angiosarcomas, gemcitabine and docetaxel for leiomyosarcomas, and trabectedin for mixoid-round cell liposarcomas and leiomyosarcomas. [18] Palliative surgery is recommended for patients diagnosed with low and intermediate grade RTs in the case of local recurrence to manage the symptoms and to increase the patients' quality of life. [19]

In this particular patient, an additional organ resection was needed due to tumor extension to spleen and engulfment of left kidney so that macroscopic clearance is achieved. Hence splenectomy and left nephrectomy was done

CONCLUSION

Primary retroperitoneal neoplasm's are rare. Usually present late and cause symptoms or become palpable once they have reached a significant size. Cross-sectional imaging is key to the evaluation of retroperitoneal masses and in the pre-operative staging and surgical planning of these lesions. Wide surgical resection for treatment of RTs remains the gold standard procedure. While the efficiency of pre-operative and post-operative radiotherapy and chemotherapy is still a controversial issue. Complete resection of retroperitoneal tumors significantly influences patient survival and should be the goal of surgical intervention.

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