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CASE REPORT

A Huge Retroperitoneal Liposarcoma with Renal Involvement

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ABSTRACT

Retroperitoneal Soft Tissue Sarcomas (RPS) are rare tumors, which account for approximately 12 per cent to 15 per cent of all soft tissue sarcomas. Early diagnosis is not easy as there is an absence of specific clinical presentations. Symptoms would only be detected if the liposarcoma presses on the surrounding organs. Surgery remains the mainstay of treatment for retroperitoneal sarcomas. We report a case of a giant retroperitoneal liposarcoma of 5.3 kg of weight in a 65 years old woman, with abdominal distension, precocious satiety, and weight loss after a cystocele surgery. The TC showed a huge abdominal mass (14 cm x 23 cm), encapsulated and heterogenic; with calcifications, which causes a large displacement of the organs. It was successfully treated with surgical excision.

Abbreviations: RPS: Retroperitoneal Soft Tissue Sarcomas; CT: Contrast Enhanced Computed Tomography; MRI: Magnetic Resonance Imaging; PET/CT: Positron Emission Tomography/Computed Tomography using 18F-fluorodeoxyglucose; IHQ: Immunohistochemistry; EUS-FNB: Endoscopic Ultrasound – Fine Needle Biopsy; GIST: Gastrointestinal Stromal Tumor; WDL: Well Differentiated Liposarcoma; DDL: Dedifferentiated Liposarcoma; LMS: Leiomyosarcoma.

Introduction

Retroperitoneal soft tissue sarcomas are rare tumors which account for approximately 12 per cent to 15 per cent of all soft tissue sarcomas with a mean incidence of 2.7 per million [1]. The most frequent sarcoma subtypes in the retroperitoneum in adults over 55 years old are WDL and DDL (40 per cent) and LMS (27 per cent). In younger age groups, leiomyosarcoma becomes more common than liposarcoma [2]. Early diagnosis is difficult as there is an absence of specific clinical presentations. Symptoms would only occur if the liposarcoma presses on the surrounding organs. The management is surgical intervention. Even with complete removal of the liposarcoma, prognosis remains poor [3].

Case Presentation

A 65 years old female, without a medical history, come to the hospital six months after a cystocele surgery, with abdominal distension, precocious satiety, weight loss, and a estrange sensation of movement of something inside

of the abdominal cavity during left lateral decubitus. The physical examination indicated a 15 cm x 20 cm flexible and mobile mass located in the upper abdomen. The laboratory examination, including routine blood reveled anemia with hemoglobin 10,5 g/dl (11,5-16 g/dl), hematocrit 32 per cent (36-51 per cent), thrombocytosis 422.000 /µL (130.000-450.000/µL), elevated C-reactive protein 212 mg/dl (<5) and erythrocyte sedimentation rate of 105 mm. The CT showed a huge abdominal mass (14 cm x 23 cm), encapsulated and heterogenic; with calcifications, that causes a large displacement of organs. The mass was in contact with the tail of the pancreas, left kidney spleen an, in appearance, it was neo proliferative. Isolated adenopathy in the hepato-pancreatic region (10-15 mm), Small amount of free liquid in pelvis minor. The EUS (5-7 MHz) showed a great solid mass, retro gastric, heterogenic, with vascularization, lobulated. Which was visualized from the upper part of the body to the gastric antrum. Due to the large size of the lesion, it was not possible its measurement and its dependence of the gastric wall in the zones visualized. Trans gastric aspiration puncture (2 passes) was performed with a 22G biopsy needle (Shark-core) guided by EUS of the lesion, obtaining a large amount of material for the histological study. The EUS suggested a possible mesenchymal tumor type GIST. The cytology showed connective tissue cells and absence of epithelial cells. The biopsy showed fibroblasts and abundant lymphoplasmacytic cells and occasionally histiocytic cells. It did not contain spindle cells, or vesicular cells, and it was not possible to determine the IHQ, which was, at the same time, suggestive of a tumor of the gastrointestinal stromal. During laparotomy in "J" of the left side, the patient received complete resection of the tumor that seems to be glued to the kidney, left nephrectomy, splenectomy and left ovarian artery ligation due to its attachment to the diaphragm, with left diaphragm reconstruction and pleural drainage placement. During the surgery, as the patient had anemia, it was required a blood transfusion (2 CCHH intraoperative), without any other complication. The total mass was 30 x 23 x 16 cm with a weight of 5330 g. Hematoxylin and eosin staining of the tissue showed a mesenchymal neoplasia with fusiform pattern, with cells with marked pleomorphism. The immune histochemically staining was positive for vimentin and S-100 and negative for desmin, actin, CD34, CKAE1AE3; highly suggestive pleomorphic liposarcoma. The kidney was partially infiltrated by neoplasiaand the ureter showed absence of histological signs of malignity.

The patient recovered well and, after surgery, she was transferred to the Oncology Department, no tumor recurrence was observed at the CT after the surgery.

Discussion

Retroperitoneal soft-tissue sarcoma is a rare disease that accounts for 10 per cent to 15 per cent of all soft-tissue sarcomas and 33 per cent of all malignant retroperitoneal tumors. Among these, Liposarcoma (LPS) is the most common histologic type [4,5] and it is classified into four subtypes: (1) undifferentiated; (2) pleomorphic; (3) well differentiated and (4) myxoid /round cell [6]. The undifferentiated and pleomorphic type are neoplasm with high grade of malignancy accompanied by remarkable biological aggressiveness and with metastatic potential while well-differentiated and myxoid/round cell forms are tumors with low grade of malignancy, associated with a more favorable prognosis [7]. It commonly occurs in patients aged between 40-60 years old with a 1:1 ratio between male and female [8].

CT is the most useful and widely available primary imaging investigation. Because soft tissue sarcoma accounts for only a third of retroperitoneal tumors arising extrinsic to the solid abdominal viscera, other diagnoses must be considered, such as: lymphoma, angiomyolipoma, neurofibroma [9]. Percutaneous core needle biopsy usually confirms the diagnosis and it is the gold standard for diagnosis, but rarely lesions are not amenable or high risk for biopsy and the differential diagnosis based on imaging becomes crucial. MRI is reserved for patients with allergy to iodinated contrast agents or problem solving where for example muscle, bone or foraminal involvement is equivocal on CT. MRI

may also be useful for delineating disease in the pelvis [10]. Due to the variability of tumor grade, FDG PET/CT has no routine role but again can be used for problem solving. It is utilized when pulmonary abnormalities are detected on CT. Following resection, surveillance with CT is useful for detection of recurrence [11].

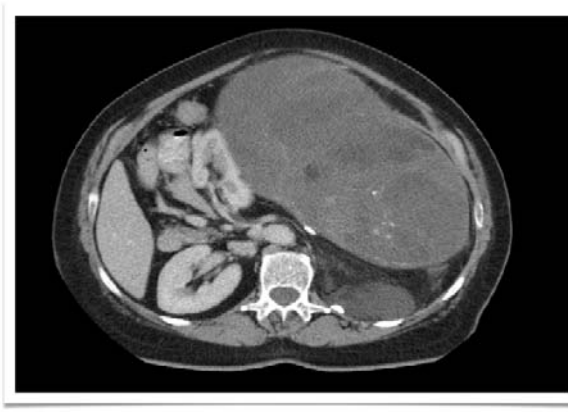

Figure 1: CT of the abdomen. Green arrow pointing out the liposarcoma; red arrows pointing out the calcifications and necrosis.

Figure 2: Intra-operative findings. The retroperitoneal tumor weighing 5.3 kg.

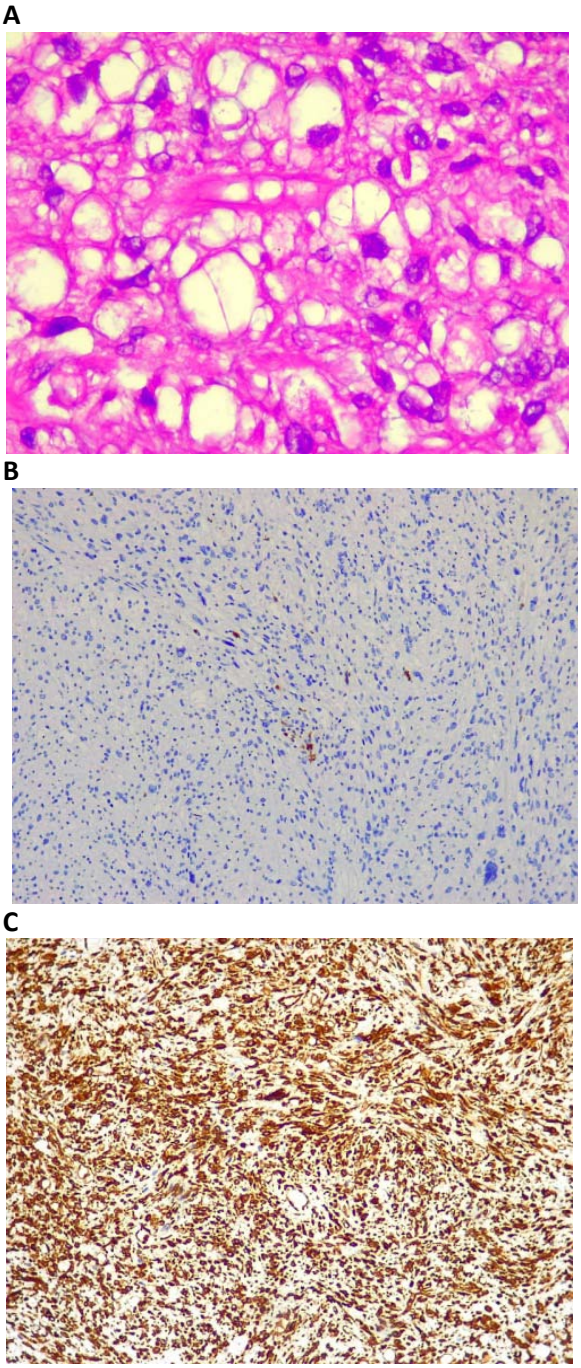

Figure 3: Histopathological findings of the tumor diagnosed pleomorphic liposarcoma (HE stain, original magnification x60). (a) Most of the tumor cells consisted of Pleomorphic lipoblasts. (b) S100+ (c) vimentin+.

Surgery remains the mainstay of treatment for retroperitoneal sarcomas. In the majority of cases, one or more organs need to be resected together with the tumor in order to achieve complete resection. Very often, the ipsilateral hemicolon and kidney are resected 'in block' with the tumor. Depending on the histological diagnosis a clear multidisciplinary discussion has to be warranted to determine if radical surgical strategy is possible and if so, whether a neoadjuvant approach is also recommended [12]. The tumor size, location and relationship to neighboring viscera and vascular structures must be defined to plan a possible adjacent visceral resection. Common causes for non resectability are involvement of the celiac axis and superior mesenteric vessels or involvement of bone (i.e., spine)

[13]. If surgery may technically be a challenge in attaining clear resection margins, radiotherapy and/or systemic therapy may be considered to improve local control [12]. Currently, there is a randomized phase III trial STRASS trial (EORTC 62092) through the recruitment of patients from both Europe and North America in order to compare radiation therapy followed by surgery with surgery alone in treating patients with previously untreated non metastatic retroperitoneal soft tissue sarcoma. It is expected that the STRASS study will provide clarification on the role of RT in the primary setting [14].

Conclusion: Retroperitoneal soft-tissue sarcoma is a rare disease, among these LPS is the most common histologic type, early diagnosis is difficult as there is an absence of specific clinical presentations. Symptoms would only occur if the liposarcoma presses on the surrounding organs. The management is surgical intervention.

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