



Primary retroperitoneal liposarcoma: a case report and literature review

Vytautas Eimontas¹, Kristina Zviniene¹, Diana Petrova¹, Darius Pranys², Daimantas Milonas³, Mindaugas Kiudelis⁴

¹*Clinic of Radiology, Lithuanian University of Health Sciences, Medicine Academy, Kaunas, Lithuania,*

²*Department of Pathology, Lithuanian University of Health Sciences, Medicine Academy, Kaunas, Lithuania,*

³*Clinic of Urology, Lithuanian University of Health Sciences, Medicine Academy, Kaunas, Lithuania*

⁴*Clinic of Surgery, Lithuanian University of Health Sciences, Medicine Academy, Kaunas, Lithuania*

Abstract

Introduction: Liposarcomas are mesenchymal neoplasms with atypical adipocytes and lipoblasts in the background of mature adipose tissue. Retroperitoneal space offers an environment in which sarcomas can reach impressive sizes before they become symptomatic.

Case Report: We are reporting a case of 45 years old previously healthy female with left flank and back slight pain and mild swelling on the left abdominal side. Woman visited her family physician, because of left flank pain. Abdominal ultrasound detected a large tumorous mass on the left retroperitoneal space. Enhanced contrast CT was performed after this and findings indicated tumor containing retroperitoneal fat also the collecting renal system was slightly expanded because of the tumor closely attached to the middle ureter. Midline laparotomy was performed and tumor en bloc with left kidney, left adrenal glands and surrounding adipose tissue was radically removed. Histological analysis revealed atypical – lipomatous tumor, G1 (well – differentiated).

Conclusions: Liposarcoma is a malignant mesenchymal neoplasm composed of lipogenic tissue with varying degrees of atypia, which grow slowly and silently and may reach the enormous size. The main treatment of retroperitoneal sarcomas is radical surgery.

Keywords: retroperitoneal space, well – differentiated liposarcoma, tumor removal.

Introduction

Liposarcomas are mesenchymal neoplasms with atypical adipocytes and lipoblasts in the background of mature adipose tissue. They can appear anywhere in the body. Retroperitoneal space offers an environment in which sarcomas can reach impressive sizes before they become symptomatic. Frequently, retroperitoneal liposarcomas are incidentally diagnosed or identified with cross-sectional imaging as the part of a workup for other problems. When patients have symptoms, however, the most common complaint is abdominal or back pain [1].

Case report

We are reporting a case of 45 years old previously healthy female with left flank and back slight pain and mild swelling on the left abdominal side. During abdominal ultrasound examination a large tumorous mass of unknown origin in left retroperitoneal space was detected. The patient was sent to surgeons' consultation for the treatment tactics decision.

Uterus myoma was removed before two

years. During physical examination, patient's arterial blood pressure (ABP) was 130/80 mm/Hg, pulse rate – 72 beats/min., pulmonary auscultation and blood test without any abnormality. Abdominal palpation was sensitive in upper part with smooth mass palpable on left side of abdomen, without signs of muscles tension or peritoneal irritation.

Abdominal computed tomography (CT) demonstrated a massive ~13.0x9.5x10.0 cm in size mixed structure tumor on the left retroperitoneal space which expanded to the left perirenal space, fat component surrounded dorsal-external renal border without direct invasion, renal displacement superiorly by dense component and some rotation was noticed, also the collecting renal system was slightly expanded because the tumor closely attached the middle ureter part.

The left kidney structurally appeared normal and any enlarged pathological lymphnodes or destruction lesions in bones were observed. Radiologist who was reviewing abdominal CT images suggested that tumor is likely to be the retroperitoneal liposarcoma. (Fig.1,2).



Figure 1 and Figure 2 Abdominal sagittal and axial postcontrast preoperative CT images showing: large tumor of mixed dense (arrow)–fat (arrowheads) structure occupying left retroperitoneal space, structurally normal kidney is displaced superiorly and rotated.

Additionally woman underwent pulmonary CT scan for possible tumor spread, but there were no metastases in lungs parenchyma, nor mediastinal lymphadenopathy revealed. Multidisciplinary team meeting recommended radical surgery - tumor removal. Midline laparotomy was performed and tumor en bloc with left kidney, left

adrenal glands and surrounding adipose tissue was radically removed and sent to histological examination. Operation was performed 12 days after abdominal CT. There were no intraoperative and postoperative complications and the postoperative period was uneventful. Removed specimen is shown in Fig.3 and Fig.4.

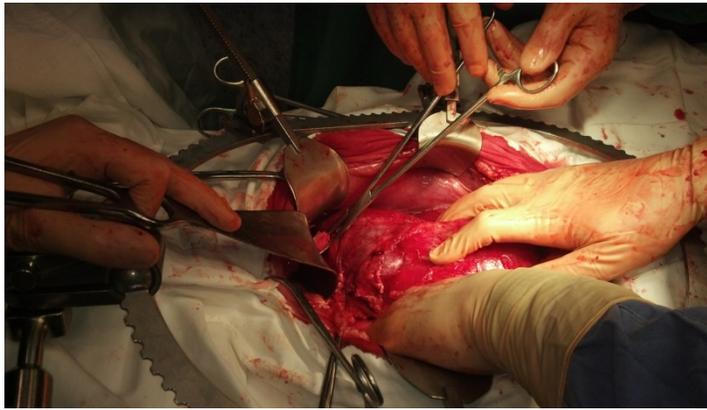


Figure 3: Tumor (liposarcoma) is in the left retroperitoneal space.

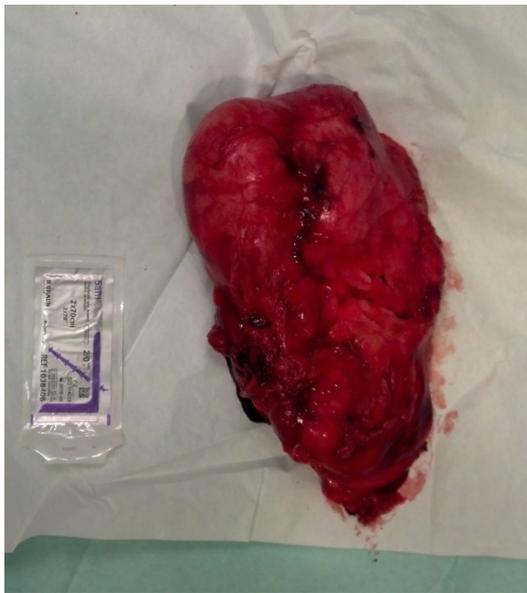


Figure 4: Macroscopic removed specimen (left kidney with proximal part of ureter, left adrenal gland and surrounding adipose tissue).

Histological analysis revealed atypical-lipomatous tumor, G1 (well-differentiated). Left kidney and adrenal gland histologically were normal. Removed kidney had normal histological structure, adjacent to kidney tumor infiltration is seen. Tumor infiltration consist of broad bands of fusiform, medium-sized chromaffin cells, between cells some lymphocytic infiltration is observed. Fusiform cells moderately has positive staining with CD34 immune marker. Lipoblasts has positive staining with S100P immune marker, 10% of tumor cell nuclei showed positive reaction with Ki67. (Fig.5)

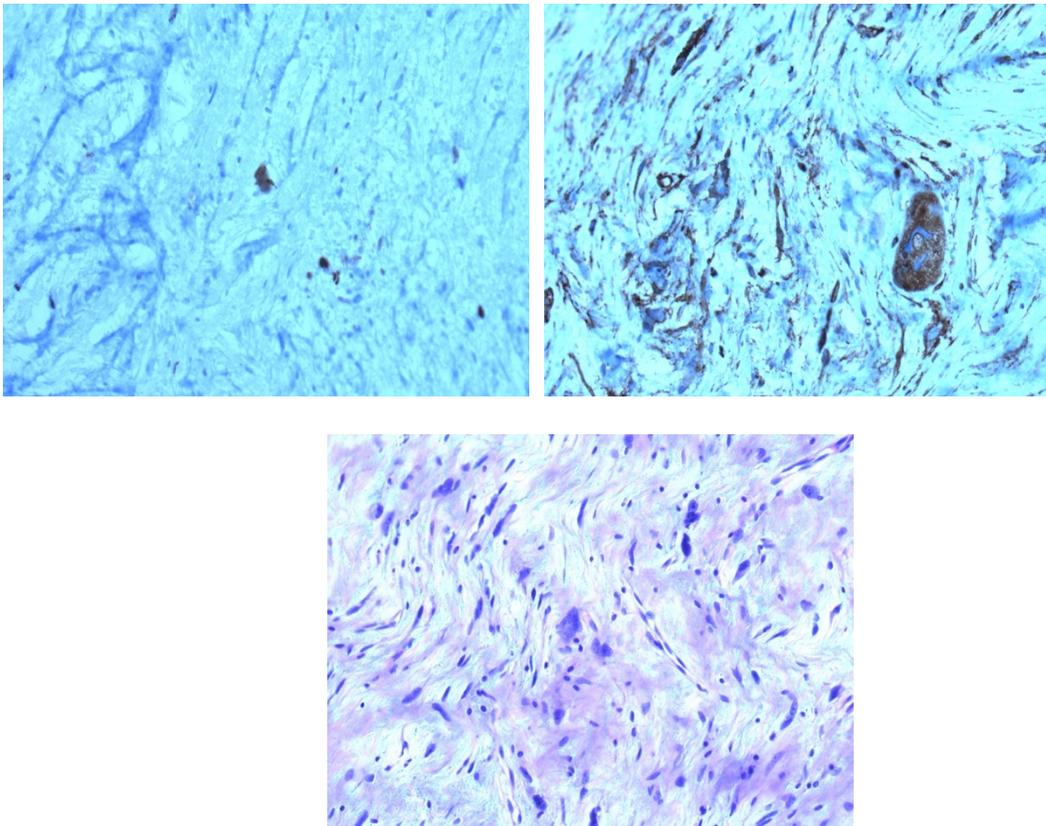


Figure 5: Histological pattern discription.

Literature review

Liposarcomas are malignant neoplasms with adipocyte differentiation. They occur most commonly in 5th – 6th decades of life. Mostly arises in deep soft tissue or in the retroperitoneum [2]. Approximately 90% of cases occurs de novo. Retroperitoneal space is the second most common site of occurrence, with up to 36% of liposarcomas occurring at this site. The tumor is often deep-seated and large at the time of the diagnosis, as it was in our case, because the retroperitoneal space provides a large potential volume allowing sizeable growth prior clinical signs and symptoms develops [3,4].

Liposarcomas arise from perinephric fat in the retroperitoneal space and they may encase kidneys or cause pyelo – ureteric obstruction. These signs were obvious in our case.

During patient's physical examination, a painless, palpable abdominal mass, which may compress and/or dislocate adjacent organs, is found in 70–80% of all cases [5]. In our case, a palpable mass was on the left abdominal side, left kidney was dislocated, proximal ureter part was compressed.

It is well known that clinical behaviour of liposarcomas is closely related to histological characteristics. Liposarcomas has three principal forms: 1) atypical – lipomatous tumor (ALT) – well-differentiated/de-differentiated 2) myxoid/round cell 3) pleomorphic [6].

Well-differentiated lesions grows slowly with more favourable outlook

than aggressive myxoid/round cell. Pleomorphic variants tend to recur after excision and metastasize to lungs [2]. Pleomorphic and myxoid types are most common intramuscular [7]. Well-differentiated variant accounts for about 40% to 45% of all liposarcomas and therefore represents the majority of adipocytes' malignancies, they show predominant presence of mature fat cells, and the amount of widely diffused lipoblasts is relatively low [8]. CT scan or magnetic resonance imaging (MRI) is performed in order to evaluate tumor

extension and relation to adjacent organs, because the determination of tumor resectability is fundamental for surgery planning.

The well-differentiated components are indistinguishable from normal fat by imaging, and thus follow fat characteristics on both CT and MRI. The fibrous septa of fatty component may be thick, irregular or nodular and can be homogeneously enhanced on contrast-enhanced CT [9,10].

Classically, de-differentiated liposarcoma is characterized by the presence of a well-differentiated lipomatous lesion juxtaposed with an area of high-grade de-differentiation [11]. In these forms imaging shows both fatty and no fatty solid components, which may be discontinuous in the retroperitoneal space. De-differentiated variant exhibits less aggressive clinical behaviour than high grade pleomorphic sarcomas [6].

A recent study by Lahat et al. examined the utility of preoperative CT imaging in determination of liposarcomas histologic classification as either well – differentiated and de – differentiated. Last lesions were more likely to be infiltrative, hypervascular with areas of necrosis and focal/nodular water density [12].

It has been recently reported that well-differentiated and de-differentiated liposarcomas have different biological behaviours, in de-differentiated tumors they tend to present as a recurrence more often, require multi-organ resection more frequently and has a shorter disease free interval when compared to well-differentiated subtypes [13]. The mainstay of treatment is complete surgical removal, which can be challenging as the tumor may be difficult to distinguish from normal retroperitoneal fat. In all cases, the surgical goal should be complete tumor removal with microscopically negative margin (R0). The extent of disease in patients with retroperitoneal sarcomas can sometimes require nephrectomy at the time of primary resection. Indications for nephrectomy included gross tumor invasion or total encasement [1]. In our case, tumor widely encased kidney and was closely attached to the

collecting system.

Prognostic factors for survival of liposarcoma are the histologic subtype and the resection margin [3]. Sepideh Gholami et al. in their clinical study investigated 41 patients with retroperitoneal sarcoma and found that high grade tumors were associated with shorter intermediate 5-years survival rates of 75% and 65-90%, respectively [14]. Close follow-up after surgery is mandatory due to high rates of recurrence.

The roles of adjuvant chemotherapy and radiotherapy are controversial. Some studies

survival and duration of symptoms was inversely proportional to prognosis [15].

From the literature review, the overall 5-years survival for well-differentiated subtypes is 90%, while 5-years survival for pleomorphic subtypes is only 30-50%. De-differentiated and myxoid/round cell subtypes have showed that local control of retroperitoneal sarcomas was enhanced with radiotherapy but none of them supported an increase in survival [14].

Conclusions

Liposarcoma is a malignant mesenchymal neoplasm composed of lipogenic tissue with varying degrees of atypia, which grow slowly and silently and may reach the enormous size. The treatment of retroperitoneal sarcomas is based on radical surgery. All diagnostic algorithms and other treatment modalities should be managed by a multidisciplinary team. Careful identification of tumor extent is also very important because incomplete removal can have an adverse effect upon the prognosis

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