

## GIANT RETROPERITONEAL LIPOSARCOMA WITH ENTERAL AND BLADDER COMPRESSION—CASE PRESENTATION AND LITERATURE REVIEW

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### Abstract

*Liposarcomas are the most common retroperitoneal sarcomas, occurring more frequently in the age range of 40-60 years. The purpose of this article is to highlight the diagnostic and therapeutic modalities of retroperitoneal giant liposarcomas exemplifying by presenting a 41 year old patient with a large retroperitoneal tumor (29/24/12 cm) excised with difficulty to healthy tissue and following the histopathological result, it was shown to be a primitive liposarcoma with variable profile. The patient had a good post-operative evolution and refused any adjuvant treatment (radiotherapy). He developed a large local relapse associated with vertebral metastases, with reintervention having a very difficult tumor reduction but with unfavorable postoperative evolution, with multiple organic dysfunction syndrome leading to the patient's death. Analyzing the data from the literature, we concluded that complete surgical excision with the affected organs and structures is the gold standard of treatment, but tumor recurrences are frequent (especially for dedifferentiated subtypes) and lead to increased mortality rates; adjuvant or neoadjuvant radiotherapy can help to prevent such relapses (there are still many ongoing clinical studies).*

**Keywords:** tumor, retroperitoneal, giant, liposarcoma, excision

### Introduction

Several pathological changes may occur in the retroperitoneal space, including retroperitoneal tumors that may be benign or malignant, the latter being divided into primitive or metastatic tumors [1].

The first description of a retroperitoneal tumor appears to belong to Giovanni Battista who presented in 1761 data about a retroperitoneal lipomatous tumor discovered at the time of a necropsy [2].

Primary retroperitoneal tumors are developed from retroperitoneal tissue, with the exception of retroperitoneal organs (kidneys, pancreas, ureters, ascending and descending colon, etc.) or embryonic remains [3]. Retroperitoneal sarcomas account for approximately 50% of all retroperitoneal tumors [4]. Liposarcomas are the most common retroperitoneal sarcomas (20-45%) with a peak incidence between 40-60 years of age, with no significant gender differences [5]. In extremely rare cases, these tumors can reach impressive

dimensions, producing compressive type symptoms (in the retroperitoneal or intraperitoneal organs).

### Case presentation

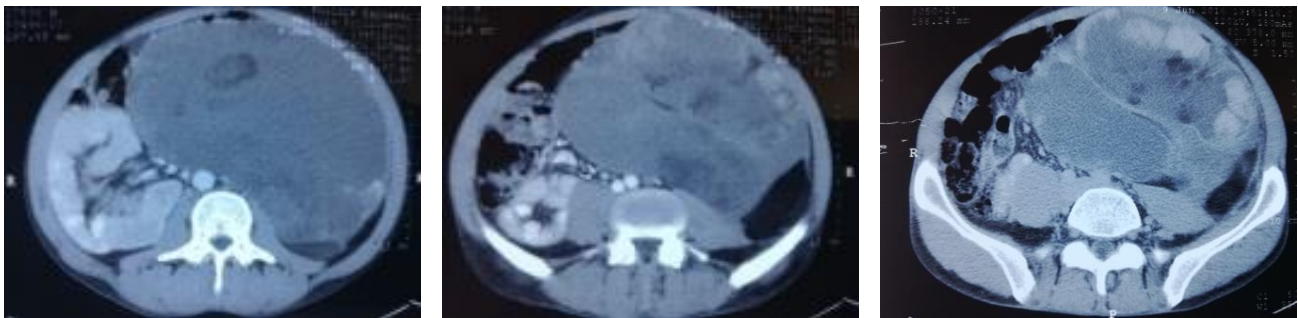
We present the case of a 41-year-old man admitted to the surgery clinic for diffuse abdominal pains that occurred a few days ago, bloating and abdominal meteorism, nausea and polakiuria. From the medical history, we do not retain significant data about previous personal or heredocolateral pathologies.

The local clinical examination showed a meteorised abdomen, mobile with respiratory movements, painful on palpation in the left hypochondrium and the left flank, without signs of peritoneal irritation and a bulky tumor (diameters about 30/25 cm), which occupied almost the entire peritoneal cavity, with no mobility.

Laboratory examinations are within

normal limits, except for fibrinogen that has a value of 974 mg /dl and a VHS of 87mm / 1h.

Thoracic, abdominal and pelvic CT performed with i.v. contrast reveals a giant (29/24/12 cm) tumor that cranially goes up to the gastric horizontal part, caudal to pelvis (in contact with bladder), laterally reaching the abdominal wall, and medially exceeds the median line by moving jejunono-ileal bowels to the sub-hepatic space (Figure 1). The tumor had fluid, parafluid and solid areas and a central area that present necrosis. In the peripheral zone there is an intense contrast enhancement, but it seems to keep a thin wall; no parenchymal origin could be determined. The bladder is moved caudally by the tumor but has the limit of demarcation to it. Intimate report with left kidney is described without ureterohydronephrosis. No other intraperitoneal pathological changes or secondary visceral or lymph node determinations were reported. Pulmonary parenchyma is homogeneous, with no focused processes.



**Figure 1 – Contrast CT of retroperitoneal giant tumor**

Simple abdominal radiography did not revealed any images suggestive for hydroaeric levels (requiring an abdominal-pelvic tumor).

There is no conclusive data on the benefits of any preoperative (neoadjuvant) oncologic treatment, therefore we didn't consider it appropriate to perform a preoperative tumor biopsy.

After a proper preparation, we have performed an open surgical intervention. We have found a 30/24 cm tumor, which was developed retroperitoneal and pushed the entire small intestine to the right and the cranial, also compressing the bladder. Left retroperitoneal access was obtained after medial mobilization of the descendent colon and the tumor (which was predominantly left-side paravertebral) was

excised with difficulty together with the left prerenal fascia which was invaded by the tumor.

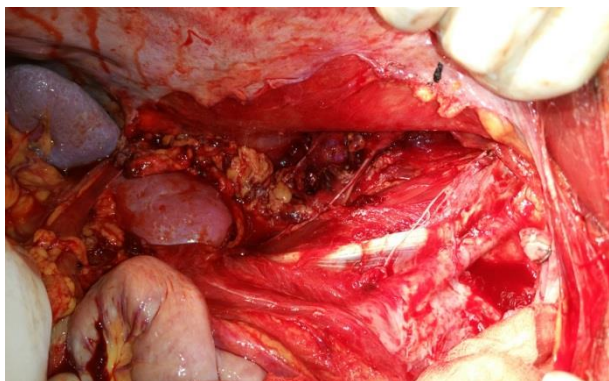
Urinary bladder and left ureter are not invaded tumorally (there is only compression of proximity to the bladder); they remained integral at the end of the surgical intervention (Figure 2).

The excised tumor weighs about 10 Kg, is polilobate and encapsulated (Figure 3).

The postoperative course was favorable, with rapid recovery of bowel function, micturitions was normal, without hematuria.

The anatomopathological result revealed that it was a primitive malignant retroperitoneal tumor, namely a giant liposarcoma with variable profile (mixoid, round cells type and dedifferentiated), large areas of necrosis (over 50% of the tumor surface) and mitosis (Figures

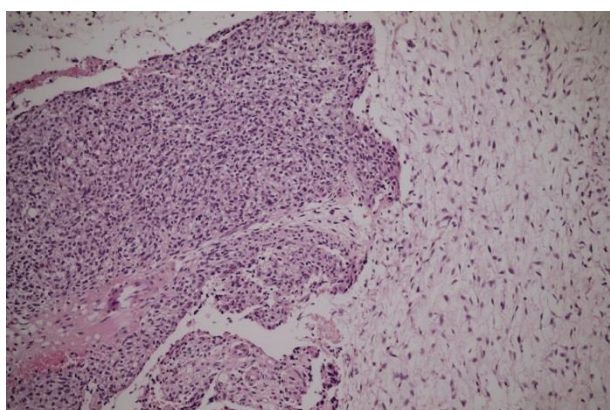
4 and 5). The dedifferentiated character of the tumor is supported by the presence of tumor areas suggesting another histological form of sarcoma (in our case-angiosarcoma), but with liposarcomatous phenotype at immunohistochemical tests (Figure 6).



**Figure 2 - appearance of left kidney and other retroperitoneal and intraperitoneal viscera after tumor removal**



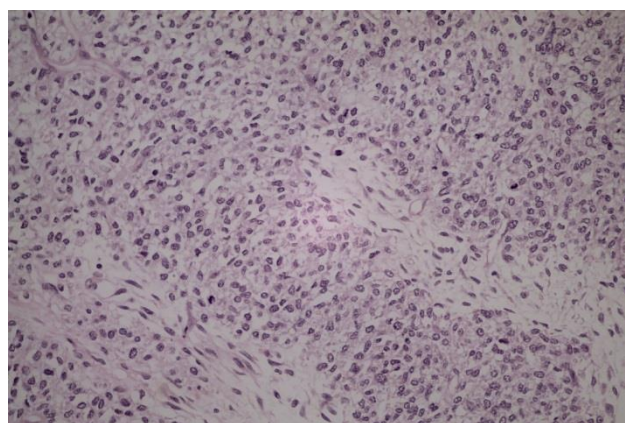
**Figure 3 - Appearance of the excised giant retroperitoneal tumor**



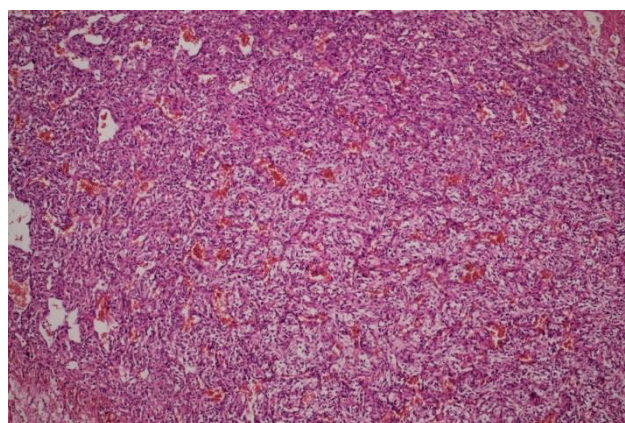
**Figure 4 - Interface between the mixoid and round cell histology. Color Hematoxylin - Eosin, X20**

At discharge, the patient was advised to go to the territorial oncology service in order to establish the postoperative oncological treatment, explaining him the importance of oncological therapy, but the patient refused this,

not even presenting to the recommended postoperative medical checkups.



**Figure 5 - Detail showing the numerous mitoses predominantly distributed in the round cell part of the tumor. Hematoxylin - eosin, X40:**



**Figure 6 - Angiosarcoma area, Coloration Hematoxylin-eosin, lens X10:**

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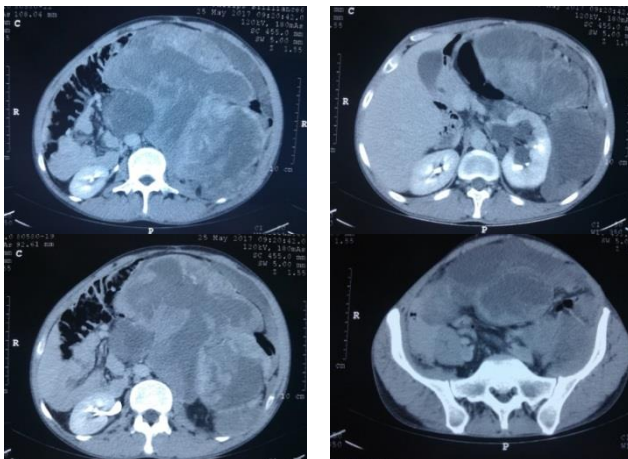
After 11 months from discharge, the patient returns to our clinic for altered general condition, diffuse abdominal pain, marked weight loss (approximately 7 kg), paresthesia and left thigh pain.

The local exam shows a painfully abdomen on palpation and a giant tumor that occupies the entire peritoneal cavity.

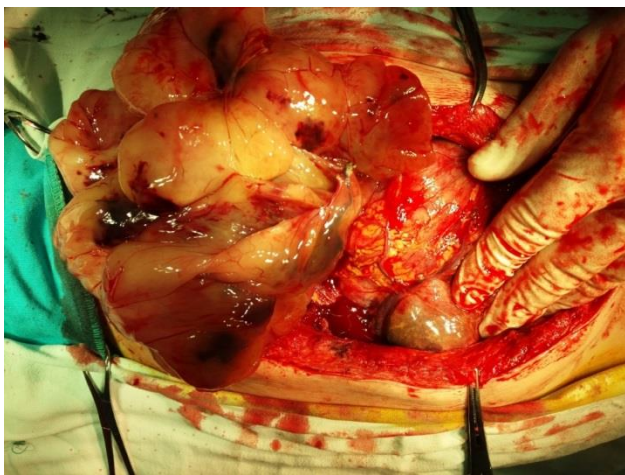
Laboratory exams show a slight leukocytosis (12,000 elements / mm<sup>3</sup>), the rest being within normal limits.

Abdominal-pelvic contrast CT shows a large expansive abdominal process that occupies

quasi-complete the abdominal cavity (Figure 7). The lesion is irregularly contoured, multilocular, and exerts a compressive effect on the stomach, transverse and descendant colon and sigmoid; small bowel - pushed to the right flank, spleen-anterior and cranial and left kidney - medial with left ureterohydronephrosis grade II and delayed excretion on the left. Urinary bladder is imprinted by the tumor and in the T12 vertebral body, posterior and on the left part there is a focal lesion with osteocondensing appearance (suggestive for secondary determination).



**Figure 7 - CT aspects with recurrent liposarcoma**



**Figure 8 - Intraoperative aspect of the recurrent tumor**

At the second open surgery we have found a giant tumor, similar to the initially excised (but local conditions were more unfavorable with major postoperative adhesions) that almost completely occupied the abdomen (Figure 8). Acytoreductional surgery was performed by excision of about 80% of the tumor tissue (about 6 kilograms in weight), a complete removal being totally impossible due to persistent intraperitoneal diffuse bleeding and

requiring hemostasis with gauze fields left in the peritoneal cavity for 48 hours.

The histopathological examination confirmed that it was a giant liposarcoma (28/22 / 9,5) cm, with the same histological characters as in the first surgical procedure.

However, the postoperative course was unfavorable this time, due to the installation of multiple organ dysfunction syndrome (with pulmonary, renal and hematological impairment); the general condition progressively deteriorated, and the exitus occurred few days after the surgery.

## Discussions

Retroperitoneal liposarcoma may develop asymptomatic for long periods of time [6]. Symptoms often occur when the tumor grows to impressive dimensions, making it difficult for a surgical approach and decreasing the life expectancy.

The clinical manifestations of giant retroperitoneal tumors are generally compressive, the most common symptom being abdominal or dorsal pain [7]. Compression over gastro-intestinal tract alters the transit even reaching intestinal occlusion, while bladder and ureteral compression can lead to urinary disorders such as polakiuria, dysuria, and various degrees of ureterohydronephrosis that predispose to recurrent urinary infections. Respiratory dysfunction with dyspnea can occur due to diaphragm ascension by bulky tumors. Medial compressions with appearance of specific neurological signs are also possible [8]. It may also appear lower limb edema of different degrees (due to retroperitoneal venous compression) or various hernias (inguinal, femoral and umbilical) due to increased intraperitoneal pressure.

Contrast enhanced CT and MRI are very useful for imaging diagnostic but the certain diagnostic is established based on the pathology examination [9].

Histologically liposarcomas are classified in 5 subtypes: well differentiated, mixed, round, pleomorphic and undifferentiated cells. Liposarcomas are spectacular due to its impressive dimensions and variable histological

aspects, especially for liposarcomas with retroperitoneal localization [10].

Differential diagnosis includes a wide range of diseases including: primary neoplasm arising from a retroperitoneal organ (duodenum, kidney, pancreas, adrenal glands) other types of retroperitoneal sarcoma (leiomyosarcoma, fibrosarcoma, angiosarcoma), secondary retroperitoneal tumors (due to testicular cancer with retroperitoneal metastases) or malignant lymphoma [7].

According to the existing data up to now, in case of the retroperitoneal giant liposarcoma, the only accepted attitude according to medical literature that has beneficial effects on quality of life and life expectancy is surgical excision through healthy tissue [11-15]. Many studies show that complete tumor excision led to the highest survival rate at 5 years, the longest disease-free interval and the lowest number of reinterventions for local or metastatic recurrence [14].

Due to tumor adherence or invasion in the retroperitoneal organs, the surgical approach is often difficult and burdened by numerous risks [5]. If tumor invades adjacent organs and structures, complete tumor excision with or without partial resection of organ (eg partial nephrectomy, caudal pancreatectomy, etc.) is indicated if this is possible without vital risk (uncontrollable haemorrhage, non-functional contralateral kidney etc.). The invasion of large retroperitoneal vessels is a negative prognostic factor due to the fact that complete removal of the tumor to healthy tissue is difficult or often impossible in such cases [16].

Regarding the laparoscopic approach of retroperitoneal liposarcomas, several authors pointed out that this technique can be feasible and safe with a rapid recovery of the patient, despite technical difficulties, being preferred for well differentiated subtypes without adrenal intraperitoneal processes. Thus preoperative MRI is very important in selecting cases where the laparoscopic approach could be feasible [17, 18]. However, there are reported few cases of laparoscopically approached retroperitoneal liposarcomas, due to the obvious difficulties being particularly evident in this case [19].

In patients where full resection of retroperitoneal liposarcoma is impossible or with unacceptable risks, most authors

recommend partial debulking of the tumor followed by radiotherapy with satisfactory results on quality of life and survival. This therapeutic attitude is more effective than radiotherapy applied on tumors subjected only to biopsy [13,14]. However there are studies that do not confirm the increase of 5-year survival rate for patients treated by surgical means compared to those undergoing radiotherapy only [16]. The adjuvant role of postoperative radiotherapy following radical tumor excision is not yet elucidated, with multiple prospective studies still under way. The results reported by some authors reveal a decrease in the incidence of local recurrences in postoperatively treated patients compared to those who underwent only radical surgery (38% vs. 49%) [20].

There are currently some centers that prefer the combination between preoperative (neoadjuvant) radiotherapy with a complete tumorexcision which seems to decrease the postoperative tumor recurrence, thus lowering the mortality rate [21]. However, the role of radiotherapy and chemotherapy used for adjuvant or neoadjuvant purposes to increase the survival rate remains controversial [11]. Radiotherapy is commonly used for palliative purposes in those for whom surgical excision (total or partial) is contraindicated [22]. The reticence in the wider use of radiotherapy derives from its side effects, primarily on the intestine and bladder.

Despite the fact that liposarcoma is one of the most frequent tumors of soft parts, the molecular mechanisms that are the cause of its pathogenesis are not very well-known yet [23]. This makes the chemotherapeutic treatment be inefficient [24]. The data in the literature based on studies of genetics associate the occurrence and progress of some histological subtypes of liposarcoma with PI3K/ AKT path activation and that is why the therapy focus on this path could become an alternative treatment for such cases in the future [23,25].

Taking into account that, in the case of retroperitoneal liposarcoma, metastasis is generally rare (except for pleomorphic and round cell subtypes), below 33% in some studies, the most frequent cause of death is due to the impossibility of radical tumor excision until healthy tissue. Otherwise the local relapse

is quite common, even after a complete surgical resection [26, 4]. The histologic subtype is very important, 80% of them presenting a local relapse in the first 3 years postoperatively many of them being of undifferentiated type that has the highest recurrence rate [10]. The 5-year survival rate is significantly higher for well-differentiated liposarcoma compared to other subtypes [26].

In our patients case, although we performed a complete surgical excision to healthy tissue, despite the impressive dimensions of the tumor, the histological subtypes present and the fact that the patient refused an adjuvant radiotherapy (as indicated by us at the patient's discharge) contributed to the unfavorable evolution, with local relapse and distal (bone) metastases relatively early (11 months postoperatively) that subsequently led to the patient's death.

## Conclusions

Retroperitoneal gigantic liposarcomas are a challenge even for an experienced surgeon, due to adhesions or invasion in surrounding organs and structures, the full excision of the tumor (which is the gold standard in the treatment of these tumors) is often difficult. The patients survival and prognosis are also closely correlated with the histologic subtype and could be enhanced by adjuvant and neoadjuvant radiotherapy and possibly the development of targeted therapies on the pathophysiological mechanisms responsible for the occurrence of retroperitoneal liposarcoma.

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