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Case Report

Giant Retroperitoneal Liposarcoma—A Renal Hazard

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Abstract: Retroperitoneal tumors are uncommon and may reach a large size prior to causing symptoms or being noticed by the patient or physician. A middle-aged female consulted us for care during her "terminal" illness. She had already undergone four previous retroperitoneal resection surgical procedures. She presented with a large recurrent protruding mass from the right side of the abdomen and related a history of a previous cholecystectomy, right nephrectomy, right colectomy, and repeated resection of a recurrent retroperitoneal liposarcoma. She thus came to us for consultation and terminal care in order to be away from her friends during treatment for this terminal condition. After our consultation, she elected to have repeated surgical excisions of the tumor. The surgical excisions yielded a giant recurrent tumor mass, which overflowed and covered all margins of the 21-inch-wide surgical scrub basin. Over the next eleven years, she had multiple surgical resection procedures involving both the right and left retroperitoneum (a splenectomy, a left colectomy, and a colostomy). Recovery from each of these resection procedures (the final combined resection weight was 120 pounds) was without complications. However, the tumor finally encased the pancreas and the left kidney. If the tumor encasement were to be palliated and resected, she would require hemodialysis. At this time, the patient elected to have no further resection surgeries, no dialysis, nor any palliative chemoradiation treatment. Over a period of sixteen years from her first resection and twelve years from our first resection, the patient had continued to work at her medical administrative and leadership position and led a functional life after our consultation, except for her surgical period. The patient was not cured but benefited from repeated palliative surgeries, prolonging her life and improving her job performance.

Keywords: liposarcoma; giant tumor; retroperitoneal tumor; palliative surgery; quality of life; perirenal tumor; giant retroperitoneal liposarcoma

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1. Introduction

The retroperitoneum is loosely defined, according to Stedman's medical dictionary, as the area located behind the peritoneum within the abdomen and may contain the aorta, inferior vena cava, the kidneys, the ureters, the pancreas, multiple lymph node channels and lymphatics, and retroperitoneal adipose tissue [1]. These systems are surrounded by adipose tissue [2]. Each of these retroperitoneal structures may develop their own pathologies such as an aortic aneurysm, renal tumors, or malignancy. When tumor development does occur, the challenges to both diagnosis and individual therapy are immense.

Sarcomas, one of the retroperitoneal tumors, are, in general, difficult to treat whether they are primary or recurrent and may occur throughout the body. For example, gynecologic leiomyosarcomas are highly malignant and intensely studied in relation to surgery in combination with chemotherapy as adjuvant therapy—with little curative benefits [3]. But a retroperitoneal leiomyosarcoma showed a long period of progression-free survival when

trabectedin was utilized as an adjunct to treat a chemo-refractory metastatic leiomyosarcoma of the retroperitoneum [4]. Whether future drug therapy research will benefit the treatment of primary retroperitoneal liposarcomas (LPS) awaits further study [5].

Retroperitoneal tumors, especially those of multi-centric origin that are malignant, present unique therapeutic difficulties for both the patient and the physician. In order to illustrate some of the treatment difficulties encountered by an individual when maintaining their lifestyle and employment, we present a patient who developed a huge (giant) recurrent retroperitoneal liposarcoma (LPS) that required multiple palliative surgical procedures.

2. Case Study

2.1. Patient

This middle-aged female presented with a history of multiple previous exploratory laparotomies and the resection of a large amount of tumor volume during each procedure. The tumor was again recurring and her surgeon had declined to perform further surgery due to the size and extent of the tumor. During her previous surgical procedures, she had a cholecystectomy, resection of a malignant right colon polyp, resection of the right kidney and ureter, a right adrenalectomy, right colectomy, and multiple resections of the retroperitoneal liposarcoma tumor mass.

2.2. Case History

Over the past several years, the patient had recurrent mild abdominal discomfort, which led to a surgical intervention for a retroperitoneal tumor mass. This required repeated surgical excisional procedures. Unfortunately, the tumor recurred despite the repeated surgical resections and she could now be seen to have a huge abdominal mass protruding from the right side of her abdomen. Her surgeon felt that no additional surgery was indicated and chemotherapy was of little value. Therefore, she sought our care during her final months of life.

2.3. Physical Examination

This middle-aged woman appeared to be alert and active, was employed, and exhibited a sense of gravitas but had a noticeable abdominal bulge occupying the right side of her abdomen. She was in no acute distress but demonstrated some mild abdominal discomfort when lying on her back on the examining table. Upon physical examination, a large, readily palpable, and visible abdominal mass was protruding from the right side of the abdomen. A well-healed long abdominal paramedian incision was noted. No other palpable mass was discovered during the physical examination. The rectal exam was negative and there were no palpable cervical lymph nodes. There was no leg edema.

2.4. Diagnostic Studies

A chest X-ray showed the right diaphragm to be somewhat elevated but no pulmonary metastatic lesions were noted. The hemoglobin was at 10 gm percent. The urinalysis was normal as was the metabolic panel. The pathology reports from the previous operative procedures and repeat slide review suggested that the tumor was a liposarcoma. Our interpretation of her electrocardiogram suggested a previous inferior myocardial infarction. A computerized tomogram of the abdomen showed that the large deviating tumor mass was thinning out at the duodenum.

2.5. Patient Options

The patient had come to us for palliative terminal care and readily discussed this approach. Chemotherapy and radiation were felt to be of little oncologic value in this circumstance. After further discussion, including the options available and possible surgical mortality, she elected to have additional surgery to again debulk the tumor mass.

2.6. Potential Outcome

Having discussed the options with her, our goal was to resect (debulk), but not cure, the huge tumor burden she was experiencing. Our goal was to avoid major injury to the other visceral structures while eliminating or removing as much tumor as possible.

2.7. Actual Outcome

During the surgery, a huge tumor mass involving and deviating towards the right retroperitoneal structures including the colon, duodenum, and vascular structures—anterior and to the left—was found. During surgery, we were able to encircle and remove a major en bloc portion of the tumor (greater than 13.5″ or 35+ cm in cross section and greater than 38″ or 98 cm in diameter) along with a portion of the right colon (Figures 1–3). Her weight, after surgical resection, dropped from 125 pounds to 79 pounds. The excised tumor mass overflowed and covered the entire surgical scrub basin margins. Following an uncomplicated recovery, she regained her weight and returned to work as a supervisor.

The tumor overlapped the surgical basin on all sides.

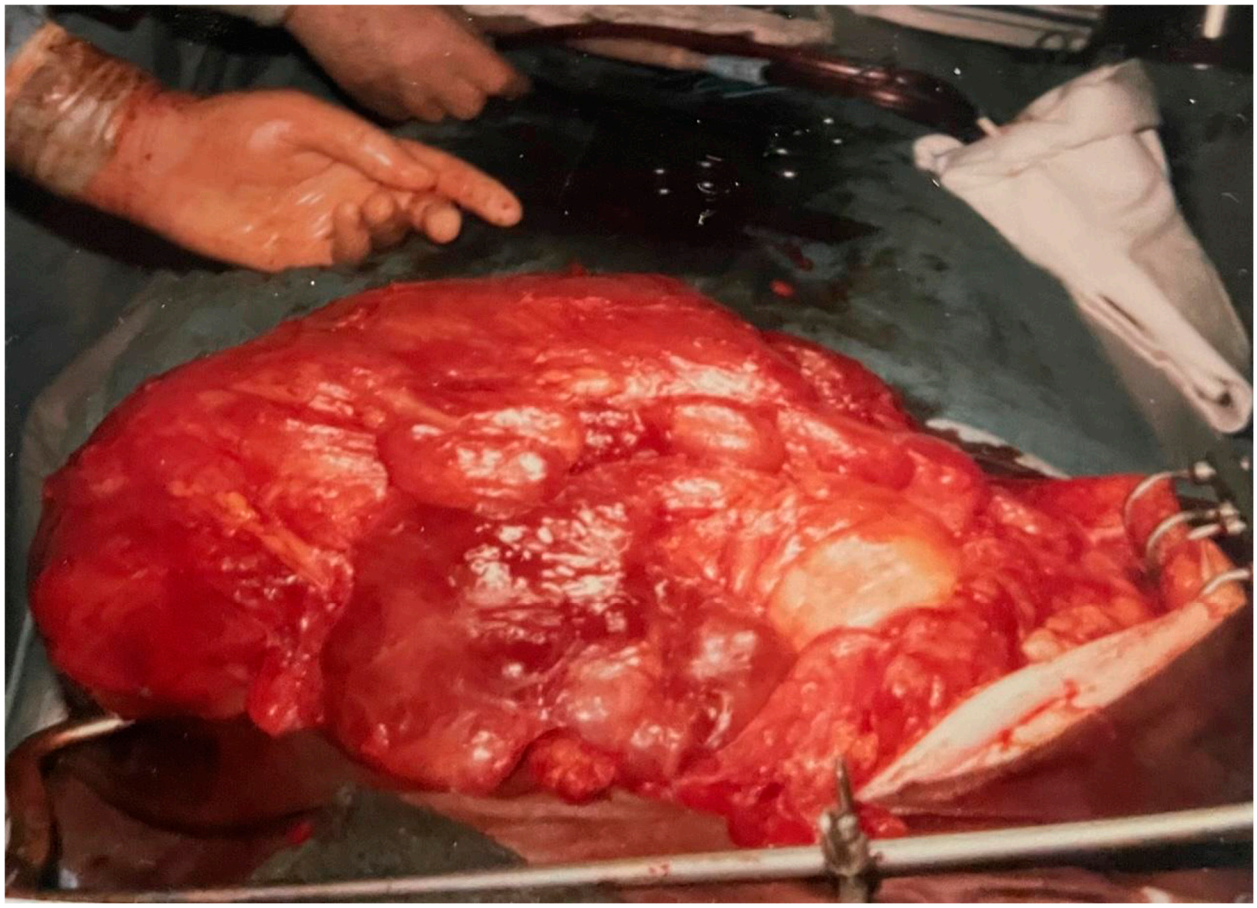

Figure 1. Recurrent Liposarcoma Reresection Specimen.

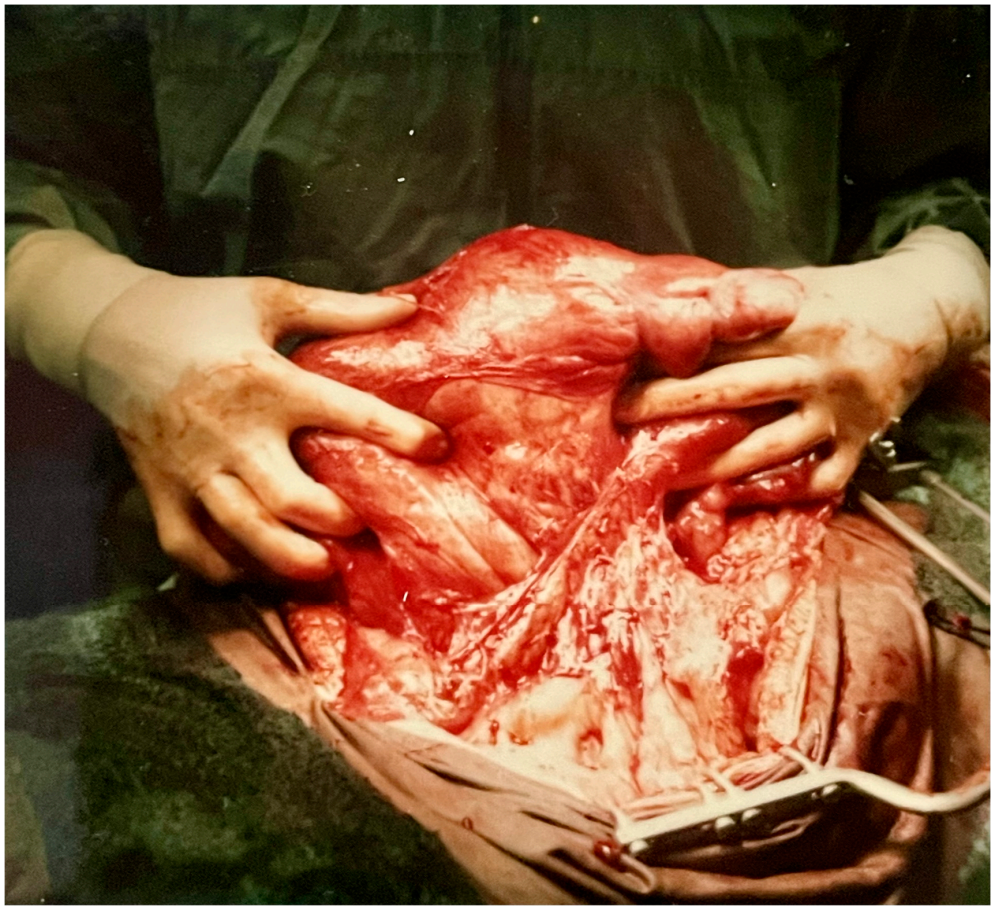

Figure 2. Tumor Retraction for Visibility.

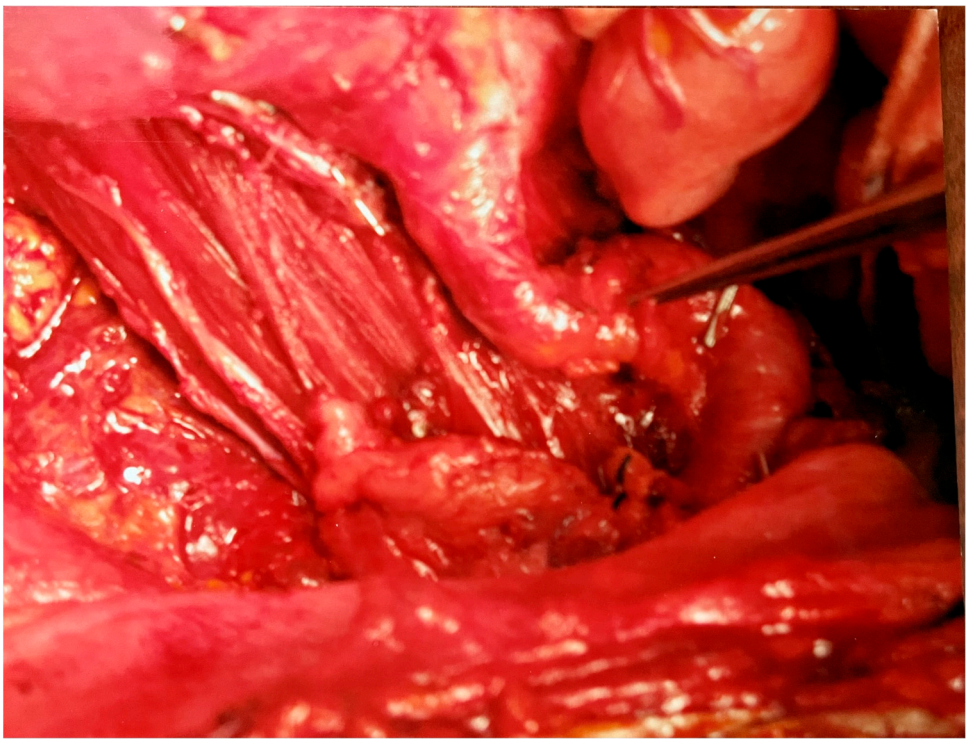

Figure 3. Marked right iliac artery deviation and psoas muscle after tumor resection.

2.8. Long Term Outcome

After a prolonged period of symptom relief, the tumor recurred and surgical resection was again her chosen option. The tumor required repeat surgery on the right two years later and again three years after that. The tumor then progressed to involve the left side of the abdomen and required a splenectomy, a left colectomy, and a left colostomy to complete the palliative "mass" resection. Another two years later, the patient required subsequent surgery after duodenum tumor compression and extensive resection. Subsequently, when the pancreas and left kidney became involved, she then declined further surgery and chronic dialysis.

3. Discussion

3.1. General

This lady developed an uncommon and incurable malignancy. Her initial surgeon, despite repeated efforts, was unable to totally resect the tumor, which involved the right kidney and the right side of the abdomen posterior to the right colon. The retro-peritoneum does not benefit from restrictive "walls" or barriers which might prevent the spread of a tumor, as seen with a benign Schwanoma or an encapsulated tumor lesion, which can aid in surgical resection while preserving other organs (Table 1). Retroperitoneal tumors may be highly symptomatic or unrelated to the cause of the person's complaints. They may be found as the primary symptom producer or as an incidental finding (such as a large lipoma). Symptoms may include pressure-like complaints, loss of appetite, and weight loss for no apparent reason, or a possible mass. And, despite surgery, the LPS tumors may repeatedly recur and encircle/involve other major abdominal organs, as with this patient.

Table 1. A few retroperitoneal masses that may be resectable.

A.	Lymphomas
B.	Adenomas
C.	Liposarcomas (LPS)
D.	Pancreatic Neoplasms
E.	Renal malignancy
F.	Vascular aneurysms
G.	Lipomas

When initially seen by us, this patient had an obvious huge right-sided tumor and a previous known diagnosis of liposarcoma. LPS retroperitoneal malignant tumors generally do not have a capsule or surrounding lining, as is seen with some benign retroperitoneal tumors, that might prevent them from infiltrating and spreading and make "clean" surgical margins possible. Not being radio-sensitive or chemo-sensitive to any degree, most of these tumors are treated with surgical extirpation. But when the tumor is large and infiltrates surrounding structures, a curative resection may not be possible.

Recently, reviews have been written that make suggestions about the diagnosis, classification, management, and treatment of retroperitoneal tumor masses. In addition, a literature review may classify retroperitoneal tumors according to the structure involved, such as the colon or kidney, or the lesion may be classified according to the cell type—including lymphoma, neurogenic, or sarcoma, and liposarcoma. Fortunately, some retroperitoneal tumors, even large ones, may be benign in nature and encapsulated, and therefore resectable. Neurogenic or angiolipomatous tumors, when benign, may be removed entirely by surgery. Others, such as lymphomas, may be biopsied and treated non-surgically with excellent results. The lymphoma class of tumors and most benign tumors may respond to a better degree than an infiltrative liposarcoma. Unfortunately, a few of these "benign" lesions may also remain very resistant to curative therapy. Continued awareness and diagnostic evaluation to delineate and hopefully provide curative treatment remain the goal as radiation and chemotherapy may have little effect—especially in the liposarcoma group.

Retroperitoneal liposarcomas (LPS) occur equally in either sex at near equal occurrence rates and, when small, usually cause no or few symptoms. LPS tumors usually occur in patients between the ages of 40 and 70 years of age. As the LPS grows, the patient may note a slow but steady abdominal girth enlargement and a protrusion of the abdomen to the right, left, or midline. When LPS tumors enlarge, other retroperitoneal structures such as the kidney, pancreas, or the spleen may become encircled and encased by the tumor—as in this patient (Table 2). As the glandular structures become involved, the tumor margins become less distinct and surgical intervention becomes difficult.

Table 2. Organs frequently involved or surrounded by retroperitoneal LPS tumors making curative resection difficult.

A.	Kidney
B.	Adrenal
C.	Gallbladder
D.	Pancreas
E.	Colon
F.	Duodenum
G.	Spleen

3.2. Literature Review

Various authors have studied patients with giant retroperitoneal tumors and have argued that a complete resection is possible, meaning that it has remained the treatment of choice. Bachmann et al. reported that the LPS tumor recurrence rate improved in those patients also receiving radio-chemotherapy [5]. Their tumor resection was complete in nine of ten patients. This was the first study to report the outcome of a giant dedifferentiated retroperitoneal LPS and to conclude that neoadjuvant radiotherapy may improve the rate of survival with LPS. Three of their patients had one organ resected, four patients had two organs resected, and one patient had three organs resected, including the kidneys in four patients, the large intestine in six, the adrenal gland in two, and an orchiectomy in one.

Xu et al. reported that a 65-year-old male patient had a two-year history of progressive abdominal enlargement without other abdominal complaints and a tumor that occupied the entire abdominal cavity measuring 37.0 cm × 32.0 × 26.5 cm and weighing 21.0 kg. Their review of the literature demonstrated 13 other cases with a giant retroperitoneal liposarcoma greater than 30 cm in diameter from 1980 to 2019 [6]. Nine patients were male, the median age was 57, and the patient complaints revolved around an enlarging abdominal girth. Needle biopsies was performed preoperatively and all patients underwent surgery with additional organ resections including seven nephrectomies, one ovariectomy, and one diaphragmatic resection. Six of these patients had well-differentiated and five had poorly differentiated tumors. They advised a preoperative tissue diagnosis, including a possible needle biopsy, of the retroperitoneal tumors for which surgery was being contemplated. They felt CT scans could be reliably utilized, complete tumor resection was the treatment goal, and that size alone was not a contraindication to surgery.

A review of 123 patients with retroperitoneal tumors (Stage I–V) by Chen et al. showed that the early tumor stages were associated with a better overall tumor-free survival [7]. A renal resection was the most common associated organ resection followed by the small intestine, spleen, and pancreas. Twelve patients required two or more organs to be en bloc resected. The length of survival was greater in patients who did not require an additional organ resection. Stage-three patients were 100% three- and five-year survivors while there were no stage-four survivors at three or five years. The risk factors for recurrence are shown in Table 3. Their goal of "total resection" was felt to be an important prognostic event, while tumor stage was an independent predictor of survival longevity.

Table 3. Risk factors for LPS recurrence.

A.	Age
B.	Tumor size
C.	Degree of differentiation
D.	Tumor stages
E.	Resection

Moyon et al. reported a 34-year-old patient with a growing LPS mass in her abdomen, severe weight loss, and recurrent postprandial vomiting [8]. The encapsulated retroperitoneal mass measured 40 × 28 × 10 cm. Lucia Casadei et al. focused on well-differentiated LPS and their current therapies, including chemotherapy strategies [9]. They presented a review of 13 ongoing trials related to these tumors, radiotherapy strategies for such tumors, and a review of the use of molecular therapy for LPS treatment—especially for recurrent LPS cases.

Xiao et al. listed five liposarcoma subtypes and stated that the well-differentiated type may convert to a dedifferentiated LPS and that the final diagnosis depends on the pathologic findings, computed CT, and magnetic resonance (MRI) (Table 4) [10]. They found that many LPS are asymptomatic prior to diagnosis and that a large amount of fat on imaging suggests LPS. They felt that the main treatment for retroperitoneal LPS, after diagnosis, was surgery and stated that survival and prognosis is highly dependent on the surgical approach and histologic subtype. In their study, the low-grade LPS five-year survival was over 90% while the survival of patients with a poorly differentiated LPS may be down to 50%. Half of the patients with a tumor less than 15 cm had no symptoms, while only 33% of patients with large tumors over 25 cm had no complaints. The most common complaints noted were abdominal distension, mass formation, and tenderness. They concluded that the most important prognosis factors included the LPS subtype, presurgical LPS size, and whether the patient was symptomatic or not. Well-differentiated LPS had the best prognosis, and pleomorphic LPS the worst prognosis. Resection of all tumors is the best treatment for all LPS subtypes while chemotherapy and radiotherapy are less effective alternative choices.

Table 4. Five liposarcoma subtypes [10].

I.	Well differentiated LPS—most common
II.	Myxoid—second most common
III.	Pleomorphic—highly malignant
IV.	Mixed LPS
V.	Dedifferentiated

Nassif et al. have discussed advances in the management and treatment of LPS [11]. After reviewing the LPS subtypes and frequency, they reviewed the unique biomarkers found in relation to LPS and their incorporation in future treatment protocols. In their opinion, neoadjuvant radiation therapy given preoperatively was associated with an increased survival and local control, especially for myxoid LPS treatment. They also reported the MRCL (the myxoid subgroup of LPS) showed a favorable response in some patients while advocating for further drug therapy studies [9].

What we have described in this study is an example of the type of highly malignant tumors which may occur in the retroperitoneum and the effect they may have on the patient. The tumors may be carcinomatous or sarcomatous in nature and occur in adults or children. An example of the difficulty in treating and curing such a retroperitoneal tumor is the clear cell sarcoma of the kidney, which may occur in children, as presented by Brillantino et al. [12]. Malignant or benign retroperitoneal tumors present a challenge for the patient and the physician.

3.3. Patient Discussion

This female patient primarily suffered weight loss and discomfort while physically showing a large abdominal tumor mass. During her previous surgery and over the decade during which we treated her, we were unable to obtain a curative resection. The process eventually progressed from the right to the left side of the abdomen for which a splenectomy, colectomy with colostomy, repair of the bowel, lysis of adhesions, and resection of another 20 pounds of tumor were performed. When the situation reached the stage of a possible left renal resection, pancreatectomy, and chronic renal dialysis, she declined further intervention. Until that time, however, she was able to function and work every day except for during her periods of hospital treatment.

During the multiple surgical procedures, this patient had approximately 120 pounds of liposarcoma removed. The photographs demonstrate the enormity of one of the tumor resections and the surgical difficulty encountered during the resection. We were able to monitor the tumor progress palpably and with recurrent X-rays, computerized tomography scanning, and ultrasounds, and to periodically palliate the disease process surgically, but we were not able to cure the tumor. None of the referenced articles discussed a loss of renal function due to tumor encroachment as being related to the demise of the patient.

4. Conclusions

This patient was aware of her diagnosis (a huge recurrent liposarcoma of the retroperitoneum) and prognosis when initially seen by us for palliative care. But, after some discussion, despite her diagnosis, we were able to provide options and a treatment program enabling her to return to productive employment for more than a decade. She elected to follow a palliative course, which provided her with many years of fruitful employment as well as the enjoyment of her family and friends. Despite her diagnosis and the rigors of treatment, as exemplified by the photographs, she had very positive surgical palliative results until the potential need for chronic dialysis after the completion of a nephrectomy and pancreatectomy.

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Informed Consent Statement: Because the patient passed away and his family members could not be contacted, this study is only a summary of the patient's entire clinical treatment process, and confidentiality is guaranteed as much as possible.

Data Availability Statement: The data presented in this study are available on request from the corresponding author.

Conflicts of Interest: The authors declare no conflict of interest.

References

1. Pugh, M.B. (Ed.) *Stedman's Medical Dictionary*, 27th ed.; Lippincott Williams and Wilkins Company: Baltimore, MD, USA, 2000; p. 1563.
2. Netter, F.H. *Atlas of Human Anatomy*; CIBA—Geiger Corporation, pharmaceutical division: West Caldwell, NJ, USA, 1989; Volume 165, pp. 257, 315.
3. Rizzo, A.; Nannini, M.; Astolfi, A.; Indio, V.; De Iaco, P.; Perrone, A.M.; De Leo, A.; Incorvaia, L.; Di Scioscio, V.; Pantaleo, M.A. Impact of Chemotherapy in the Adjuvant Setting of Early Stage Uterine Leiomyosarcoma: A Systematic Review and Updated Meta-Analysis. *Cancers* **2020**, *12*, 1899. [CrossRef] [PubMed] [PubMed Central]
4. Reichinger, A. Long-Time Progression-Free Survival with Trabectedin in Chemorefractory Metastatic Leiomyosarcoma of the Retroperitoneum: A Case Report. *Case Rep Oncol.* **2023**, *16*, 1013–1019. [CrossRef] [PubMed] [PubMed Central]

5. Bachmann, R.; Eckert, F.; Geleert, D.; Strohaker, J.; Beltzer, C.; Ladurner, R. Perioperative strategy and outcome in giant retroperitoneal dedifferentiated liposarcoma—Results of a retrospective cohort study. *World J. Surg. Oncol.* **2020**, *18*, 296. [CrossRef] [PubMed]
6. Xu, C.; Ma, Z.; Zhang, H.; Yu, J.; Chen, S. Giant retroperitoneal liposarcoma with a maximum diameter of 37 cm: A case report and review of literature. *Ann. Transl. Med.* **2020**, *8*, 1248. [CrossRef] [PubMed]
7. Chen, J.; Hang, Y.; Gao, Q.; Huang, X. Surgical diagnosis and treatment of primary retroperitoneal liposarcoma. *Front. Surg.* **2021**, *8*, 672669. [CrossRef] [PubMed]
8. Moyon, F.X.; Moyon, M.A.; Tufino, J.F.; Yu, A.; Mafla, O.L.; Molina, G.A. Massive retroperitoneal dedifferentiated liposarcoma in a young patient. *J. Surg. Case Rep.* **2018**, *10*, rjy272. [CrossRef]
9. Casadei, L.; Costas Casal de Faria, F.; Lopez-Aguiar, A.; Pollock, R.E.; Grignol, V. Targetable pathways in the treatment of retroperitoneal liposarcoma. *Cancers* **2022**, *14*, 1362. [CrossRef] [PubMed]
10. Xiao, J.; Liu, J.; Chen, M.; Liu, W.; He, X. Diagnosis and prognosis of retroperitoneal liposarcoma: A single asian center cohort of 57 cases. *J. Oncol.* **2021**, *2021*, 7594027. [CrossRef]
11. Nassif, N.A.; Tseng, W.; Borges, C.; Chen, P.; Eisenberg, B. Recent advances in the management of liposarcoma. *F1000Research* **2016**, *5*, 2907. [CrossRef] [PubMed] [PubMed Central]
12. Brillantino, C.; Rossi, E.; Minelli, R.; Irace, D.; Castelli, L.; Zeccolini, R.; Bignardi, E.; Tufano, A. A rare case of renal tumor in children: Clear cell sarcoma. *Il G. Chir.-J. Ital. Surg. Assoc.* **2019**, *40*, 217–224. [PubMed]