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Challenges in the treatment of a giant retroperitoneal liposarcoma

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Challenges in the treatment of a giant retroperitoneal liposarcoma

BACKGROUND: *Retroperitoneal soft-tissue sarcoma is a very rare neoplasm, the most frequent histological subtype is liposarcoma with up to 45% of all cases. Unspecific clinical presentation, late diagnosis and high local recurrence rate represent important problems in clinical practice. We present the case of an adult patient with an unusual large liposarcoma of the retroperitoneum analyzing diagnostic workup, surgical approach and therapeutic strategies.*
CASE REPORT: *A 68-years old female was admitted with weight gain (+12 kg) and increasing abdominal girth. Computed tomography scan imaging showed a retroperitoneal tumor with 40 cm maximum diameter. Biopsy revealed a myxoid liposarcoma. The interdisciplinary curative surgical treatment included preoperative ureteral splinting, en-bloc tumorexstirpation, ileocecal resection, right ureteral resection and vascular reconstruction of the Arteria iliaca communis. The postoperative course was uneventful. After sixteen months the patient developed multifocal local recurrence requiring extensive surgical resection of tumor and retroperitoneal fat (Figs. 3, 4). However, thirteen months later the tumor reappeared and the patient was assigned to palliative chemotherapy. The patient is still alive with stable tumor disease.*
CONCLUSION: *The removal of a huge retroperitoneal sarcoma is a significant challenge for the surgeon. Accurate planning, interdisciplinary treatment options, and radical surgery are essential. However, the recurrence risk is exceptionally high because of the enormous tumor dimensions and the big tumor surface, multimodal therapeutic approaches may improve the outcome in these patients.*

KEY WORDS: Liposarcoma, Retroperitoneum, Surgery

Introduction

Adult soft tissue sarcomas are rare tumors with an estimated incidence of 4-5/100000/year in Europe, they represent 1-2% of all solid tumors. The majority of soft tissue sarcomas affect the extremities, only 10-20% originate in the retroperitoneal space [1-3]. Retroperitoneal soft tissue sarcomas are sporadic tumors with many different histological subtypes, the most frequent histological subtypes are liposarcoma (20% of all soft tissue sarcomas in adults), leiomyosarcoma, and malignant fibrous histiocytoma [4]. The peak incidence is detected between 50–70 years [5]. It has been reported that 20% of these tumours are greater than 10 cm at the time of diagnosis [6]. Depending on tumor grading (well differentiated vs. undifferentiated) and location mortality rates in patients with liposarcoma vary between 1% to 90%, recurrence rates range from 5% to 83% [4]. We report on a challenging unusual case of a giant retroperitoneal liposarcoma managed by interdisciplinary resection (patient´s informed consent was obtained for publication), and analyse preoperative workup and options of surgical treatment.

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Case Report

A 68-year-old slim female (Body Mass Index 20, history of appendectomy and cholecystectomy) presented in a peripheral hospital with rapid weight gain (12 kg) and increase of abdominal girth within a few months. On clinical examination, the whole abdomen was hard and stiff, the laboratory examinations were normal. A computed tomography scan demonstrated the presence of a giant mass of about 40 cm maximum diameter from the subhepatic space to the pelvic cavity displacing and rotating the right kidney to the diaphragm and the small bowel to the left hemiabdomen (Fig. 1 A-C). The tumor tissue appeared heterogeneous with areas of different density, consistent with the radiological diagnosis of a giant liposarcoma without metastases, originating from the right retroperitoneum. After percutaneous biopsy and histological diagnosis of a myxoid liposarcoma, the patient was referred to the university hospital. The multidisciplinary tumorboard recommended radical oncological surgery; preoperatively the patient underwent prophylactic bilateral ureteral splinting to facilitate intraoperative identification of the ureter. The giant liposarcoma was resected successfully by an interdisciplinary team (visceral surgeon, vascular surgeon and urologist) en bloc and in toto including ileocecal resection, right ureteral resection (terminoterminal anastomosis) and partial right A. iliaca communis resection (goretex patch reconstruction). The natural intraoperative aspect of the tumor is depicted in Figure 2. The tumor weighed 12.7 kg and measured 40 x 30 cm, the postoperative Body Mass Index was 15. The histological exam showed - as far as possible to assess in such a case - a surgical resection in sano (R0), no tumor infiltration in the resected bowel and negative presacral lymphnodes. The postoperative course was uneventful, the tumorboard recommended regular follow-up. Sixteen months later the periodical computed tomography scan showed local recurrence with three tumor nodules with a maximal diameter of 10 cm in liver hilus, small bowel mesentery and right lower abdomen. The multidisciplinary tumorboard recommended attempting radical oncological surgery again. The three tumor nodules could be resected successfully including the complete, but sparse surrounding adipose tissue (R0 resection) (Fig.4). Histology showed both dedifferentiated and well-differentiated areas of liposarcoma. After another twelve months of follow-up, at computed tomography scan a local recurrence was found again. According to the patient´s wish, surgery was not taken into account anymore and a palliative chemotherapy at the referring hospital was initiated. After two cycles of doxorubicin monotherapy, the disease was progredient and ifosfamide was started. At the last follow up exam, 52 months after the first operation, the patient presented in satisfying conditions with stable disease.

Discussion

Retroperitoneal malignant neoplasms are very rare, unfortunately, diagnosis is difficult and often delayed because of unspecific symptoms. In contrast to other locations like extremities or head and neck, tumors arising from the retroperitoneum can grow for a long time occupying the whole abdominal cavity before developing symptoms. Pain is the symptom most complained in about 60% of cases, other clinical manifestations may be nausea, hydronephrosis, haematuria, dysuria, bleeding, peripheral paralysis, thrombosis, oedema of the lower limbs, weight loss, and fever [2,4,6]. Also, very rare and unusual clinical presentations like hernias have been reported in literature.[7] In the present case, only weight gain and increasing abdominal girth were observed, two rather unspecific and trivial symptoms.

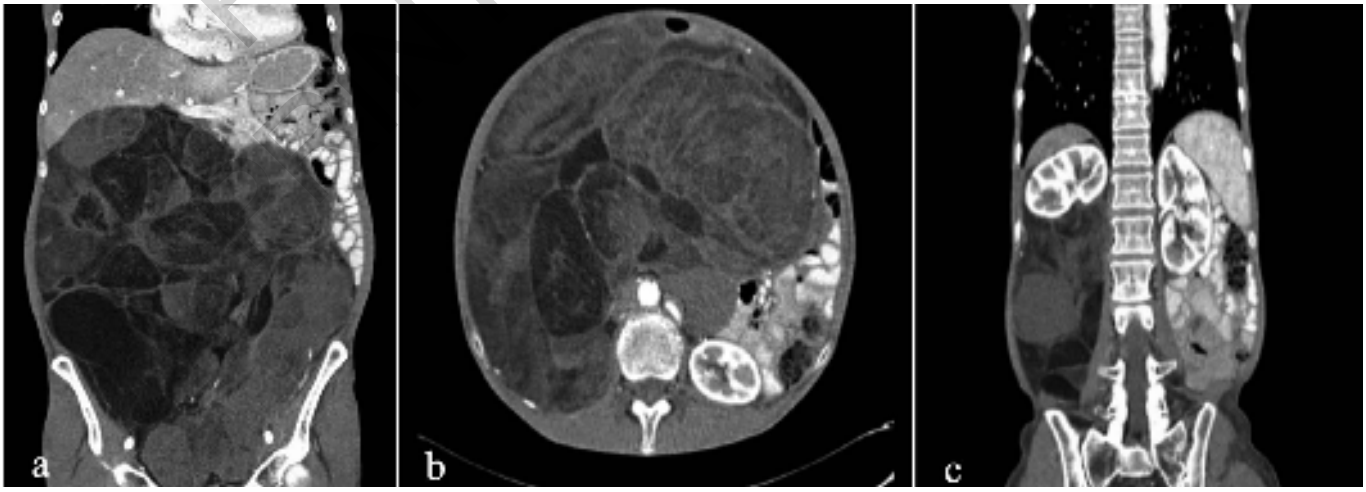

Fig. 1: A,C) Coronal and B) axial preoperative contrast-enhanced abdominal computed tomography scan showing the giant retroperitoneal liposarcoma going from the subhepatic region to the pelvic cavity (40cm x 30cm) displacing and rotating the right kidney to the diaphragm.

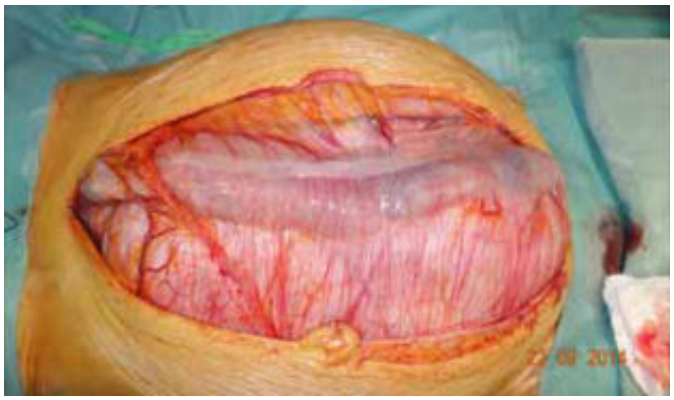

Fig. 2: Natural intraoperative aspect of the tumor at the beginning of surgery with the stretched right hemicolon displaced left to the median line.

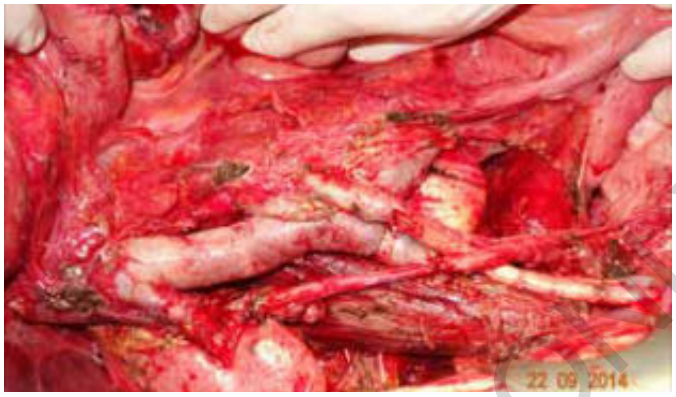

Fig. 3: Intraoperative situs after tumor resection and reconstruction of ureter and A. iliaca comm.

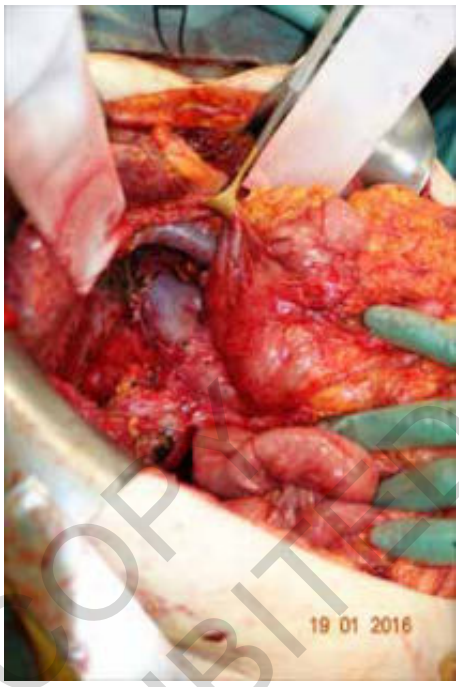

Fig. 4: Intraoperative situs after resection of the local recurrence in the liver hilus between V. cava and V. portae.

Concerning diagnosis, computed tomography scan is essential to analyse tumor's dimensions and exclude metastases, especially in the lung. Magnetic resonance imaging is indicated to gain additional information about subtyping or local invasion [8]. The preoperative knowledge about adjacent, adherent or invasive tumor may help to plan the extent of surgery and involve other specialists like urologist, vascular surgeon or gynaecologist. In case of eventual involvement of the kidney, functional assessment by kidney scintigraphy can be done to facilitate the decision on nephrectomy. Following appropriate imaging assessment, some guidelines recommend performing of multiple core needle biopsies. The risk of needle track seeding is minimal and should not be a reason to avoid a biopsy [8]. However, preoperative biopsy is still a matter of debate and many surgeons support biopsy only if the histological exam is connected with a clinical consequence like preoperative chemotherapy or radiation.

The only curative treatment of a retroperitoneal sarcoma is complete radical resection (R0) [5]. This, however, can be difficult to achieve, especially in giant tumors with limited surrounding healthy tissue and close proximity to critical structures. Tumor size, age, histological subtype, vascular invasion, resection status (R0) and necessity of multivisceral resection (≥ 3 organs) were shown to correlate with local recurrence rate and prognosis. However, especially the last parameter, the extent of multivisceral resection, is hard to anticipate preoperatively, and thus, the success of surgery is not easily guaranteed [6,9-12]. The recurrence rate in case of large tumors amounts to 52%, in contrast, only 15% of tumors develop distant metastases [9]. Previous analyses showed significantly higher 5-year overall survival rates for patients with R0 resection compared to R1 resection (93% and 45%, respectively) [9-12]. The rate of negative resection margins generally ranges from 42 to 76% in literature. Repeated resections of tumor manifestations in case of recurrence are possible, but the probability of a R0 resection decreases with relapses: the R0 resection rate after the first recurrence surgery amounts to 57% and declines to 20% after the second recurrence and 10% after the third recurrence. This data reflects the technical difficulty associated with multiple abdominal operations [13-15]. Thus, on the one hand, initial complete radical surgery is essential and should be pursued at any price, on the other hand multivisceral resections, especially including kidney, left hemicolon, pancreas, stomach and big vessels have been shown to significantly increase morbidity [10,13]. In summary, the approach to the organs and structures neighbouring a retroperitoneal sarcoma is crucial regarding morbidity, prognosis and local recurrence. Organ preserving and simple excision

of the tumor has been shown to be inferior to more aggressive surgical strategies such as liberal multivisceral compartmental resection of contiguous organs even if uninvolved [13,16-19]. The main surgical problem in giant retroperitoneal sarcomas is that almost all intraabdominal organs are somehow affected from the large tumor and the large tumor surface. Additionally, even a compartmental resection may be difficult, because the tumor displaces all surrounding tissue and there remains only little healthy tissue. We suggest, that resection of important organs like kidney, pancreas or liver should be evaluated on individualised basis, weighing the potential for local control or local recurrence against possible long-term dysfunctions. In our case, a complete radical resection with removal of the whole compartment was attempted, the right kidney however was leaved in situ, because of patient´s suboptimal kidney function and patient´s desire.

Although data of studies of neoadjuvant therapy versus resection alone for retroperitoneal soft tissue sarcomas have shown inconsistent results, neoadjuvant treatment options like chemotherapy or radiotherapy may be considered in carefully selected patients [20]. The goal of neoadjuvant therapy approaches is not a change of the surgery extent, but an improvement of the quality of the resection margins. Adjuvant and intraoperative radiotherapy is of limited importance, because associated with significant short- and long-term toxicities, adjuvant chemotherapy is not established routinely [8]. The patient in this case report was not a candidate for radiotherapy or intraoperative radiotherapy because of the large tumor dimensions, a palliative chemotherapy was initiated after diagnosis of the second local recurrence, since surgery was declined by the patient. The current mainstay of systemic treatment for patients with metastatic or unresectable disease is doxorubicin with or without ifosfamide in the first-line setting, however in dedifferentiated tumors the survival benefit is negligible [8,21]. The postoperative surveillance should include regular computed tomography scans every 3 or 6 months to prematurely detect asymptomatic local recurrence and evaluate the possibility of repeated surgical options.

Conclusion

The treatment of a giant retroperitoneal liposarcoma is challenging because of the extremely large surface area of the tumor, which involves many surrounding structures and promotes a high rate of local recurrence. Multimodal treatment options are limited, the only curative treatment is complete radical surgery including compartmental resection or in the majority of cases multivisceral resection.

Riassunto

Il sarcoma retroperitoneale è un tumore molto raro, il tipo istologico più frequente (45 %) è il liposarcoma. Per l'assenza di sintomi specifici e per la diagnosi spesso tardiva si tratta frequentemente di lesioni piuttosto voluminose, in grado di raggiungere dimensioni notevoli occupando gran parte dell'addome e dislocando organi e strutture circostanti. La terapia standard è l'asportazione chirurgica completa del tumore, che determina significativamente la sopravvivenza a lungo termine. Il rischio oncologico più importante consiste nello sviluppo di una recidiva locale. Progettazione accurata (con o senza biopsia preoperatoria) e resezione radicale, preferibilmente una resezione dell'intero compartimento retroperitoneale coinvolto, rappresentano gli elementi centrali dal punto di vista del chirurgo. Nel caso clinico proposto, viene descritto il decorso di una paziente con un grande liposarcoma retroperitoneale con un diametro di 40 cm trattato con resezione multiviscerale interdisciplinare. In base a questo caso analizziamo e discutiamo l'iter diagnostico e terapeutico di sarcomi retroperitoneali giganteschi.

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