Case Report

Giant retroperitoneal liposarcoma: A case report and literature review

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Abstract

Retroperitoneal liposarcomas are a heterogeneous group of mesenchymal tumours that have a wide spectrum of histological subtypes and vague clinical presentations.

Introduction

Liposarcomas are rare malignant tumours that commonly originate in the thigh or retroperitoneum.1 Their worldwide incidence varies between 0.07% and 0.2% of all retroperitoneal tumours.2 The estimated incidence of liposarcoma in Europe is 4–5/100,000/year; however, in
Germany, these are amongst the most common malignant soft tissue tumours in adults. Retroperitoneal liposarcomas (RPSs) present at a median age of 56 years, but they have been reported to occur at all ages (2–98 years). The gender distribution of RPSs is approximately equal, although some studies have reported female predominance.

The European Society for Medical Oncology (ESMO) has identified certain predisposing factors for RPS: genetic aberrations, ionizing radiation exposure, particularly in children undergoing repeated computed tomography (CT) and those with diabetes mellitus. Approximately 20%–25% of the patients with RPS have been reported to have diabetes mellitus; the co-existence of these two diseases has been associated with adverse prognosis. Interestingly, most RPSs rarely metastasise, even when they grow to a large size. According to the ESMO, although they are malignant lesions, only about 10% of these tumours are found to have metastasised to distant organs, particularly the lungs and liver, through haematogenous spread. The pathological diagnosis of RPSs should include morphology, molecular pathology, and immunohistochemistry. RPSs are broadly classified into four histologic subtypes: well-differentiated, myxoid, pleomorphic, and dedifferentiated. Dalal et al. have found that the well-differentiated subtype is the most common variant of RPS (46%), followed by the myxoid (28%), dedifferentiated (18%), and pleomorphic subtypes (8%).

As RPSs grow in an expandable space, they attain a large size before being discovered as a palpable lump or as the cause of increasing abdominal girth or gastrointestinal, urologic, or neurological compression symptoms. This is in sharp contrast with hollow viscus malignancies which manifest relatively during luminal obstruction or intra-luminal bleeding. Approximately 60%–80% of the patients present with a palpable abdominal mass and half of the patients have vague abdominal pain at presentation. RPS is probably one of the largest neoplasms in the human body, invariably measuring >5 cm when diagnosed.

Herein, we report the successful management of a giant RPS, weighing 11.6 kg, in a 75-year-old man, at the Department of Surgery of the Hospital Reinbek St. Adolf-Stift, Germany. We aim to convey an important message to physicians to keep a high index of suspicion for RPS in patients presenting with vague abdominal symptoms. We also provide an update on the diagnostic and management guidelines for RPS.

Case report

A 75-year-old man presented to the Surgical Clinic of the Department of Surgery of the Hospital Reinbek St. Adolf-Stift, Germany, with complaints of rapid weight loss of 7 kg, reduced appetite, and a growing abdominal circumference within the previous 6 weeks. The patient also experienced intermittent constipation. He had a history of bilateral hip arthroplasty due to arthrosis and a left-sided inguinal hernia repair. Physical examination revealed a non-tender, distended abdomen with a palpable mass extending from the epigastrium to the pelvic region. Routine

Figure 1: Preoperative CT scan of the giant liposarcoma. Sagittal (A), coronal (B, C), and axial images (D, E); *left kidney.
biochemical parameters were within normal limits. Abdominal ultrasound revealed a heterogeneous soft tissue mass that filled nearly the entire abdominal cavity. CT of the abdomen showed a giant heterogeneous mass, measuring 31 × 23 × 30 cm, leading to the displacement of the left kidney, descending colon, the small bowel loops, and the mesenterial axis (Figure 1). No ascites, locoregional or distant metastases, or calcifications were observed. Staging was performed using endoscopy and the findings were unremarkable.

In consultation with our multidisciplinary tumour board, it was decided that exploration and surgical resection of the tumour be performed. Laparotomy was performed using the midline approach; an enormous, lobulated, retroperitoneal mass presented (Figure 2). After resection of the tumour mass, the left kidney was also removed as it appeared to be infiltrated by the tumour. Furthermore, multiple nodular lesions were observed on the left hemi-diaphragm. Partial diaphragmatic resection was performed with subsequent reconstruction.

On macroscopic examination, the tumour’s dimensions were 35 × 29 × 20.5 cm and it weighed 11,685 g. The cut section showed a yellow, lobulated tumour with a large, central necrotic area of 19 × 10 × 25 cm, comprising approximately 70% of the tumour mass (Figure 3). The attached diaphragm and muscles were infiltrated by the tumour. Examination of the left kidney revealed infiltration of the capsule without any infiltration into the parenchyma. Histopathological examination revealed a lipomatous tumour with a low-grade, dedifferentiated liposarcoma. Apart from well-differentiated areas with only scarce atypical adipocytes and lipoblasts, regions with spindle cell, pleomorphic, or myxoid/chondroid components were also observed (Figure 4). Mitotic activity was low, with a Ki67 proliferating index of 5%−10%. The tumour showed typical MDM2 gene amplification.

The patient had an uneventful recovery and was discharged on the twelfth postoperative day. His follow-up plan consisted of visits every 3 months for 2 years. During the short-term follow-up, the patient remained free of pain, and his physical condition improved. Additionally, the physical examination and abdominal ultrasound were unremarkable.

Discussion

We present a case of an exceptionally large RPS which was diagnosed by abdominal CT and was successfully removed. The tumour had infiltrated into the adjacent left kidney and hemi-diaphragm, which necessitated nephrectomy and partial excision of the diaphragm. Postoperatively, the patient had uneventful recovery. Our case is similar to other reported cases in terms of non-specific presentation; however, an RPS with such enormous weight and dimensions has not been previously reported.

According to the Transatlantic Retroperitoneal Sarcoma Working Group (TARPSWG), abdominal ultrasound can be the initial imaging test, but CT or magnetic resonance imaging should be performed in all cases. The TARPSWG has emphasized the value of collaboration among various disciplines in the treatment of RPSs. Thus, multidisciplinary team work involving surgeons, pathologists, imaging specialists, radiation therapists, and...
medical oncologists is recommended. In our case, CT of the abdomen and pelvis was helpful in diagnosing RPS. Thereafter, the case was managed under the guidance of the multidisciplinary tumour board of the hospital.

Percutaneous biopsy is recommended prior to surgery for suspected RPS; it can also guide the surgical approach and follow-up protocol. According to the ESMO recommendations, following an appropriate imaging evaluation, the standard approach to RPS diagnosis includes multiple core-needle biopsies using ≥14–16 G needles. However, such biopsies could also jeopardize the accurate estimation of malignancy in large RPSs. In our case, neither fine-needle nor core-needle biopsy was performed prior to surgery.

Complete surgical resection is the gold standard in the treatment of RPSs. There are no size restrictions that would contraindicate a surgical resection. Our case also validates the observations that even enormous RPSs may be surgically resected under the guidance of a multidisciplinary team, which is consistent with the consensus management of liposarcomas in adults. The significance of an R0 resection was first demonstrated by Lewis et al. in 1988. In their cohort of 500 patients with RPS, the median survival after an R0 resection was 103 months compared to 18 months after an R1 or R2 resection. In a review by Zeng et al., the long-term 5- and 10-year survival rates without complete resection were described as 16.7% and 8.0%, respectively. This reaffirms the curative role of surgical excision of RPS.

Due to the wide spectrum of histological subtypes with different biological behaviours, a standard recurrence rate of RPSs cannot be determined. In a review by McCallum et al., the researchers investigated the correlation of the local recurrence and distant metastasis rates with tumour size. The estimated 5-year metastasis rate in tumours <2.5 cm
was approximately 3%, whereas in tumours ≥20 cm, the rate was up to 60%. Recently, neoadjuvant chemotherapy as a radiosensitiser in combination with radiotherapy has been described. In the recently reported phase I/II clinical trial, researchers investigated the long-term outcomes of 83 patients with RPS who underwent three cycles of ifosfamide along with radiotherapy before surgical excision of the tumour. This trial endorsed the effectiveness of a combination therapy; however, the long-term outcomes of the treated patients remained poor. Local recurrence of RPSs, particularly within the first 6 months to 2 years, was found to be more common than distant metastasis.

The consensus guidelines by the TARPSWG for the management of recurrent RPS recommend a careful review of the preoperative imaging, operative notes, and histological findings by a multidisciplinary team. Unfortunately, the outcome and prognosis deteriorate with each surgical resection with a concomitant increase in morbidity. Thus, oncologists should weigh the risks and benefits when managing recurrent RPSs. The prognosis of dedifferentiated liposarcomas is better than that of the pleomorphic type, but worse than that of the atypical lipomatous tumours/well-differentiated liposarcoma. These variations are influenced by the different biological behaviours of liposarcomas.

Conclusion

Our case suggests that dedifferentiated liposarcomas should be treated by radical surgical excision, with multi-visceral resection if required. A high index of suspicion for RPS should be maintained in patients presenting with non-specific abdominal symptoms. A multidisciplinary tumour board should be consulted before embarking on any intervention. Surgery remains the cornerstone of curative therapy for intraabdominal liposarcomas.

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Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

The authors declare, that the involved patient gave his informed consent for participation in research. The study was done according to the declaration of Helsinki.

Authors contributions

JH conceived and designed the study, conducted research, provided research materials, and collected and organized, analyzed and interpreted data. KN and HH performed surgery and postoperative follow-up and collected data. KHU performed histopathology examination and provided research materials. SYG and TS wrote final draft of article, and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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