



Giant retroperitoneal liposarcoma: case report and review of literature

Gigantski retroperitonealni liposarkom: prikaz primera in pregled literature

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ABSTRACT

Retroperitoneal liposarcoma (RPLS) is a rare malignant tumour representing 40–50% of retroperitoneal sarcomas, with an incidence of less than one case per 100,000 people per year. Most cases are diagnosed between the ages of 40 and 60. Because the retroperitoneal space allows tumours to grow silently, RPLS are often only discovered once they have reached a considerable size. ‘Giant’ RPLS, defined as tumours exceeding 30 cm or 20 kg, are exceptionally uncommon, with only a limited number of cases reported worldwide. The prognosis is mainly determined by the histological subtype and the extent of resection, while tumour size primarily indicates the complexity of the surgery. We present the case of a 52-year-old male patient with a giant well-differentiated retroperitoneal liposarcoma measuring 53 cm.

IZVLEČEK

Retroperitonealni liposarkom (RPLS) je redek maligni tumor, ki predstavlja 40–50 % retroperitonealnih sarkomov, z incidenco manj kot en primer na 100.000 prebivalcev na leto. Večina primerov je diagnosticirana med 40. in 60. letom starosti, retroperitonealni prostor pa omogoča tiho rast, zato so RPLS pogosto odkriti šele, ko dosežejo precejšnjo velikost. ‘Gigantski’ RPLS, opredeljeni kot tumorji večji od 30 cm ali težji od 20 kg, so izjemno redki, v strokovni literaturi so opisani redki primeri. Prognozo določata predvsem histološki podtip in radikalnost resekcije. Tehnična zahtevnost operacije je odvisna predvsem od velikosti tumorja. Predstavljamo primer 52-letnega moškega z gigantskim dobro diferenciranim retroperitonealnim liposarkomom velikosti 53 cm.

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INTRODUCTION

Retroperitoneal liposarcoma (RPLS) is a rare mesenchymal malignancy arising in the retroperitoneal space. Because the retroperitoneum can expand without early symptoms, tumours often present late with abdominal distension or compression of adjacent organs [1].

‘Giant’ RPLS, usually defined as tumours larger than 30 cm in diameter or weighing more than 20 kg, are exceptional. Only 34 cases with diameters greater than 30 cm had been reported in the English literature so far [1]. A systematic review of 157 cases emphasised their rarity and the technical challenges involved in treatment [2].

According to the 2020 WHO classification, liposarcomas are divided into well-differentiated (WDLPS), dedifferentiated (DDLPS), myxoid, pleomorphic, and myxoid-pleomorphic variants. Subtype strongly influences biological behaviour, with WDLPS prone to local recurrence but low metastatic risk, and DDLPS or pleomorphic variants showing more aggressive behaviour [1, 2].

Surgery remains the cornerstone of treatment, with complete resection offering the best chance of long-term survival [1-3]. For giant tumours, achieving negative margins may require multiorgan resection. The role of adjuvant radiotherapy and chemotherapy remains a matter of debate [2, 4, 5].

CASE REPORT

We present the case of a 52-year-old male patient, a significant age as it is within the typical range for the development of certain conditions, who was referred to the Medical Emergency Unit due to malaise and fever with suspected ascites. A clinical examination revealed a distended and markedly enlarged abdomen and bilateral lower-extremity oedema. Laboratory investigations revealed elevated levels of C-reactive protein (CRP) and procalcitonin (PCT), suggesting a bacterial infection. Kidney injury

markers were also elevated. Liver enzymes, bilirubin, and prothrombin time were within normal limits; however, hypoalbuminemia was noted. Blood cultures were obtained, and empirical antibiotic therapy with amoxicillin–clavulanic acid was initiated.

We performed a contrast-enhanced CT scan of the abdomen. Imaging revealed a massive, heterogeneous lipomatous mass measuring approximately $53 \times 48 \times 30$ cm, occupying the entire abdominal cavity. The sheer size of the mass which displaced all abdominal organs posteriorly and cranially towards the diaphragm, underscores the urgency of the situation. The mass consisted of lipomatous tissue of variable density, with areas of marginal calcification and solid components. Notably, there were no signs of ascites, and the imaging characteristics were consistent with a giant liposarcoma.

The patient was subsequently referred to the Institute of Oncology in Ljubljana, where he was hospitalised for further diagnostics and treatment. Blood

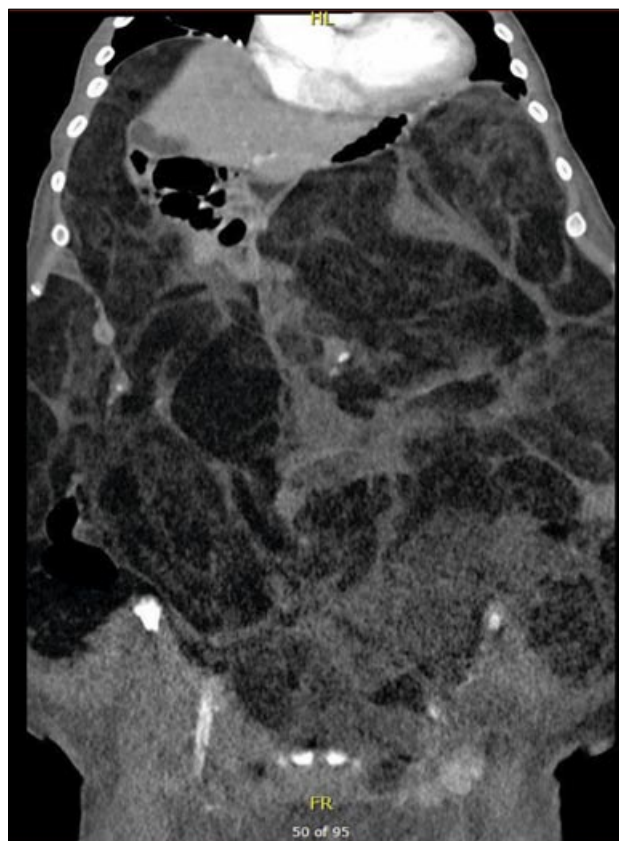


Figure 1: CT scan of giant liposarcoma – axial plane

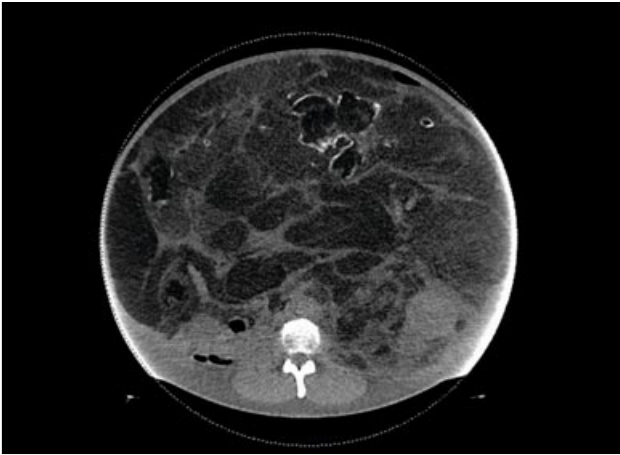


Figure 1: CT scan of giant liposarcoma – coronal plane)

cultures confirmed growth of *Streptococcus pyogenes*. Based on the antibiogram, vancomycin was added to the treatment regimen. Following antibiotic therapy, inflammatory markers declined, and kidney function returned to normal.

Further diagnostic evaluation included a CT scan of the chest, which revealed no signs of metastasis. However, it did show two solitary pathological axillary lymph nodes, each measuring 13 mm in diameter.

An ultrasound-guided biopsy of the abdominal mass was performed, and histopathological examination revealed a well-differentiated liposarcoma, with amplification of the MDM2 gene.

Additionally, ultrasound-guided fine-needle aspiration of a pathological lymph node was performed. Histopathology suggested partial infiltration by small B-cell lymphoma, which was CD20 negative.

The case was presented at the mesenchymal tumour board. After re-evaluation of the CT scans, the board concluded that radical tumour resection was technically not feasible, but a debulking operation could be considered. Furthermore, the patient was deemed at high operative risk due to the size of the tumour, severe malnutrition, and the risks associated with general anaesthesia.

Subsequently, the patient was referred to Germany for a second opinion regarding the surgical strategy.

DISCUSSION

Epidemiology and Reported Cases

Retroperitoneal sarcomas are rare, accounting for approximately 10–15% of all soft-tissue sarcomas. Within this group, liposarcomas are the most frequent histological subtype, accounting for roughly 40–50% of retroperitoneal sarcomas [1, 2]. The overall incidence of retroperitoneal liposarcoma is estimated at less than one case per 100,000 people per year, most often diagnosed between the ages of 40 and 60, with a slight male predominance [1, 2].

Although retroperitoneal liposarcomas are already rare, the subset of “giant” tumours—usually defined as measuring more than 30 cm in diameter or weighing more than 20 kg—is extraordinary. Sun et al. reviewed the English literature and identified only 34 such cases, with well-differentiated tumours being the most common [1].

Histopathology and Classification

The histological subtype is the key prognostic factor. Well-differentiated liposarcoma (WDLPS) tend to recur locally but rarely metastasise, while dedifferentiated liposarcomas (DDLPS) are associated with higher recurrence and metastatic potential. Myxoid subtypes may be more responsive to systemic therapy, whereas pleomorphic subtypes are rare and carry the worst outcomes [1–3]. Immunohistochemistry and molecular testing, such as MDM2 amplification, are important diagnostic tools [1].

Treatment

Complete surgical excision with negative margins is a curative treatment. In giant cases, this often requires en bloc resection of adjacent organs such as the kidney or colon [1, 4]. Preoperative imaging with CT or MRI, sometimes combined with 3D reconstruction, facilitates surgical planning [1].

When radical resection is not feasible due to patient condition or invasion of critical structures, debulking surgery may be considered for symptom relief [1, 2, 5]. The role of adjuvant therapy is controversial: radiotherapy may reduce local recurrence in selected cases, while chemotherapy is mainly reserved for unresectable or metastatic disease [2, 3, 5].

Prognosis

Size alone is not the main prognostic factor. Histological subtype, tumour grade, and completeness of resection are more important predictors [2]. In significant cases, short-term outcomes are favourable when resection is complete, with reports of no recurrence at 6–12 months [1, 4]. However, recurrence remains common in the long term, especially for DDLPS and pleomorphic variants [2].

CONCLUSION

Giant retroperitoneal liposarcoma is an infrequent tumour entity. Despite the tumour's size, complete surgical resection remains the cornerstone of therapy and offers the best chance of long-term control. Histological subtype and surgical margin status are the key prognostic factors, whereas tumour size mainly reflects technical difficulty. Multidisciplinary planning and specialised surgical expertise are essential.

In cases where radical resection is not possible due to tumour extension or patient condition, debulking surgery may provide temporary relief but with limited oncologic benefit. Given the high risk of recurrence, close and long-term follow-up is mandatory. Each case must be evaluated individually, balancing operative risks with potential benefits.

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