Case Report

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Surgical management of giant recurrent retroperitoneal liposarcoma: a case report and review of the literature

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Introduction: Liposarcoma is a rare, primary, malignant mesenchymal tumor. It represents ~7% of all mesenchymal sarcomas and 1% of all cancers. Their incidence does not exceed to 2.5 cases/million inhabitants/year. This tumor is locally invasive, diagnosed at a late stage, and can reach a significant size and weight, resulting in a locally advanced tumor.

Case presentation: A 59-year-old female patient consulting for a large abdominal mass. The abdominal computed tomography showed three retroperitoneal masses, and the surgical exploration revealed a huge process in the retroperitoneal cavity, which takes the left renal compartment, and the left colon. The intervention consisted of a mono-bloc excision of the mass taking the spleen, the left renal compartment, and the left colon with colonic anastomosis. The histological examination concluded the existence of a well-differentiated myxoid liposarcoma of grade I, the postoperative follow-up was simple. One year later, she underwent an excision of a recurrence of the same retroperitoneal location, but of a pleomorphic cell histological type of grade II according to the FNCLCC classification. We review the literature, the pathological, therapeutic, and prognostic aspects of this tumor.

Discussion: Retroperitoneal liposarcoma is a rare tumor. Its gravity is due to an often-late diagnosis, a complete imaging workup including ultrasound, computed tomography and often MRI is necessary preoperatively to determine the relationship with the different organs. The definitive diagnosis is histological, surgery is the most effective treatment and can be extended to neighboring organs. the frequency of recurrence requires particular surveillance.

Conclusions: We highlight the importance of radical surgical excision to avoid retroperitoneal liposarcoma tumor complications and to minimize the recurrence risk.

Keywords: case report, giant masses, liposarcoma, myxoid cells, pleomorphic cells

Introduction

LPS is a rare, primitive, malignant mesenchymal tumor; it most often develops from soft tissue, it represents ~7% of all mesenchymal sarcomas, 1% of cancers[1] and their incidence does not exceed 2.5 cases/million inhabitants/year[2].

It is locally invasive, late diagnosed, and can reach a significant size and weight, resulting a locally advanced tumor. The diagnosis of retroperitoneal LPS is often delayed due to the compliance of the abdominal cavity. In addition, there is a higher rate of recurrence compared with LPSs in other locations[1].

LPS risk factors identified by the American Cancer Society include radiation (especially radiation therapy used to treat other malignancies), certain family cancer syndromes, damage/trauma to the lymphatic system, and exposure to toxic chemicals[3].

We report the case of a 59-year-old woman who underwent complete excision of a giant retroperitoneal LPS. Three months later, she had a recurrence of the same retroperitoneal location, but of a different histological type and grade. We review the literature on the pathological, therapeutic, and prognostic aspects

HIGHLIGHTS

- Liposarcoma (LPS) is a rare, primitive, malignant mesenchymal tumor. It is locally invasive, of late diagnosis.
- Computed tomography (CT) scan is the best examination for the diagnosis of these tumors and specifies the relationship with the surrounding organs.
- The definitive diagnosis is anatomopathological of the tumor resection specimen.
- Surgical resection with a negative margin is considered a primary treatment for LPS.
- The prognostic factors are: the histological grade of the tumor, local recurrence, and distant metastases.

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of this tumor. This work has been reported in line with the SCARE criteria[4].

Case presentation

A 59-year-old female patient with no previous pathological history presented with vague abdominal pain for 9 months, progressive increase in abdominal volume, and weight loss.

The examination revealed edema of the lower limbs, two large abdominal masses, one located in the iliac fossa and the right flank, the other in the left flank, as well as a shifting dullness.

The biology revealed an inflammatory syndrome with C-reactive protein at 56 mg/l, hypoproteinemia at 57 g/l, and hypoalbuminemia at 29 g/l. Abdominal ultrasound showed the presence of a large tissue and cystic mass of the abdominopelvic region and significant infiltration of the mesenteric fat.

The abdominal CT helical (64 strips), 1–2 mm slice thickness, 2–3 mm axial slices and multiplanar reconstructions, thick volumetric three-dimensional sections for vascular assessment showed three abdominopelvic masses in three locations.

One on the right side, hyperdense and poorly circumscribed, measuring 16 cm, one on the left side, cystic, measuring 11 cm, and the third one , has a tissue aspect with cystic areas measuring 12cm in the left iliac fossa. These aspects were suggestive of a LPS. The thoracic CT scan was normal (Figs. 1 and 2).

The patient underwent a midline incision that extended from the subxiphoid to the suprapubic area. The intraoperative exploration revealed:

A huge bilobed retroperitoneal process compressed the peritoneal cavity, which took the left renal compartment, and the left colon which was pushed forward between the two lobes of the tumor.

After reaching the retroperitoneal space and identification of the limits of the tumor, the mass was immediately exposed and extensively dissected using coagulating shears (HARMONIC ACE; Ethicon). A mono-bloc excision of the retroperitoneal mass taking the spleen, the left renal, and the left colon with laterolateral colonic anastomosis, we did not place any drains.

The operation lasted 3 h and a half, the intraoperative bleeding was evaluated at less than 200 ml and the postoperative follow-up was simple.

The macroscopic examination showed a well-limited, polylobed tumor mass, encompassing the left kidney, weighing 3000 g (Fig. 3). On section, the tumor was of variable consistency with a multilobed fatty appearance associated with nodules of fasciculate appearance, myxoid in places (Fig. 4).

The second tumor was a well-limited, polylobed tumor weighing 6000 g and included the spleen and the colon free of tumor invasion. In the section, the tumor was made of round or oval cells.

The histological examination concluded that a well-differentiated grade I LPS according to the FNCLCC classification with resection limits pass in healthy areas.

The postoperative follow-up was simple, the patient had a smooth postoperative course and was discharged on the sixth postoperative day.

There was no indication for postoperative radiotherapy or chemotherapy, which is why the patient was put under a surveillance protocol with a control CT scan after 6 months.

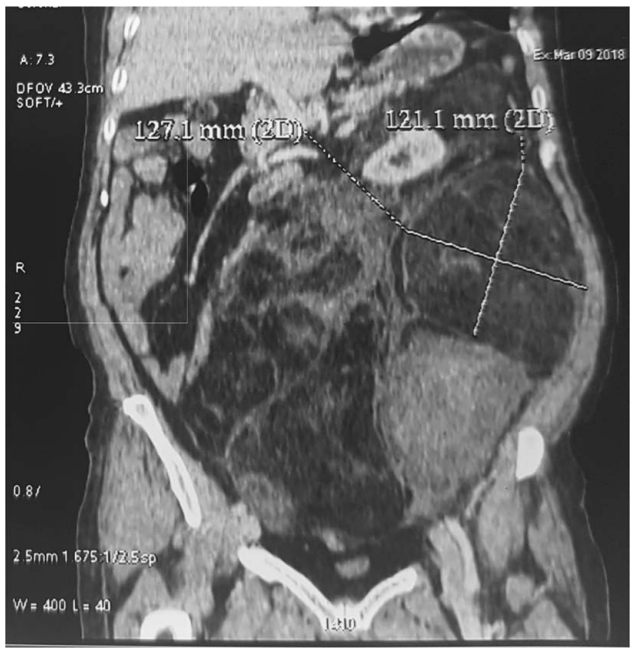

Figure 1. Computed tomography images showing the tumors their boundaries and extensions.

One year later, during the surveillance, a CT scan showed a mass on the left flank in retroperitoneal contact with the psoas muscle measuring 67×56 mm and a pelvic mass lateralized to the left measuring 67×45 mm.

The surgical exploration of the second procedure revealed a pelvic process invading the left ureter, the left adnexa, a loop of bowel, and coming into contact with the external iliac artery and vein.

The surgical procedure consisted of a resection of the pelvic mass enlarged to the left adnexa, and a gallbladder resection involving the psoas muscle (Fig. 5). The histological examination

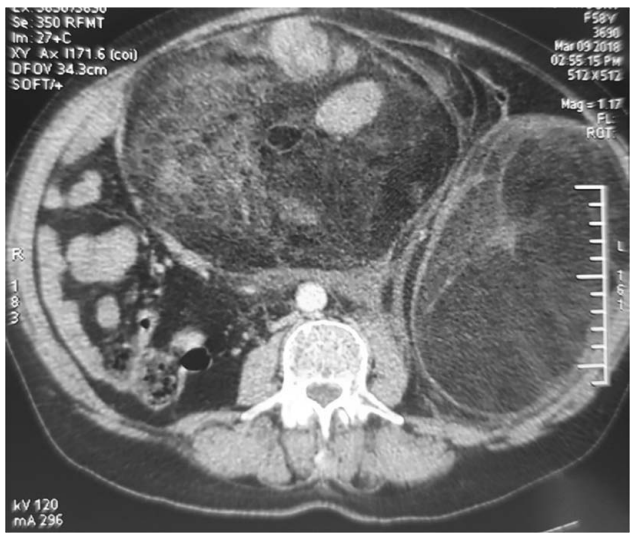

Figure 2. Computed tomography images showing the tumors their boundaries and extensions.

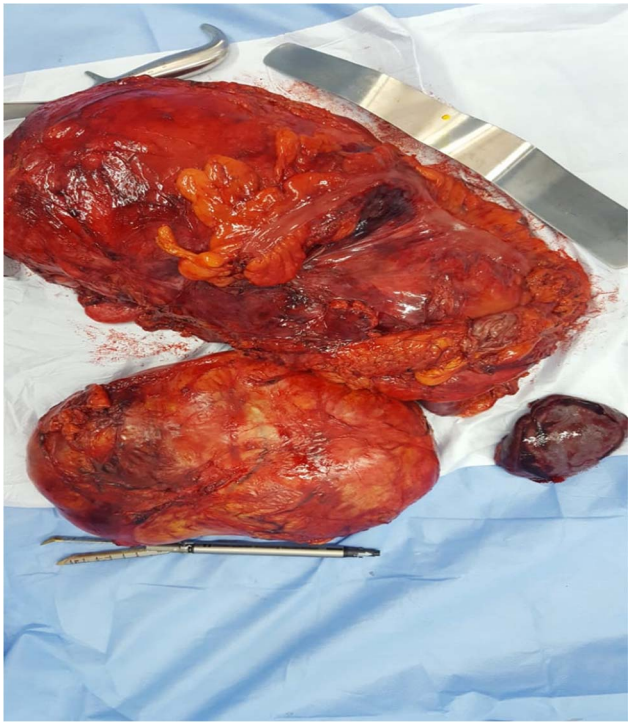

Figure 3. Image of the surgical specimen after tumor resection in one piece.

revealed a pleomorphic cell sarcoma of grade II according to the FNCLCC classification, the small intestine invaded at the level of the subserosa, with healthy proximal and distal limits (Fig. 6).

The patient's postoperative course was uneventful. Abdominal drainage was stopped on the fifth postoperative day, and the patient was discharged on the ninth postoperative day, once deemed physically fit to live independently by physiotherapy and occupational therapy and the patient was then referred to the oncology center for adjuvant radiochemotherapy.

Discussion

LPS represents ~7% of all mesenchymal sarcomas, 1% of cancers.

The average age of the occurrence is 50 years with extremes of 3 and 76 years. This tumor can reach a considerable size of 76 cm

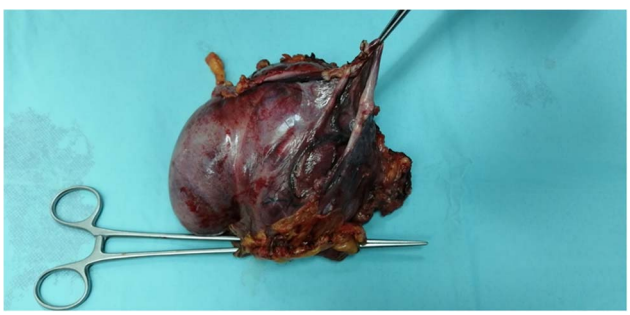

Figure 4. Paucicellular proliferation, made of spindle-shaped cells arranged on a myxoid background (hematoxylin and eosin; × 100).

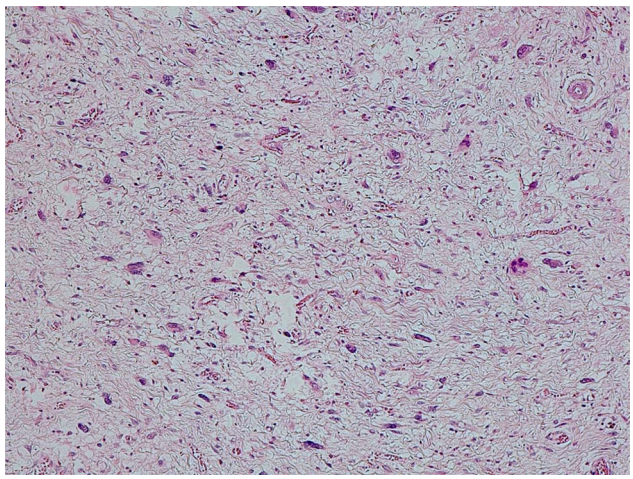

Figure 5. Image of the recurrence piece after resection.

and a significant weight of 30 kg[1] and their incidence does not exceed 2.5 cases/million inhabitants/year[2].

The characteristic of the genetic alteration for myxoid/round-cell LPS is classically a t(12;16) (q13;p11) or t(12;22)(q13;q12) translocation found in more than 95% of myxoid/round-cell LPS, whereas well-differentiated/dedifferentiated LPS is related to the amplification of the 12q13–15 region that comprises the MDM2 and CDK4 genes[5].

The diagnosis of LPS is often delayed due to the compliance of the abdominal cavity.

The clinical symptoms are dominated by abdominal pain with a feeling of heaviness associated with an abdominal mass in 80% of cases[1].

The CT scan is the best examination for the diagnosis of these tumors and specifies the relationship with the surrounding organs. Its appearance is typically a heterogeneous mass with areas of fatty density and others of muscular density. Currently, MRI is increasingly replacing the CT scan in the radiological assessment of LPS. It specifies the characteristics of the tumor and

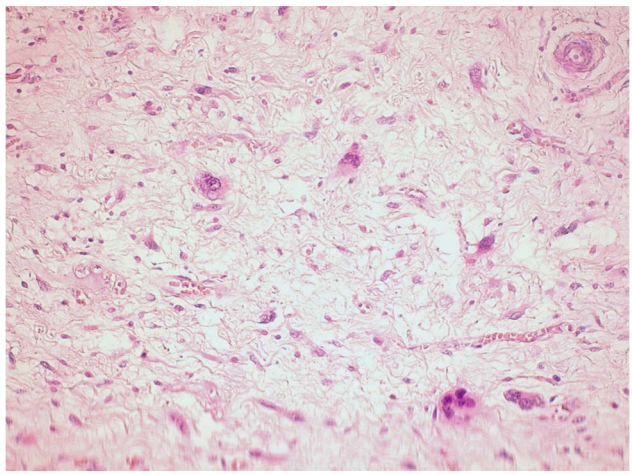

Figure 6. Atypical appearance of tumor cells. It presents a nuclear pleomorphism, frequent multinucleations with numerous mitoses (hematoxylin and eosin; × 400).

assesses the existence of a vascular thrombus and its extent. LPSs have a different signal on MRI depending on the degree of differentiation: for well-differentiated forms, they have a signal intensity close to that of soft tissues, whereas the other types appear in hypo signal in T1 and hyper signal in T2[6].

The definitive diagnosis is anatomopathological of the tumor resection specimen which allows the classification of the tumor according to its histological variety. Several histological varieties have been described: well-differentiated LPS, round-cell LPS, myxoid LPS, pleomorphic LPS, and dedifferentiated LPS[7].

Well-differentiated LPS has a better prognosis than all others LPSs. It can recur locally after excision, but it has a low metastatic potential[8]. The myxoid form is the most malignant and the most frequent; 50%, recurs rapidly and has a worse prognosis. A combination of two or three histological types is possible but rare: mixed-type LPS[9].

The management of myxoid LPS needs to be discussed in multidisciplinary meetings including surgeons, oncologists, and radiotherapists.

Surgical resection with a negative margin is considered a primary treatment for primary retroperitoneal LPS that improves local control[10].

Studies show that clean microscopic margins are associated with longer postoperative survival time compared with resections with a microscopic tumor-positive margin[11].

Large retroperitoneal LPSs present unique challenges and require a more aggressive surgical approach that may include multiple resections for recurrences[12]. Multiple re-operations for recurrent disease may result in a significant increase in long-term survival, even despite the overall higher rate of local recurrence of primary retroperitoneal LPS compared with other sarcomas[13].

A study by Mäkelä *et al.*[14] showed that the rate of complete resection and subsequently, postoperative survival time, is influenced by the inaccessible, deep location of retroperitoneal LPSs, rather than their size alone. Studies reported that the median survival of patients who underwent complete resection was 103 months, as compared with 18 months in patients undergoing incomplete resection[15]. R0 resection of a large retroperitoneal LPS was associated with an 85.7% 5-year survival compared with 33.3% following R1 resection[16]. Wang *et al.*[17] suggested that extended resection that includes adjacent organs is beneficial to achieve radical treatment. Bradley *et al.*[18] reported that over 50% of successful complete excisions also included adjacent organs. The structures most commonly resected are kidneys, ureters, and large bowel.

Although some reports are in support of en-bloc resection of uninvolved adjacent organs to improve local control[19,20], these studies fail to show any improvement in overall survival for extended resection beyond R0. Previous studies[21,22] have demonstrated that organ resections can reduce the local recurrence rates but do not prolong the survival time[21,23].

Complementary treatments are based on adjuvant radiotherapy and/or chemotherapy depending on the stage, histological type, and recurrence of the tumor. Neoadjuvant radiotherapy is indicated preoperatively for unresectable tumors to make them resectable and postoperatively to avoid recurrence[24]. Myxoid forms are more sensitive to radiotherapy than other histological types[24]. Chemotherapy is of little interest since LPSs have low chemosensitivity. The used molecules are doxorubicin and alkylating agents. It is used for forms with a poor prognosis and in adjuvant or neoadjuvant situations in the treatment of metastasized tumors[25].

The prognostic factors are the histological grade of the tumor, local recurrence, and distant metastases. Other factors have been implicated such as: complete resection, tumor volume, age, synchronous metastases, retroperitoneal location, and invasion of neighboring organs[26].

The risk of recurrence is less differentiated and more infiltrative than the first tumor and is accompanied by more distant metastases. They appear within 2 years after the first resection. The frequency of recurrence is 20–85% of cases. Distant metastases can occur in the lungs, pleura, liver, and lymphatics[27].

The median survival at 5 years depends on the grade. It is 85% for grade I, 60% for grade II, decreases to 21% for type III, and 10% for type IV[28].

Conclusions

Retroperitoneal LPS is an invasive tumor. Its severity is due to an often-late diagnosis because of the complacency of the space in which they develop. A complete imaging workup including ultrasound, CT and often MRI is necessary preoperatively to determine the relationship with the different organs. The definitive diagnosis is histological, surgery is the most effective treatment and can be extended to neighboring organs. Surgical treatment may be associated with radiotherapy and chemotherapy in case of an advanced tumor. The high frequency of recurrence requires special surveillance.

Ethical approval

NA.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

M. Ayoub has written the article, consulted the patient, prescribed all of the tests, prepared the patient for surgery, and participated in the surgery. B.L. and J.R. have helped in writing the article and data collection. M. Achraf has helped in the interpretation of histological data. S.M. and P.B.M. (oncology surgery professors) have supervised the writing of the paper and has been the leader surgeon of the case.

Conflicts of interest disclosure

The authors declare that they have no financial conflict of interest with regard to the content of this report.

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