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Giant recurrent retroperitoneal liposarcoma initially presenting as inguinal hernia: Review of literature

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Abstract

INTRODUCTION

Liposarcomas comprise around 15% of soft tissue tumors. These tumors of mesodermal origin arise as single tumors, present one histologic type and diverse locations (including the retroperitoneum). Diagnosis of liposarcomas of retroperitoneum is difficult because of this unspecific presentation and in 50–100% of the cases there is recurrence from residual tissue.

PRESENTATION OF CASE

An 86 year old male patient was admitted in 1996 due to a right and voluminous inguinal hernia. During the herniaplasty, a right paratesticular tumor was isolated and removed. The histologic exam revealed a well-differentiated liposarcoma. A CT scan was performed and a large abdominal mass was detected. The patient underwent a laparotomy and an incomplete resection of the tumor was achieved. After the surgery the patient remained asymptomatic during a long period. Nine years later, the patient underwent another laparotomy with partial removal of the giant recurrent retroperitoneal liposarcoma.

CONCLUSION

The purpose of this publication is to report the recurrence of giant retroperitoneal liposarcoma, which is an unusual presentation in surgery today. Furthermore, we would like to emphasize the long-term survival of this patient despite partial resection and the possibility of performing a re-resection in this type of cases.

Keywords: Liposarcoma, Retroperitoneal, Hernia, Recurrence, Surgery

1. Introduction

Soft tissue sarcomas are rare tumors that represent approximately 1% of all diagnosed malignant neoplasms, 10–15% of which are located in the retroperitoneum.1 Liposarcomas comprise around 15% of soft tissue tumors, thereby constitute the most frequent retroperitoneal tumors.2,3 These tumors of mesodermal origin can reach significant dimensions, despite their poor vascularization.

Liposarcomas frequently arise as single tumors, being rare multicentric presentations. They generally present one histologic type and diverse locations, such as upper and lower extremities, trunk, head and neck, retroperitoneum and mediastinium. The peak of incidence is the age group of 50–70 years.3,4 Due to

their asymptomatic presentation, they are difficult to diagnose. Following surgical removal, 50–100% of the cases recur from residual tissue.

The purpose of this publication is to present a case report of a patient with a rare neoplasm, of unfrequent size and location that recurred nine years following its resection. We also show that this type of liposarcomas progress very slowly and causes no major symptoms.

2. Case report

An 86 year old male with no relevant past medical history was admitted in June 1996 due to a right inguinal hernia which impaired his gait. The patient presented asthenia without other symptoms. During hernioplasty, a 1500 g and 30 cm of diameter right paratesticular tumor was isolated and removed. Histological exam revealed a well-differentiated liposarcoma with areas of the inflammation and sclerosis. Following surgery, a thoracic–abdominal–pelvic computed tomography (CT) scan was performed and detected a large mass extending from the pelvic cavity to the surface of the liver (Fig. 1A).

Afterwards, the patient underwent exploratory laparotomy. The surgery revealed the presence of a voluminous, lipomatous retroperitoneal mass extending from the inferior surface of the liver to the retrovesical recess; the ascending and transverse segments of the colon were displaced anteriorly and as the root of the mesentery anteriorly and to the left. There were no signs of ascites, lymphadenopathies, or liver metastases. At gross examination, the surgical piece revealed many irregular masses of tissue, weighing a total of 7500 g and measuring 40 cm × 30 cm × 12 cm. The histological exam revealed a liposarcoma composed predominantly of well-differentiated, lipoma-like, sclerosing and inflammatory areas; in addition, there was reference to a small portion of the myxoid type. After recovering from the surgery the patient was discharged. The patient was followed in the outpatient section of general surgery and during the first year of follow-up, a recurrence of the liposarcoma was detected (Fig. 1B), but the patient refused surgical treatment and left outpatient appointments, primarily due to his asymptomatic state.

On the April of 2005 (nine years after initial surgery) he was readmitted in our hospital due to a large abdominal mass, as well as dyspnea, asthenia, and anorexia. At physical examination the patient presented a distended abdomen (Fig. 2). A CT scan detected heterogeneous masses compatible with adipose tissue (Fig. 3). One week later, the patient underwent laparotomy with partial removal of the recurring retroperitoneal liposarcoma (Fig. 4). Enterorraphy was performed due to iatrogenic injury of a segment of the small intestine. Gross examination revealed multiple portions, the largest of which with 32 cm of its largest diameter, weighing 15,000 g. The histologic exam revealed a grade 1 mixed liposarcoma, predominantly of the myxoid type, including areas of the lipoma-like and sclerosing type (Fig. 5). There were no complications during the period immediately following the surgery. However, the patient died on post-operative day 7 due to cardio-respiratory arrest.

3. Discussion

Retroperitoneal tumors are an extremely heterogeneous group of neoplasms, 85% of which are malignant. Liposarcomas constitute between 45 and 55% of retroperitoneal masses.5 The clinical manifestations of these tumors are usually unspecific, including asthenia, anorexia and dyspnea. The most frequently referred symptom is diffuse abdominal pain. At physical examination, the patient reveals in 70–80% of the cases a painless, palpable abdominal mass, which may compress and/or dislocate adjacent organs.5 In about 5–6% of the patients, the retroperitoneal tumor is diagnosed accidentally during an exploratory laparotomy or CT scanning for non-related complaints.5 In this present case, the tumor was detected during the right inguinal hernioplasty and further confirmed by CT scanning and exploratory laporatomy.

Basic abdominal imaging studies, as well as fine-needle aspiration (FNA) are auxiliary diagnostic tools of great utility for diagnosis and staging of these tumors.3 A simple abdominal radiograph provides indirect information of the existence of a retroperitoneal mass, but ultrasonography permits a more precise localization of the tumor. In order to determine the stage of the tumor, it is important to search for the presence of pulmonary metastases.3 CT scanning is highly sensitive in determining the size, extension, as well as the presence of the tumor in relation to neighboring organs, in order to permit an adequate preoperative evaluation and in order to exclude hepatic metastases.3 MRI is superior to CT scanning due to its capacity to differentiate tissue types. In this case, MRI was not performed since CT scanning was sufficient to determine the boundaries of the tumor.

Fine-needle aspiration guided by CT or ultrasonography permits a preoperative determination of the histologic type of the tumor. Nevertheless, a definitive diagnosis is only possible after surgical resection, gross and histologic examination. In this clinical case, during the inguinal hernia repair we removed the tumor which allowed the histological characterization/classification of the tumor.

Histopathologic variety is the main prognostic factor. Five histologic types are recognized. The well-differentiated or lipoma-like type represents around 30% of liposarcomas and has the best prognosis. The myxoid type is the most frequent liposarcoma, constituting around 50% of all tumors. It has a less favorable progression, as it often recurs early. The pleomorphic, round cell and undifferentiated types display the worst prognosis.6,7 Five to ten percent of the cases, such as the one presented herein, have a mixed presentation.8,9 In summary, one could conclude that the well-differentiated and myxoid types are low-grade tumors, characterized by a tendency to recur locally. On the other hand, the pleomorphic, round cell, and undifferentiated types are high-grade tumors and tend to spread early.4 Metastases from soft tissue sarcomas are rare, appearing in 1–3% of all primary tumors,5 in this case, we also did not observe any metastases. The myxoid and well-differentiated liposarcomas present a five-year survival of 75%. In contrast, less than 20% of patients with high-grade tumors survive 5 years.9

Surgical removal is considered the gold standard in treatment of retroperitoneal liposarcomas in both primary and recurring tumors.10–12 The resectability of the tumor does not rely upon its size, subtype or histologic grade.10 The difficulty in achieving complete and curative removal of the tumor lies in its relations to neighboring structures.7 In order to obtain a complete removal of the tumor, 50% of the patients must remove some part of adjacent organs, such as the kidney, ureter, and portions of the large intestine.5 In this patient, the tumor infiltrated the retroperitoneum which precluded the complete removal of the mass. The removal of adjacent organs should not be considered a contraindication for surgical intervention, due to the low pre, intra-, and postoperative mortality rates.5 In one study, Lewis attributes a peri-operative mortality of 4%, indicating hemorrhage, sepsis, acute myocardial infarction, and multiorganic failure as the principal causes of death.10 In partial surgical removals, the primary cause of death is the recurrence of the remaining tumor tissue.5 Nevertheless, some authors deny an increase in survival following complete surgical resection of the tumor with adjacent uninvolved organs.2 According to some authors, surgical removal with negative gross margins is of great importance, stating that the survival rate of patients with incomplete removals equals that of patients not submitted to a surgical intervention.10 In accordance, partial resections are only indicated in patients with significant symptoms and unresectable tumors.10 Linehan, however, states that negative gross margins do not exclude the presence of residual tumoral tissue. The presence of local recurrence is observed both patients with negative and positive gross margins, which excludes gross surgical margins as a factor predictive of recurrence.13

The importance of adjuvant therapies is also disputable.7 Contrary to the majority of mesodermal tumors, which are resistant to radiotherapy (RT), liposarcomas are sensitive to this form of treatment. However, the primary limitation of RT lies in its toxic effects on the adjacent intra-abdominal and retroperitoneal organs.7,10 As a result, there is no consensus on the efficacy of RT as an adjuvant treatment. Some authors

argue the use of RT to raise the survival rate, while others deny any importance of RT as an adjuvant treatment to complete surgical resection.7,11 In respect to RT as a palliative treatment, however, all authors recommend its application in cases where the tumor is inoperable or not completely resectable. The efficacy of chemotherapy has not been demonstrated in any hitherto published study. In fact, significant side effects and high morbidity rates have been detected. Therefore, one could conclude that chemotherapy presents very limited importance regarding the treatment of liposarcomas.7,10,11

The prognosis of liposarcoma depends on the degree of differentiation, size, histological type and tumor staging. The gold standard treatment is total surgical resection with free margins, which might be predictive of cure. However, if total resection is not possible, RT should be considered in order to decrease the recurrence of the disease. Regarding the presented case, the patient refused any treatment, so this option was discarded. With this case report, we also show that re-resection of liposarcoma is a viable option.

The case report here presented strengthens the idea that these patients need tight surveillance, which should be stimulated by the clinician, in order to achieve a better outcome that the one presented here.

Conflict of interest statement

The authors declare no conflict of interest.

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None.

Ethical approval

Written informed consent was obtained from the patient for 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.

Author contributions

PL, SV, MO and JF have helped in the acquisition of data. PL wrote the manuscript.

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Figures and Tables

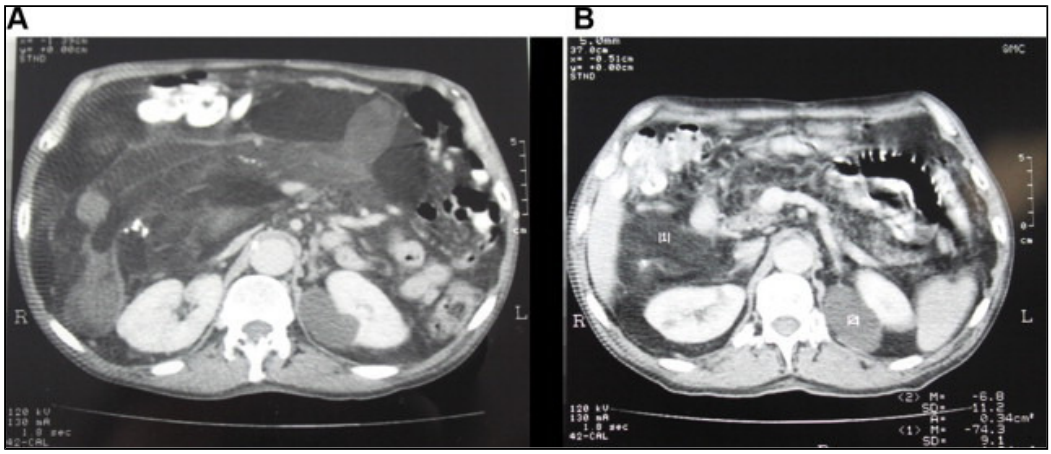

Fig. 1

Abdominal CT scanning after hernioplasty (for liposarcoma staging) (A), and control CT scan after first removal of the liposarcoma and showing the recurrence of the same (B).

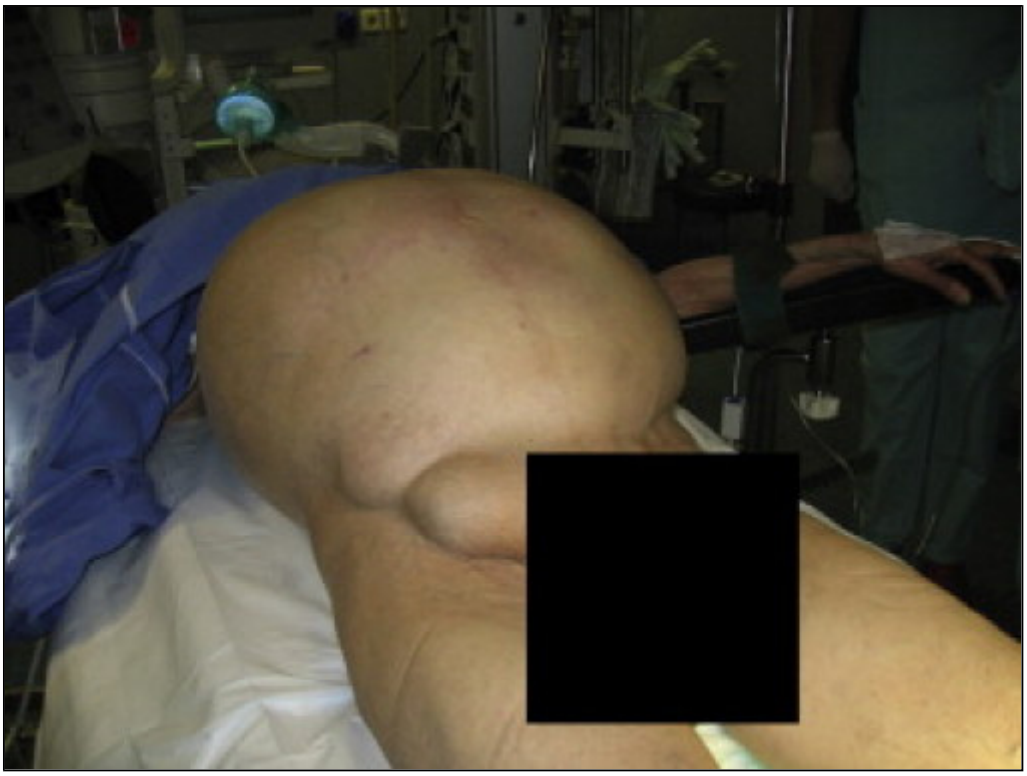

Fig. 2

Patient with large abdomen moments before intervention.

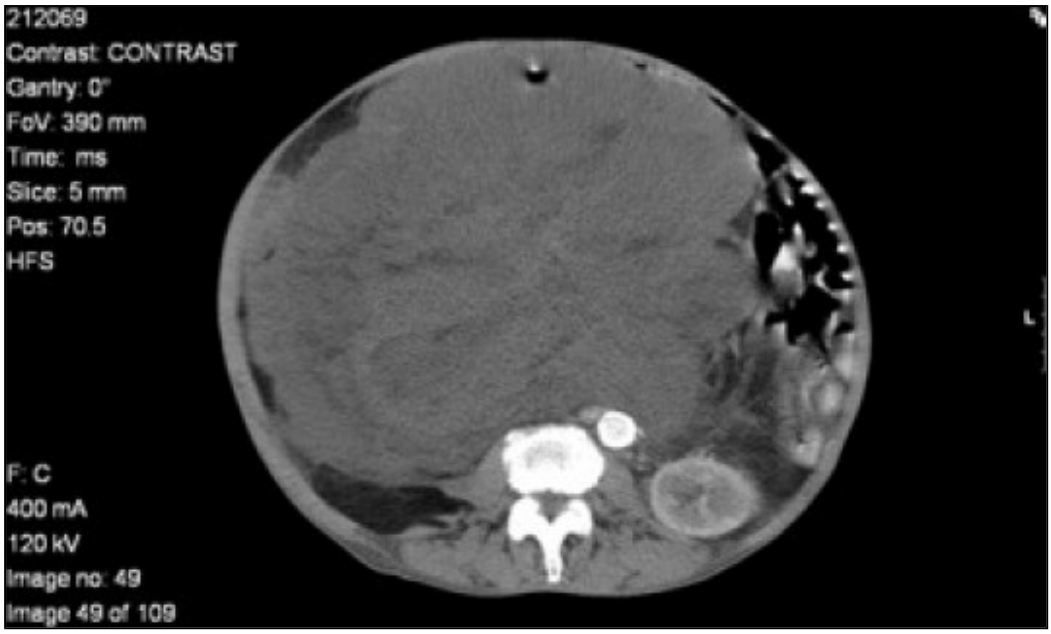

Fig. 3

Abdominal CT scanning revealing a unique mass consistent with liposarcoma recurrence 9 years later.

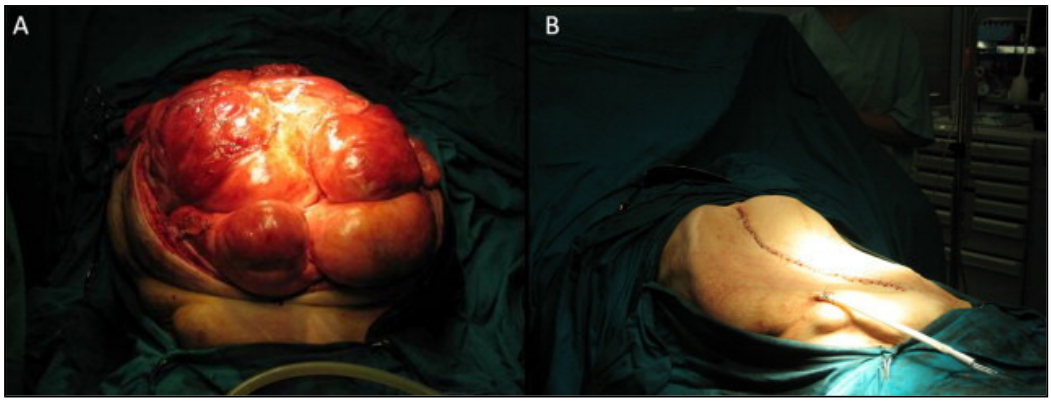

Fig. 4

Liposarcoma *in situ* (A) and patient after removal of the tumor (B).

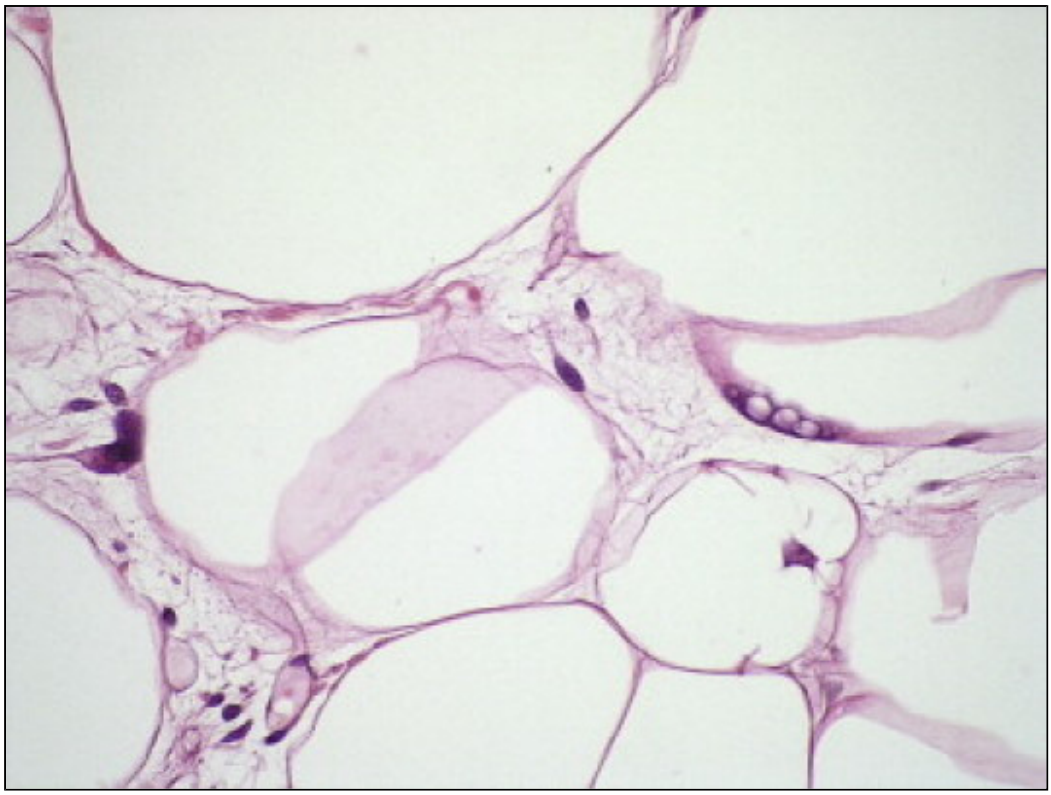

Fig. 5

Grade I mixed liposarcoma, predominantly of the myxoid type, including areas of the lipoma-like and sclerosing type.