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Is compartmental surgery always mandatory in retroperitoneal liposarcoma?
A case report of multifocal synchronous well-differentiated liposarcoma.

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Is compartmental surgery always mandatory in retroperitoneal liposarcoma? A case report of multifocal synchronous well-differentiated liposarcoma.

AIM: *Sarcomas are rare tumors representing 0.7% of all cancer cases in adults, and approximately 15-20% of those occur in the retroperitoneum. Diagnosis is usually late. Liposarcoma and leiomyosarcoma are the most frequent forms. Liposarcomas have high local recurrence rates (35-60%) and a high metastasis rate only if dedifferentiated (30%), whereas leiomyosarcoma has a high distant metastasis rate (60%) and a low local recurrence rate (20%).*
CASE REPORT: *A case report of multifocal synchronous well-differentiated liposarcoma is presented. The patient underwent a surgical excision of all the masses. The postoperative period was uneventful, with a postoperative hospital stay of 9 days. The patient underwent systemic chemotherapy and clinical and instrumental follow-up. A relapse of the disease was observed 24 months after surgery: a 25 mm mass was diagnosed close to the pancreatic stump, as well as a 24 mm mass in the left upper abdominal quadrant. The patient underwent a second laparotomy: recurrent lesions were identified and excised en-bloc with the body* of the pancreas.
CONCLUSION: *Surgery is the gold standard of therapy. The best chance for curative resection is at the time of the first diagnosis of the disease. Compartmental surgery is a macroscopically complete resection through en bloc excision of adjacent structures, even if not clearly infiltrated. Many controversies still exist in the treatment of retroperitoneal liposarcoma, such as the extent of primary and secondary resections, the benefit of chemotherapy and radiation therapy, and when these treatments should be delivered.*

KEY WORDS: Compartmental surgery, Liposarcoma, Surgery, Retroperitoneal sarcoma,

Introduction

Retroperitoneal sarcomas are rare tumors [1-4]. Liposarcoma is the most frequent histotype. It shows a high rate of local recurrence with, on the contrary, a lesser and later tendency to metastasize [5]. It is important to carefully evaluate the preoperative radiological images, to precisely define the exact extent of the tumor. Some areas are particularly difficult to assess and may require specialized radiological evaluation. Furthermore, the specific identification of well-differentiated and dedifferentiated components in liposarcomas is particularly insidious and is essential for a correct treatment plan.
Surgery is the primary treatment [6-10]. The best chance for curative resection is at the time of the first diagnosis of the disease. The tumor must be operated on with tumor-free macroscopically large margins. The retroperitoneal space does not allow for compartmental surgery. The surgery of these sarcomas is not recoverable with a second intervention and there is no evidence that a second intervention is able to recover the prognostic sig-

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nificance negative of a first intervention carried out in an inadequate manner. Incomplete resection of a primary retroperitoneal liposarcoma is associated with a very poor prognosis and can only be considered for palliation in highly selected and symptomatic patients [8,11,13]. In the clinical history of this disease, repeated interventions are possible, especially for liposarcomas, due to frequent recurrences, but the interventions following the first never manage to achieve the same level of disease control obtainable at the first intervention. The optimization of the first approach in fact drastically reduces the risk of recurrence [4]. In case of loco-regional recurrence, a multi visceral surgery with the same intentions as surgery on the primitive can be considered if the first intervention consisted of a simple excision. Otherwise, the aim of recurrence surgery should be aimed at obtaining a macroscopically complete resection that includes the surrounding organs only when clearly infiltrated.

Clinical Case

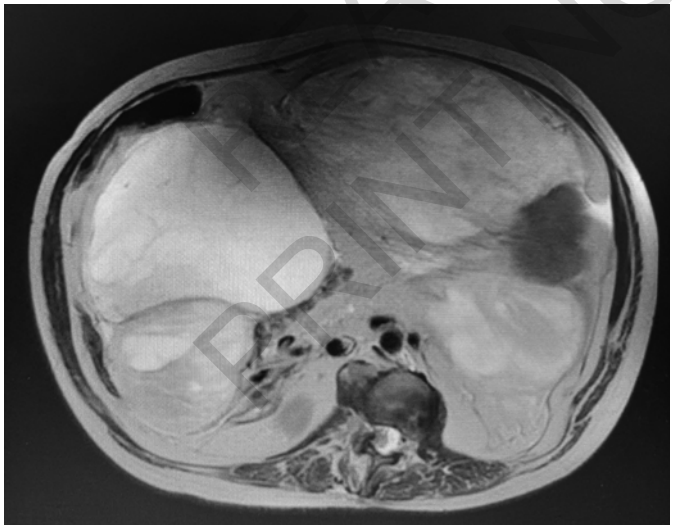
A 72-year-old Caucasian female complaining of severe abdominal discomfort and nonspecific pain, constipation, early satiety, and weight loss presented at our clinic at the General Surgery Operative Unit, San Giovanni Hospital, Crotone (IT) in August 2017. No concomitant diseases were documented in medical history, except for a total hysterectomy due to uterine fibromatosis. Contrast-enhanced CT-scan did not show distant metastasis, while contrast-enhanced MRI documented four large abdominal and retroperitoneal masses with markedly non-homogeneous signal and adipose areas in their context, compatible with liposarcoma (Fig. 1).

After multidisciplinary evaluation, the patient underwent a surgical excision of all the masses. The most superficial intraperitoneal mass was removed en-bloc with the omentum (Fig. 2A, Fig. 2B).

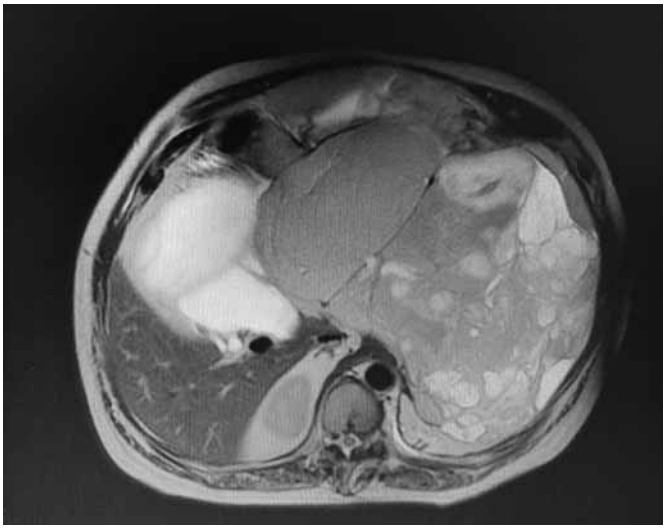
The left retroperitoneal mass was removed en-bloc with the fascia of Gerota and perirenal fat (Fig. 2C). The subhepatic lesion was removed en-bloc with the fascia of Gerota, perirenal fat and the fat around the inferior vena cava (Fig. 3A). The upper left quadrant abdominal mass was removed en-block with the tail of the pancreas and the spleen (Fig. 3B), resulting in R0 resection. Histopathological examination documented a well-differentiated liposarcoma, with microscopically disease-free margins in all surgical specimens. The postoperative period was uneventful, with a postoperative hospital stay of 9 days.

The patient underwent systemic chemotherapy and clinical and instrumental follow-up. At 14 months follow-up, an incisional hernia was identified. A relapse of the disease was observed 24 months after surgery: a 25 mm mass was diagnosed close to the pancreatic stump, as well as a 24 mm mass in the left upper abdominal quadrant.

The patient underwent a second laparotomy: recurrent lesions were identified and excised en-bloc with the body of the pancreas. Histopathological examination documented a well-differentiated liposarcoma, with tumor-free resection margins. During chemotherapy treatment, six-month after the second surgery, the patient died due to pneumonia and fever during SARS-CoV-2 pandemia.

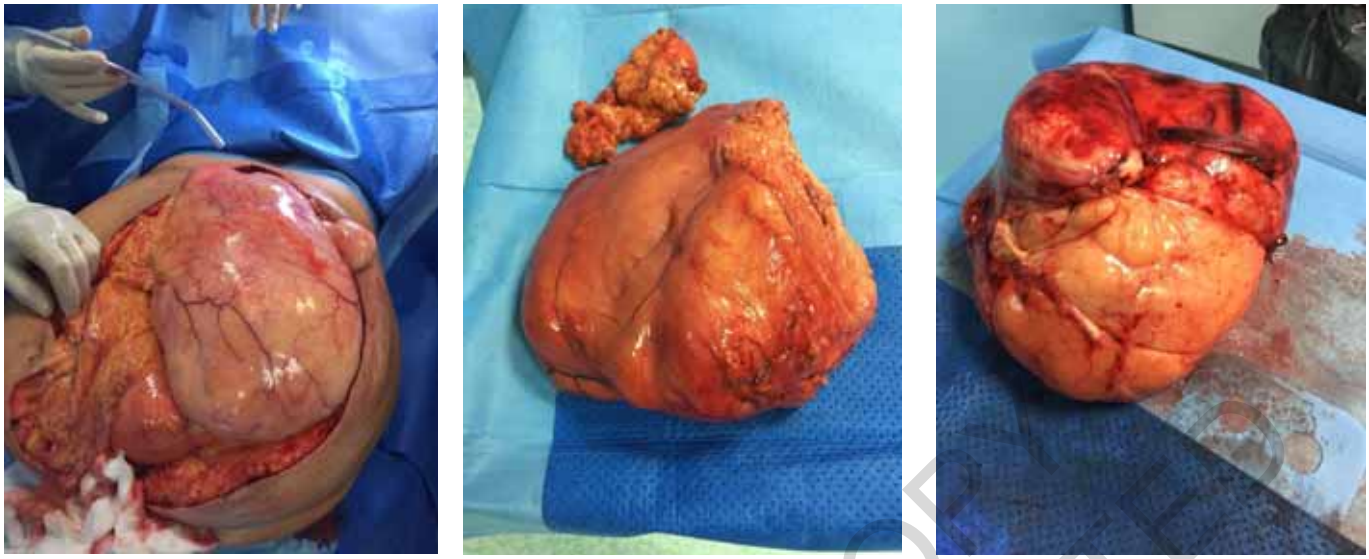


A)



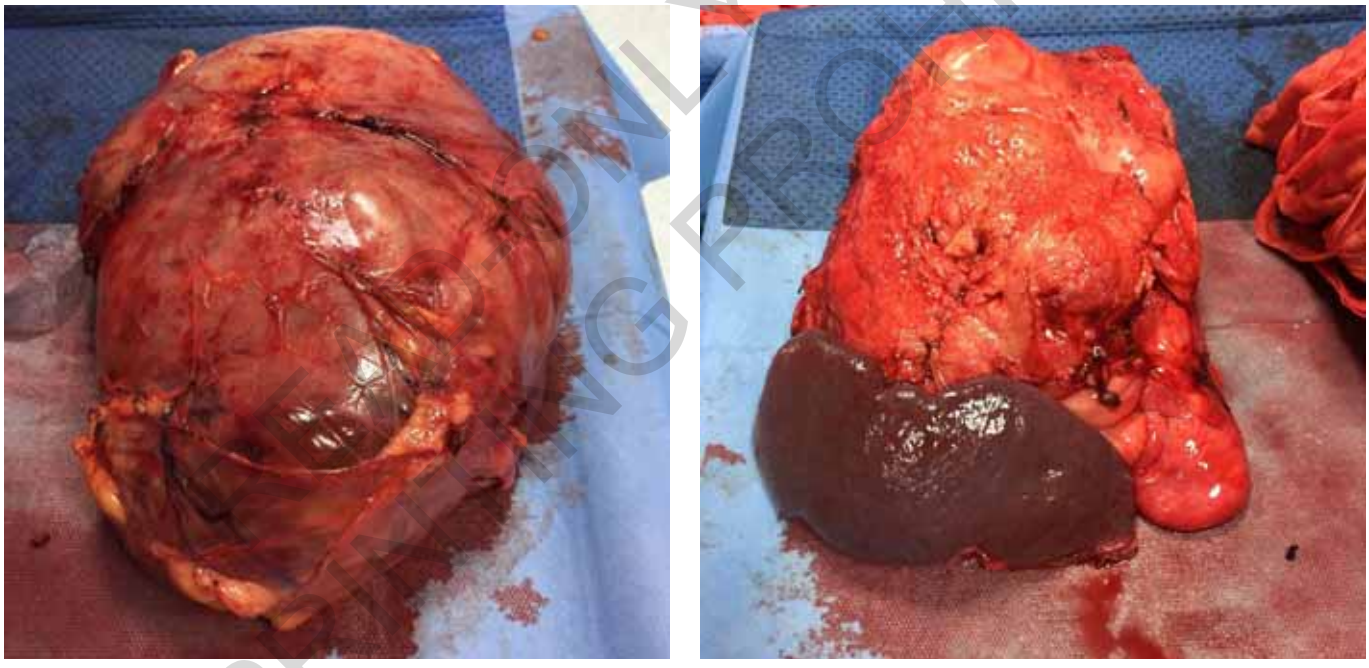
B)

Fig. 1: Contrast-enhanced MRI. Mass in the omental fat with a transverse diameter of 21 cm and craniocaudal extension comparable to that of the entire omentum, with markedly non-homogeneous signal and adipose areas in its context. (A) Expansive lesion of 18 cm in the subhepatic area, with markedly non-homogeneous signal, fluid content with internal septa. (B) Mass in the left retroperitoneal space that displaces the left kidney and spleen anteriorly. (C) Voluminous expansive lesion of 13 x 7.5 cm, caudal to the great gastric curve, indissociable from the tail of the pancreas.



A) B) B)

Fig. 2: (A) Intraoperative photo in which the most superficial of the masses is apparent. (B) The mass excised with omentum. (C) Operative specimen of the mass in the left retroperitoneal space.



A) B)

Fig. 3: (A) Operative specimen of the mass in the subhepatic space. (B) Surgical specimen of the mass removed en-bloc with the spleen and tail of the pancreas.

Discussion

Retroperitoneal liposarcomas are rare neoplasms [1]. Surgery should be aimed at achieving a macroscopically complete excisions with negative margins [1,8].
The histologic subtype significantly impacts the patterns of recurrence, with the low grade liposarcoma being at higher risk of recurrence. Since locoregional control is important for liposarcomas, there is some controversy over the appropriate extent of resection when there is no obvious evidence of tumor involvement of adjacent organs/structures at the time of surgery. Some sarcoma specialists would consider this as rationale to perform compartmental resection [6,14]; this procedure is controversial, as, in some liposarcoma patients, recurrence may develop in areas outside of the resection field. While the

orientation towards compartmental surgery in case of a single retroperitoneal liposarcoma appears univocal, there is no "one size fits all" approach to the treatment of multiple/multifocal synchronous sarcomas. However, other sarcoma specialists have criticized studies on compartmental surgery, pointing out their retrospective nature, inherent selection bias, and the potential for higher complication rates.

Multifocal disease carries in itself a poorer prognosis, with a very high rate of recurrence [9,11]. Despite a complete surgical resection, well-differentiated liposarcoma is fraught with a high local recurrence rate. Reasons for this are not completely understood but may be related to a field defect of the retroperitoneal fat that creates a niche for recurrence. Furthermore, the well-differentiated component of liposarcoma is virtually undistinguishable from normal fat. Therefore, the extent of surgery should be aimed to remove all ipsilateral retroperitoneal fat en-bloc with the mass at the price of sacrificing at least the ipsilateral kidney and colon and part of or the entire psoas muscle.

Extended surgery may raise concern for added morbidity. Depending on the organs resected during combined resection, the complication risk differs [1,8]. It is low when the intestinal tract and kidneys are involved, while it is high when the pancreas, spleen and duodenum are involved.

In well-differentiated multiple liposarcomas, as in the present case, a compartmental approach may be less useful. In our patient, adding bilateral nephrectomy and colectomy, in association with the performed distal spleno-pancreatectomy, was not a clinically feasible procedure. While both renal capsules were removed and found to be disease-free, while bilateral nephrectomy would have made hemodialysis necessary. In addition to this, the disease recurrence occurred in the left upper abdominal quadrant, close to the pancreatic body and not in the peri-renal spaces. It is important to consider that the oncologic outcome deteriorates with each surgical resection while the risks of major associated morbidities increase. Risk and benefits need to be judiciously weighed when planning and timing treatment for multiple retroperitoneal and abdominal liposarcomas.

Riassunto

I sarcomi sono tumori rari che rappresentano lo 0,7% di tutti i casi di tumore negli adulti e circa il 15-20% di questi si manifesta nello spazio retroperitoneale. La diagnosi è generalmente tardiva. I liposarcomi hanno un alto tasso di recidiva locale (35-60%) e un alto tasso di metastasi solo se sdifferenziati (30%), mentre il leiomiosarcoma ha un alto tasso di metastasi a distanza (60%) e un basso tasso di recidiva locale (20%).

Viene presentato un caso clinico di liposarcoma sincrono multifocale ben differenziato. La paziente è stata sottoposta a escissione chirurgica di tutte le masse. La paziente è stata sottoposta a chemioterapia sistemica e follow-up clinico e strumentale. 24 mesi dopo l'intervento chirurgico è stata osservata una recidiva della malattia: è stata diagnosticata una massa di 25 mm vicino al moncone pancreatico e una massa di 24 mm nel quadrante addominale superiore sinistro. La paziente è stata sottoposta a una seconda laparotomia: sono state identificate le recidive ed asportate in blocco con il corpo del pancreas.

La chirurgia rappresenta il gold-standard della terapia. La migliore possibilità per la resezione curativa è al momento della prima diagnosi della malattia. La chirurgia compartimentale è una resezione macroscopicamente completa mediante escissione in blocco di strutture adiacenti, anche se non chiaramente infiltrate. Esistono ancora molte controversie nel trattamento del liposarcoma retroperitoneale, come l'estensione delle resezioni primarie e secondarie, il beneficio della chemioterapia e della radioterapia e quando questi trattamenti dovrebbero essere somministrati.

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