

C A S E R E P O R T

Well-differentiated mesenteric liposarcoma: report of two cases

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Abstract. Liposarcoma is a rare tumor that can be treated by surgery in the absence of distant metastases. Management of liposarcoma, including diagnosis and therapy, is challenging because it has no characteristic symptoms and no established effective treatment. Here, we report two rare cases of primary mesenteric liposarcoma. In the first case, the tumor caused small bowel obstruction, and the patient presented with abdominal distention and severe abdominal pain. The second case is an occasional finding that occurred during laparoscopic surgery for incisional hernia. Both patients underwent successful resection of the tumor. Histopathology found a well-differentiated liposarcoma in both cases. (www.actabiomedica.it)

Key words: Well-differentiated liposarcoma, small bowel, mesentery, rare tumors.

Introduction

LiPoSarcoma (LPS) is one of the most common types of soft tissue tumors in adult patients. It has a peak incidence between 40 and 60 years of age and shows a slight male predominance. LPS are usually found in the deep soft tissues of the extremities, retroperitoneum, or cervical area (1). Other sites are uncommon. Primary mesenteric LPS is rare, with only a few cases reported in the English literature. There are four histopathological types of LPS: well-differentiated, pleomorphic, myxoid, and dedifferentiated (2). Well-differentiated LPS is the most common histological subgroup; they are locally aggressive but incapable of metastasis.

There are no standard guidelines for the management of mesenteric LPS, given the rarity of the tumor. Currently, the treatment strategy is surgical resection with a wide surgical margin if no distant metastases

are detected, often followed by radiation and/or adjuvant chemotherapy for high-risk patients only. Here we present two sporadic cases of primary mesenteric LPS that were removed by surgical treatment.

Case Presentation

Case 1

A 43 years-old man, in good general condition, presented to the emergency department with abdominal distention, severe pain since the day before, and progressive constipation. He did not report a significant weight loss, but he accused cramping and abdominal swelling; he denied diarrhea, fever, and vomiting. The patient had been examined in another hospital the day before, where an abdomen X-ray had been performed without significant signs of bowel obstruction.

There was no history suggestive of bowel cancer, melena, or urinary complaints, but he reported chronic colitis. He reported smoking and drinking habits. No other significant medical history was obtained. His family history was unremarkable. The abdomen examination revealed abdominal distention, slight pain, and tenderness without any palpable masses. Laboratory tests on admission were within normal values but showed an increased C-Reactive Protein (CRP). Computed Tomography (CT) scan showed a 12 cm x 12 cm x 9 cm low-density mass located in front of the hepatic flexure and volvulus of the small bowel with a

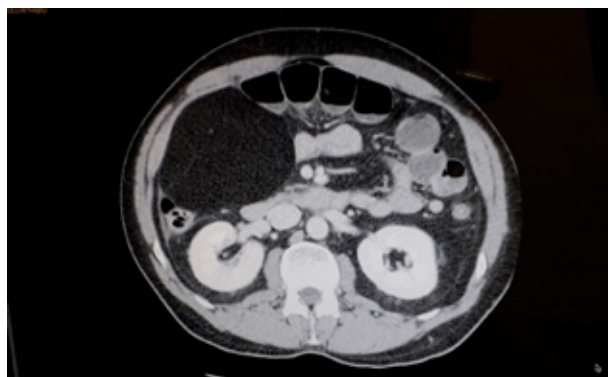


Figure 1. Axial section of the CT abdomen shows a well-defined heterogeneously hypodense mass in the peritoneal cavity predominantly on the right side; there is no calcification or necrotic area.

caliber change without any evidence of distant metastasis or ascites (Fig. 1).

A laparotomy was performed, detecting a large mass, well-encapsulated, arising from the mesentery and causing the volvulus. The tumor was resected en-bloc with almost 8 cm of the small bowel, and a later-lateral isoperistaltic anastomosis was fashioned. No additional tumor or evidence of metastatic disease was found (Fig. 2).

The tumor weighed 704 g with a size of 15 cm; the cut surface had a yellow color, and the mass infiltrated the intestinal wall. The histopathological sections revealed a well-differentiated LPS lipoma-like; it was composed of mature adipocytic cells, rare stromal cells, and monovacuolated lipoblasts (Fig. 3).

The postoperative period was free from complications. The patient was discharged after five days. Adjuvant chemotherapy was not administered. The last CT scan performed did not show any signs of disease. The patient was referred for oncological management; now, he is under follow-up without any oncological therapy.

Case 2

A 60 years-old man underwent laparoscopic surgery for an incisional hernia. The patient reported a previous laparotomy for unspecified surgery performed in Moldova ten years before. No other significant

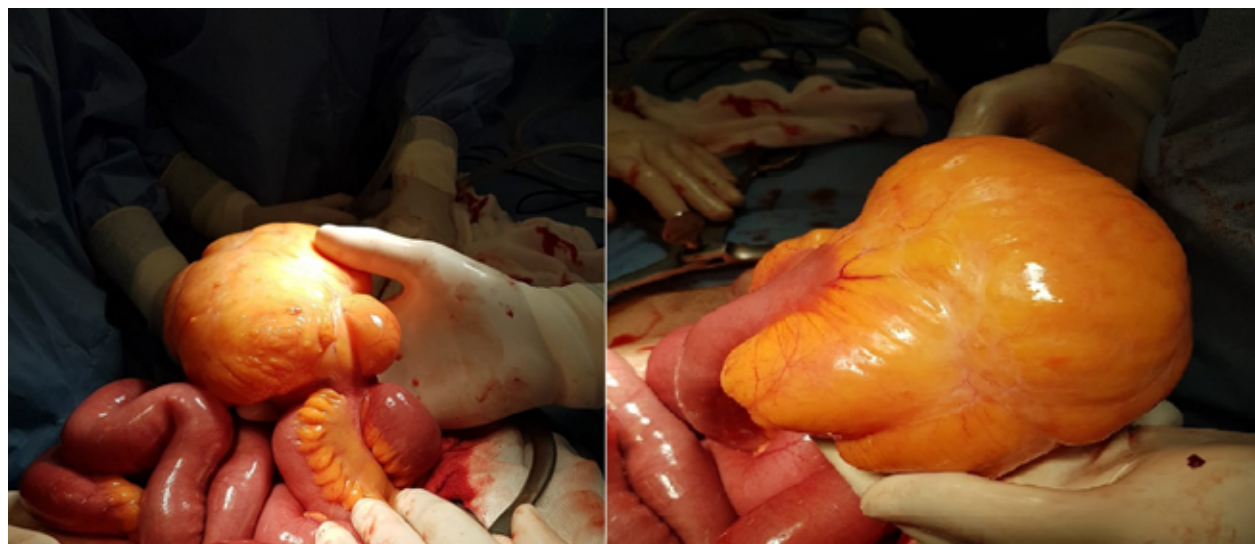


Figure 2. Mesenteric liposarcoma during resection.

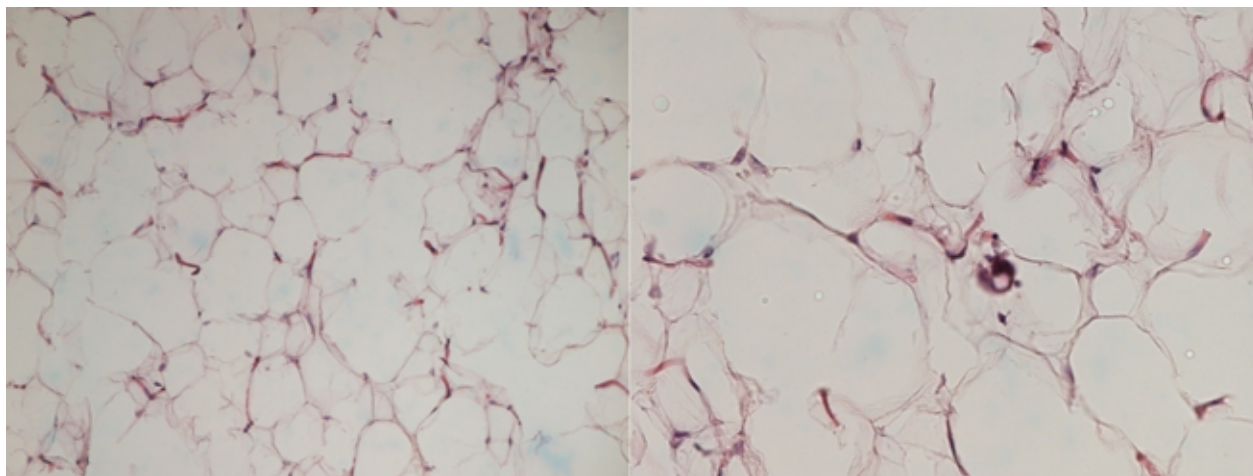


Figure 3. Histology of the well-differentiated mesenteric liposarcoma: it shows mature adipocytic cells and monovacuolated lipoblasts.



Figure 4. Three mesenteric liposarcomas during and after surgical resection.

medical history was obtained. Preoperative laboratory tests were normal except for increased CRP and blood sugar levels. During laparoscopy, three roundish neoplasms of about 8 cm in diameter each, arising from the mesentery, were found: therefore, a laparotomy was performed. The tumors were indissociable from the bowel wall, and they were resected en-bloc with 37 cm of the small bowel. A latero-lateral isoperistaltic anastomosis was performed. No additional tumor or evidence of metastatic disease was found (Fig. 4).

Tumors had a maximum diameter of 9 cm, 10 cm, and 7.5 cm. The histopathological sections revealed a well-differentiated LPS sclerosing variant; in the smallest tumor, the presence of chronic xanthogranulomatous inflammation was found, with prints of crystals of cholesterol as for previous bleeding (Fig. 5).

The postoperative period was unremarkable except for a subcutaneous seroma of the abdominal wall of 11 cm x 2.5 cm. The patient was discharged in good general condition after two weeks. The patient is currently on oncological follow-up without adjuvant chemotherapy.

Discussion

Sarcomas are mesenchymal tumors that arise from skeletal and smooth muscle, adipose and fibrous tissue, bone, and cartilage. They have a mesodermal origin (3). Soft tissue sarcomas account for approximately 1% of all new cancer diagnoses. LPS is the most common sarcoma of the soft tissue in adults, occurring most

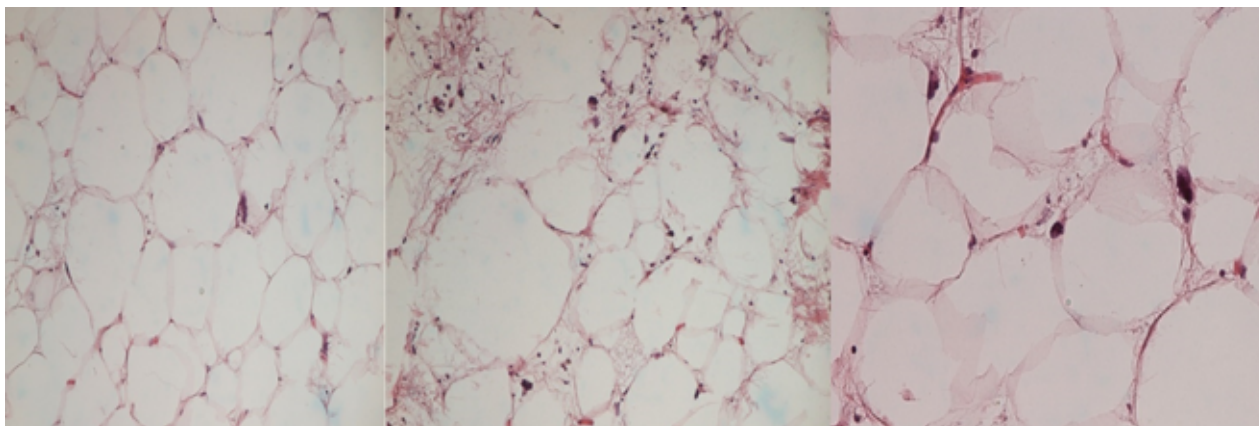


Figure. 5 Broad fibrous septa containing atypical cells and floret-like giant cells; they are embedded in a sclerotic background (sclerosing variant).

commonly in the extremities (accounting for 56% of all the LPS) and retroperitoneum (15% to 20%) (4). Primary LPS arising from the bowel mesentery is an extremely rare lesion. The early symptoms of this disease are nonspecific: some of the most common presenting symptoms include increasing abdominal distention, abdominal pain, weight loss, early satiety, and freely mobile abdominal lumps. These tumors rarely cause perforation, obstruction, intussusception, acute appendicitis, or symptoms mimicking prostatism (5). Usually, ultrasonography, CT scan, and Magnetic Resonance Imaging (MRI) can be used for radiological assessment of the tumor (6). In particular, CT scan and MRI play an essential role in the preoperative diagnosis of LPS; they are of immense value to assess the size of the tumor and the involvement of adjacent organs (7). The reported characteristics of LPS on CT images are inhomogeneity, infiltration or poor margination, CT numbers greater than normal fat, and contrast enhancement. Well-differentiated LPS are hyperintense on T2-weighted MRI with minimal or no enhancement (8). Mesenteric angiography has been used to ascertain the location of the tumor preoperatively (9). A needle biopsy can be fallacious due to inadequate sampling, making it difficult to distinguish from high-grade sarcomas (10,11).

According to World Health Organization (WHO), LPS can be divided into four morphological subtypes: well-differentiated, pleomorphic, myxoid, and dedifferentiated (2). Well-differentiated LPS, which is

the most common variant, was previously classified as liposarcoma not otherwise specified. Pleomorphic is the least common variant, accounting for less than 5% (12); at clinical presentation, these tumors range from small superficial low-grade to large infiltrating poorly differentiated retroperitoneal tumors. Prognosis is unfavorable and worsened by the presence of metastases. Myxoid LPS recur very frequently. In 2013 the WHO removed the standalone “round cell” classification and included it in the category of high quality- worse prognosis- myxoid liposarcoma. Dedifferentiated LPS can be found in up to 10% of well-differentiated LPS of any type and has a more aggressive course and the worst prognosis because approximately 40% show local recurrence, 17% metastasize, and 28% of the patients ultimately die as a result of the tumor (2).

Well-differentiated mesenteric LPS tends to occur in the fourth to seventh decade of life (mean age 57.9 years) with no difference in frequency based on the patients’ sex (men/women: 7/5) (13). They are relatively large at the time of surgery and can be subdivided into the following groups: lipoma-like, sclerosing, inflammatory, and spindle cell (2). More than one histologic variant is common in single tumors, and subclassification does not indicate any prognostic significance. The most common differential diagnostic problem of well-differentiated LPS is its distinction from spindle cell/pleomorphic LPS. Lipoblasts are the hallmark of any liposarcoma subtype (2). Lipoma-like well-differentiated LPS can be identified by the

population of various-sized fat cells, including atypical nuclei and immature fat cells (13).

Only a few cases of mesenteric LPS have been described, and therefore, there are no validated guidelines for the treatment of these tumors. Surgical excision is the recommended primary treatment whenever feasible; wide excision with negative margins (R0) is the standard surgical procedure if no distant metastases are detected.

Before surgery, it may be helpful to evaluate the option to perform the combination of Ifosfamide and external beam radiotherapy, to reduce the mass and perform a complete resection (14). The combination of preoperative radiotherapy and chemotherapy with surgery and intraoperative radiotherapy seems superior to surgery alone or surgery with radiotherapy in local control (14,15).

After surgery, radiotherapy gives no advantage, and it should be applied in selected cases only, while chemotherapy seems to provide limited benefits in a very low number of patients (15).

The prognosis is affected both by the histological subtype and the clinical and pathological characteristics of the tumor. Well-differentiated LPS subjected to radical surgical treatment usually exhibit a low incidence of local recurrences (<10%) and remote metastasis (near to 0%). Complete surgical resection, which often includes adjacent organs, offers the only reasonable chance of long-term survival, with overall average rates between 23% and 59%. These outcomes are worse in patients with myxoid, pleomorphic, and dedifferentiated LPS as well as in case of recurrence (14). Aggressive re-operation is recommended in cases with local recurrence.

In conclusion, primary mesenteric LPS is extremely rare and is treated by aggressive surgical management. Complete resection and long-term follow-up are necessary for well-differentiated mesenteric LPS due to the risk of local recurrence. A multidisciplinary approach with neoadjuvant or adjuvant therapies can be proposed to improve outcomes: the results are still not significant in few cases reported in the literature.

Abbreviations: LPS: LiPoSarcoma; CRP: C-Reactive Protein; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; WHO: World Health Organization

Declarations: • Informed consent: written informed consent was obtained from the patients to publish this case report and any accompanying images; • All authors have reviewed and approved the final manuscript.

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Ethical Approval: This study was not conducted with research intervention; thus, ethics committee approval was not necessary.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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