Huge retroperitoneal liposarcoma: a case report and review of literature

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Introduction
Liposarcoma is the most common malignant tumor of the retroperitoneum. We report a case of huge retroperitoneal liposarcoma weighing 44 kg that was successfully resected.
Case presentation
A 53-year-old Egyptian male patient was presented to our department with progressive abdominal enlargement. Computed tomography scans of the chest, the abdomen, and the pelvis revealed a huge pelviabdominal mass highly suggestive of liposarcoma. Abdominal exploration was performed with resection of a huge retroperitoneal mass weighing 44 kg that was proven pathologically to be liposarcoma.
Conclusion
Despite the huge size of retroperitoneal liposarcomas, they can be surgically resected successfully.

Keywords:
liposarcoma, retroperitoneal, sarcoma

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Introduction

Liposarcoma is considered the most common pathological type of soft tissue sarcoma that affects adults. Retroperitoneal liposarcoma may vary in weight and size reaching up to 20 kg, which is rarely reported, and is considered to be a 'giant liposarcoma'. Here, we report our experience of a giant retroperitoneal liposarcoma that weighed 44 kg and caused marked abdominal swelling, marked leg edema, and respiratory distress [9].

Case report

A 53-year-old male patient complained of progressive abdominal enlargement for ~10 months. When he sought medical advice, an ultrasound scan (US) of the pelvis and the abdomen revealed a large well-defined lobulated pelviabdominal mass that measured ~15×15 cm. According to that pattern, the US report suspected angiomyolipoma; therefore, a computerized tomographic (CT) scan on the chest, the abdomen, and the pelvis was performed looking for possible metastases and more accurate imaging of the tumor. The CT revealed a large pelviabdominal mass with enhancing soft tissue component inside that was displacing the intestinal loops highly suggesting liposarcoma as the type of the tumor (Fig. 1). An US-guided biopsy was performed, which was nonrepresentative.

After these investigations were done, the patient was referred to the Oncology Center of Mansoura University (OCMU). A CT-guided biopsy was performed, and the result was highly suggestive of liposarcoma. Then abdominal exploration was performed in October 2014. In the operation, we visualized multiple intra-abdominal and retro-peritoneal huge masses. We dissected all these masses and did a limited resection anastomosis of a part of the small intestine that was stretched by one of the masses. We also dissected another mass from the descending colon and the left ureter intact (Fig. 2).

On postoperative follow-up, the patient's condition was stable, and the laboratory investigation findings were normal. The patient was discharged 5 days after the operation.

The pathology report confirmed that the tumor was dedifferentiated liposarcoma. The resected specimens weighed 44 kg, making this tumor nearly one of the biggest reported resected liposarcomas in the world (Fig. 3).

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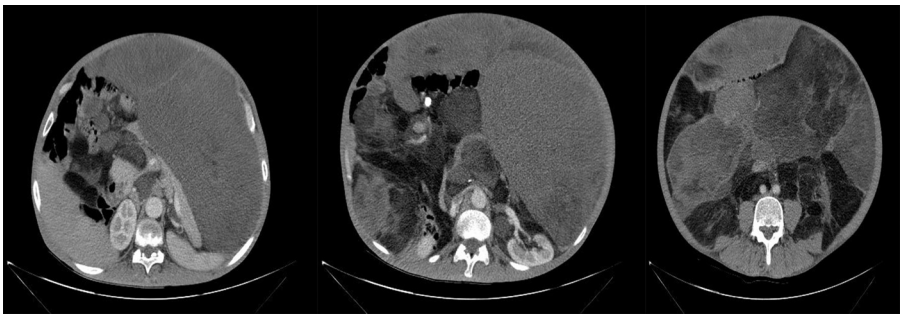
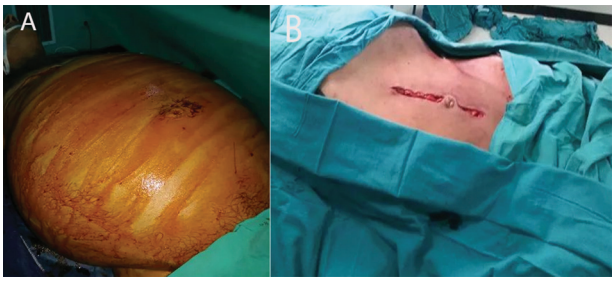
Figure 1



Computed tomography (CT) scan of the patient's abdomen and pelvis. CT scan revealed a large pelviabdominal mass with enhancing soft tissue component inside displacing the intestinal loops.

Figure 2



Patient pictures in the operating room (OR). (a) A picture before the skin incision. (b) A picture after the resection of the mass before skin closure.

Microscopically, the lesions consist of areas of atypical lipomatous neoplasm/well-differentiated liposarcoma and a nonlipogenic (dedifferentiated) component. The interface between the two zones is abrupt, although in some regions there is a gradual transition between the two components. The areas of well-differentiated liposarcoma consist of typical lipoblasts having hyperchromatic indented nucleus and vacuolated cytoplasm, whereas the dedifferentiated zones are formed of spindle-shaped cells that show hyperchromatic nuclei, scanty cytoplasm with indistinct cell borders, and separated by interwoven collagen fibers. These cells show moderate degree of pleomorphism with few scattered mitotic figures. There are multinucleated giant cells and giant cells of bizarre size admixed with inflammatory cellular infiltrate. Foci of divergent rhabdomyosarcomatous and leiomyosarcomatous differentiation are also present. Moreover, wide areas of necrosis are detected (Fig. 4).

The most common problem in the differential diagnosis is distinguishing between a pleomorphic sarcoma infiltrating the fat and a dedifferentiated liposarcoma. The presence of clear-cut evidence of atypical lipomatous neoplasm/well-differentiated liposarcoma some distance away from the dedifferentiated areas helps in the diagnosis of dedifferentiated liposarcoma.

Long-term follow-up and regular radiological scans were done on the patient looking for recurrence and survival.

Review of the literature

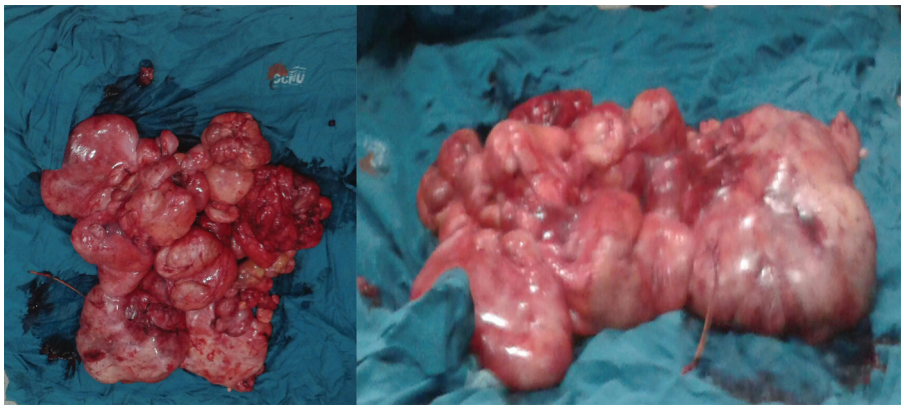
Liposarcoma is the most common type of soft tissue sarcoma that is encountered in 10–20% of all cases. It affects both sexes equally, with a peak age group of 40–60 years [9]. There are four histological subtypes of liposarcoma: well differentiated, undifferentiated, pleomorphic, and myxoid [10].

Several factors have been reported to affect the prognosis, namely, the histological subtype, tumor site, and the completeness of surgical resection. As an example, the overall 5-year survival rate is reported to reach 88–100% in myxoid and well-differentiated types. Moreover, the primary cutaneous liposarcoma has an indolent course with subsequently better prognosis, whereas deep high-grade sarcomas are reported to have a 5-year overall survival (OS) of ~50% [6].

Unfortunately, the myxoid and well-differentiated types are not well circumscribed and are accompanied by higher risk of local recurrence, particularly with incomplete surgical excision. This eventually leads to differentiation into higher grades and higher risk of metastases, which are more commonly encountered in the lungs followed by the liver [16].

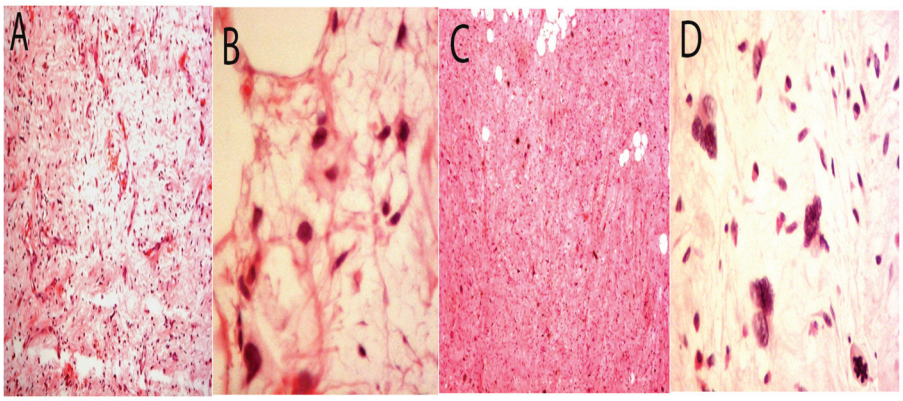
The risk of local recurrence and distant metastases is higher in both poorly differentiated and round cell types with a reported 5-year OS rate of 50%. The OS rate of dedifferentiated liposarcoma reaches up to 75%,

Figure 3



Tumor after resection. The resected specimens weighed 44 kg.

Figure 4



Histological features of dedifferentiated liposarcoma. (a) Areas of well-differentiated liposarcoma H&E: ×100, with numerous lipoblasts. (b) H&E: ×400. (c) Areas of dedifferentiated liposarcoma H&E: ×100, with scattered bizarre giant cells and mitosis. (d) H&E: ×40.

whereas it varies from 30–50% in the pleomorphic subtype depending on the tumor location [7].

Retroperitoneal liposarcoma has worse prognosis than limb liposarcoma; this can be attributed to the difficulty of attaining negative margins by surgical resection, as it is poorly circumscribed. As a consequence, retroperitoneal liposarcoma tends to recur more frequently and even may lead to death. It is commonly of large size (>10 cm) owing to the capacious potential space of the retroperitoneum [3].

CT and MRI remain the gold standard imaging tools in diagnosis of liposarcoma. On the contrary, ultrasonography is more commonly used during the follow-up period to detect any recurrence, which is more encountered in pleomorphic types than in the well-differentiated types [13].

CT reveals liposarcoma as a large space-occupying lesion that displaces or compresses the surrounding organs with a negative attenuation coefficient and sometimes internal septations. Well-differentiated liposarcomas appear mainly hypodense, whereas pleomorphic liposarcomas are heterogenous [13].

Under MRI, liposarcoma appears as a mass with a thick peripheral ring and linear septae with internal nodules and preserved contiguous structures. Well-differentiated liposarcomas have the same signal intensity as that of the subcutaneous fatty tissue. It is mainly formed of fat, thin or thick fibrous septae – that mainly appears hypointense in T1-weighed images – and intrinsic nodules. After intravenous contrast administration, no significant enhancement is noticed, and only a weak signal increase is detected compared with the unenhanced phase, which is better visualized in the fat-suppressed sequences. The myxoid type appears isodense compared with the muscles in MRI sequences. In the unenhanced phase, it appears mainly cystic. However, after intravenous contrast administration, it shows solid heterogenous pattern, where some areas show no enhancement, which corresponds histologically to areas of mucin

accumulation, and other areas appear more enhanced, which corresponds to areas of more cellular aggregation. The pleomorphic subtype appears heterogenous in T1 and T2 sequences owing to poor adipose content. Areas of hemorrhage and necrosis are mainly encountered. It is hypointense compared with fatty tissue in T1 sequences, whereas in T2 sequences, it appears hyperintense with heterogenous internal structure [14].

Risk assessment in patients with liposarcoma depends mainly on tumor grade and subtype. However, the tumor standardized uptake value obtained by positron emission tomography before any intervention was found to be a more useful parameter for risk assessment than tumor grade and subtype. Reduced survival and increased risk of early recurrence and distant metastasis were reported with SUV greater than 3.6 [15].

Biopsy is the gold standard diagnostic procedure for liposarcoma. Excisional biopsy is recommended for small, superficial, fatty tumors, whereas in case of deep large tumors (>3 cm), open incisional biopsy followed by definitive resection is the procedure of choice [4].

Surgery with or without radiotherapy remains the mainstay of treatment of liposarcoma. Moreover, there may be also a role for chemotherapy. Myxoid liposarcoma was found to be the most chemosensitive histological type, whereas well-differentiated and dedifferentiated types are less sensitive to chemotherapy. Multimodality treatment – including chemotherapy and radiotherapy – is the most effective in case of high-grade liposarcoma. Surgical resection should be done with wide safety margins to minimize the risk of recurrence and dedifferentiation. This could be challenging in some cases, as it could be hard to differentiate between the tumor and retroperitoneal fat. Moreover, it may be difficult to resect any adjacent organs. Achievement of complete surgical resection can be sufficient alone to obtain satisfactory outcomes. This can minimize the need for postoperative radiotherapy, that too which is mainly applied in large doses and fields owing to the size and location of liposarcoma [5,8].

Huge retroperitoneal liposarcomas have been reported many times in literature. In 2007, Haber *et al.* reported a giant retroperitoneal liposarcoma that weighed 10.7 kg and involved the descending colon in a 37-year-old male patient. Complete surgical resection and partial colectomy were done, and pathological examination revealed well-differentiated liposarcoma [17].

Hashimoto *et al.*, in 2010, reported a case of a 41-year-old male patient with huge retroperitoneal liposarcoma that was related to the right kidney. Complete surgical resection with scarification of the right kidney was done. Pathological examination revealed dedifferentiated liposarcoma and the specimen weighed 22 kg [2].

In 2011, Akhoondinasab and Omranifard reported complete surgical resection of a giant retroperitoneal liposarcoma in a 54-year-old male patient. The mass was dissected from the aorta, kidneys, and ureters and was excised *en bloc*. It weighed 32 kg, and pathological examination revealed a well-differentiated type [18].

Saini and Kapoor in 2012 reported a huge retroperitoneal liposarcoma in a 65-year-old male patient. It measured 54×43×38 cm^3 and was infiltrating the descending and sigmoid colon. Complete surgical resection and partial colectomy were done with an end colostomy procedure. Pathological examination revealed myxoid liposarcoma with areas of dedifferentiation, and the mass weighed 41 kg [19]. In 2015, Caizonne *et al.* reported a giant retroperitoneal liposarcoma in a 64-year-old female patient. She complained of progressive abdominal enlargement. CT scan of the abdomen and pelvis revealed a huge retroperitoneal mass that measured 42×37×18 cm^3 and extended from the subhepatic area to the pelvis displacing the right kidney to the left. Pathological examination revealed a pleomorphic liposarcoma with areas of myxoid changes originating from the perinephric fat of the right kidney and infiltrating the periureteral tissue [20].

Conclusion

The mainstay of treatment of retroperitoneal liposarcoma is complete surgical resection even in huge ones whenever possible.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor in chief of this journal.

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Conflicts of interest
There are no conflicts of interest.

References

1 Dei Tos AP, Mentzel T, Fletcher CD. Primary liposarcoma of the skin: a rare neoplasm with unusual high grade features. Am J Dermatopathol 1998; 20:332–338.

2 Hashimoto Y, Hatakeyama S, Tachiwada T, Yoneyama T, Koie T, Kamimura N, *et al.* Surgical treatment of a giant liposarcoma in a Japanese man. Adv Urol 2010.

3 Coindre JM, Mariani O, Chibon F, Mairal A, De Saint Aubain Somerhausen N, Favre-Guillevin E, *et al.* Most malignant fibrous histiocytomas developed in the retroperitoneum are dedifferentiated liposarcomas: a review of 25 cases initially diagnosed as malignant fibrous histiocytoma. Mod Pathol 2003; 16:256–262.

4 Crago AM, Singer S. Clinical and molecular approaches to well differentiated and dedifferentiated liposarcoma. Curr Opin Oncol 2011; 23:373–378.

5 McCallum OJ, Burke JJ, Childs AJ, Ferro A, Gallup DG. Retroperitoneal liposarcoma weighing over one hundred pounds with review of the literature. Gynecol Oncol 2006; 103:1152–1154.

6 Nishida Y, Tsukushi S, Nakashima H, Ishiguro N. Clinicopathologic prognostic factors of pure myxoid liposarcoma of the extremities and trunk wall. Clin Orthop Relat Res 2010; 468:3041–3046.

7 Singer S, Antonescu CR, Riedel E, Brennan MF. Histologic subtype and margin of resection predict pattern of recurrence and survival for retroperitoneal liposarcoma. Ann Surg 2003; 238:358–370. discussion 370–371.

8 Rutkowski P, Trepka S, Ptaszynski K, Kołodziejczyk M. Surgery quality and tumor status impact on survival and local control of resectable liposarcomas of extremities or the trunk wall. Clin Orthop Relat Res 2013; 471:860–870.

9 Hogg ME, Wayne JD. Atypical lipomatous tumor/well-differentiated liposarcoma: what is it? Surg Oncol Clin N Am 2012; 21:333–340.

10 Nijhuis PH, Sars PR, Plaat BE, Molenaar WM, Sluiter WJ, Hoekstra HJ. Clinico-pathological data and prognostic factors in completely resected AJCC stage I–III liposarcomas. Ann Surg Oncol 2000; 7: 535–543.

11 Gardner JM, Dandekar M, Thomas D, Goldblum JR, Weiss SW, Billings SD, *et al.* Cutaneous and subcutaneous pleomorphic liposarcoma: a clinicopathologic study of 29 cases with evaluation of MDM2 gene amplification in 26. Am J Surg Pathol 2012; 36: 1047–1051.

12 Ishida H, Naganuma H, Konno K, Sato M, Hamashima Y, Konno S, *et al.* Retroperitoneal liposarcoma: sonographic findings. Abdom Imaging 2000; 25:554–558.

13 Barile A, Zugaro L, Catalucci A, Caulo M, Di Cesare E, Splendiani A, *et al.* Soft tissue liposarcoma: histological subtypes, MRI and CT findings. Radiol Med (Torino) 2002; 104:140–149.

14 Arkun R, Memis A, Akalin T, Emin Ustun E, Sabah D, Kandiloglu G. Liposarcoma of soft tissue: MRI finding with pathologic correlation. Skeletal Radiol 1997; 26:167–172.

15 Brenner W, Eary JF, Hwang W, Vernon C, Conrad EU. Risk assessment in liposarcoma patients based on FDG PET imaging. Eur J Nucl Med Mol Imaging 2006; 33:1290–1295.

16 Dalal KM, Antonescu CR, Singer S. Diagnosis and management of lipomatous tumors. J Surg Oncol 2008; 97:298–313.

17 Hrabar D, Sulentic P, Tomasic V, Beslin MB, Bilic B, Pavic T, *et al.* Giant "lipoma like" liposarcoma of the retroperitoneum: a case report. Acta Clin Croat 2007; 46:321–323.

18 Akhoondinasab MR, Omranifard M. Huge retroperitoneal liposarcoma. J Res Med Sci 2011; 16:565–567.

19 Saini NS, Kapoor R. Giant retroperitoneal liposarcoma. JK Sci 2012; 14:51–53.

20 Caizzone A, Saladino E, Fleres F, Paviglianiti C, Iaropoli F, Mazzeo C, *et al.* Giant retroperitoneal liposarcoma: case report and review of theliterature. Int J Surg Case Rep 2015; 9:23–26.