Recurrent giant retroperitoneal liposarcoma with 10 years follow up. Case report and review of literature

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1. Introduction

Retroperitoneal Liposarcomas are a malignant transformation of fat tissue, found in two major subtypes according to their differentiation. Enhanced CT scanning of the abdomen and pelvis is essential for diagnosis, which can be confirmed by performing a transcutaneous core needle biopsy. Surgery remains the standard practice in treating non-metastatic liposarcomas [1].

The patient discussed in this report was diagnosed with a well-differentiated liposarcoma that was then fully excised. Over the course of 10 years, he had 3 more recurrences and 2 more surgeries with the same pathology. This is the longest duration of follow-up in this type of tumor reported in the English literature.

This case was reported in accordance with the SCARE criteria [2].

2. Case description

A 70-year-old man, heavy smoker with a history of hypertension, hyperlipidemia, and diabetes presented to our outpatient clinics with dizziness and fatigue. His past surgical history included a left carotid endarterectomy and partial distal gastrectomy with Billroth 1 anastomosis for complicated peptic ulcer disease. The physical examination showed a distended abdomen and hepatomegaly. Chest auscultation was normal. The rest of the exam was non-significant.

Routine labs were ordered, and the patient was found to have anemia (hemoglobin 9.1 mg/dl). Gastroscopy and colonoscopy were performed but showed no suspicious lesions or bleeding. CT scan of the abdomen (complemented by an MRI) was done and showed a large retroperitoneal mass extending from the posterior subdiaphragmatic region down to the pelvis and reaching the right

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superolateral vesical wall. The mass severely displaced the intraabdominal viscera to the side. The right kidney was also displaced anteromedially. The vascular structures including the mesenteric and inferior vena cava at the intrahepatic level are slightly compressed but still patent. The mass was isointense, resembling subcutaneous fat. Hypo signal heterogeneous STIR sequence confirmed that the mass was of fatty composition, which lead to a preliminary diagnosis of a retroperitoneal liposarcoma. No invasion of nearby structures was mentioned (Figs. 1 and 2).

The patient was scheduled for laparotomy. In the operation room, a right double J catheter was first inserted to protect the ureter. A midline incision extending from the subxiphoid to the suprapubic area was then done (Fig. 3). After reaching the retroperitoneal space and identification of the limits of the tumor, en bloc removal was achieved after liberating it from the surrounding structures followed by lymph node harvesting (Fig. 4). The mass weighed 9 kg and measured 50 × 30 × 18 cm (Fig. 5). A 1.5 cm extrinsic nodule found during bowel inspection was also excised from the distal ileal loop.

The patient had a smooth postoperative course and was discharged home on the 6th postoperative day.

The mass was adipose, heterogeneous, and had necrotic areas. Microscopically, the tumor showed lobulated neoplastic prolifer-

ation made up of mature adipose cells of variable size with some hyperchromatic and irregular nuclei (Figs. 6 and 7). Necrotic foci with lymphoplasmocytic infiltrate were observed. The diagnosis of well differentiated liposarcoma was made. A fibrohyaline calcification was attributed to the resected ileal nodule.

Three months after the initial surgery, a follow-up PET/CT scan was performed and showed no signs of local recurrence (Fig. 8).

Five years later (2015), the patient was readmitted for dizziness and was found to have microcytic anemia (Hemoglobin = 8.9 mg/dl). Occult blood was negative. An endoscopic evaluation also failed to explain his anemia. An abdominal CT scan (complemented by MRI) showed a 12 × 11 cm mass in the right infrarenal space, isointense with parietal fat, and with good contrast uptake. This time the mass was pressing on the right colon. This was in favor of recurrence of the previously operated retroperitoneal liposarcoma.

The patient underwent a second laparotomy for mass removal. This time the mass was found to be multilobulated and engulfing the inferior vena cava and right ureter. Complete liberation was achieved, but en bloc excision was not possible due to the encountered difficulty. The massed was excised in two pieces, along with the appendix that was adherent to it. The patient ameliorated well postoperatively. Pathology reported a well-differentiated

Fig. 1. CT scanner and MRI: 450 × 250 mm right retroperitoneal liposarcoma pushing the kidney and bowels to the other side.
 retroperitoneal liposarcoma with the same findings seen with the original tumor. The patient was started on chemotherapy, and a PET CT scan follow-up 6 months later showed no focal recurrence.

Four years later (2019), a follow up enhanced CT Scan of abdomen and pelvis showed the presence of a right retroperitoneal multilobulated mass measuring 23.5 × 17 × 16 cm, displacing the pancreas and other viscera, and compressing the right colon. The aspect was suggestive of a local recurrence of the retroperitoneal liposarcoma. The scan also noted the presence of a right inguinal hernia with a multinodular adipose component of 5 × 3 cm occupying the right scrotal bursa and pushing away the testicle infero-posteriorly. This was suggestive of a right para-testicular liposarcoma.

The patient was admitted for a third operation. Intraoperatively, a midline laparotomy was done. Excision of the retroperitoneal mass measuring 16 × 10 × 7 cm was done. The mass was extending towards the inguinal canal, so an inguinal incision was performed. A nodular adipose mass of 2.3 × 1.6 × 0.6 cm was identified and excised. On histopathology, both tumors were consistent with a well-differentiated liposarcoma, similar to the ones described previously.

One year later (2020), MRI showed local recurrence of the retroperitoneal mass, measuring now approximately more than 22 cm in axial diameter and exceeding 12 cm in AP diameter. It is extending anteriorly to the pre-aortic space and left anterior para-renal space, displacing the pancreas and the viscera anteriorly with no sign of invasion (Fig. 9). A fourth laparotomy was offered to excise the mass.

The timeline of diagnosis, surgeries and recurrences is resumed in Fig. 10.
Fig. 4. Intraoperative Photo: Excision of the retroperitoneal mass en bloc.

Fig. 5. Intraoperative Photo: Giant Liposarcoma excised en block.

Fig. 6. Well-differentiated liposarcoma, mature adipocytes and atypical cells (×100).
Fig. 7. Well-differentiated liposarcoma, atypical cells with inflammatory cells (x400).

Fig. 8. PET-CT Scan of the abdomen and pelvis: No FDG enhancement.
3. Discussion

Liposarcomas are rare tumors that are most commonly found in the extremities, followed by the retroperitoneum as the second most common location. The latter location represents a poorer prognosis [3]. Most of the time, Retroperitoneal liposarcomas, are incidentally found on non-related imaging of the abdomen, and they are usually asymptomatic despite their large size, but can later lead to abdominal pain, a palpable mass, or bowel obstruction [4].

There are multiple subtypes of retroperitoneal liposarcomas with a diverse genomic variation, and they are usually classified into well-differentiated, dedifferentiated, pleomorphic, and myxoid types [3]. The dedifferentiated component is usually more belligerent and can metastasize to secondary locations, thus, assessment of extension and the presence of secondary lesions is essential [1]. Well-differentiated and dedifferentiated liposarcomas consist 40% of the retroperitoneal sarcomas in adults aged over 55 years. Differential diagnosis of retroperitoneal soft tissue masses includes undifferentiated pleomorphic sarcoma, synovial sarcoma, solitary fibrous tumor, extraosseous Ewing’s sarcoma, and malignant peripheral nerve sheath tumor [4].

The diagnosis is usually done by enhanced CT of the abdomen and pelvis [1]. The low attenuation fat content of liposarcomas is usually distinctive. On ultrasonography, liposarcomas are classically hyperechoic [5]. Calcification can be present and they usually indicates poor prognosis and dedifferentiation, or represent inflammatory or a sclerosing variant of the well differentiated liposarcoma [4]. MRI is thought to be more sensitive in detecting retroperitoneal liposarcomas [5]. FDG PET/CT is not used in the routine diagnosis of these lesions due to the vast variability of histological types and tumor grading [4]. Certain subtypes have metastatic potential [3], so a full staging CT scan may be required [4].

When in doubt, and radiology can’t confirm the diagnosis, multiple percutaneous large core needle guided biopsies of the solid tumor can safely be done. The risk of needle tract seeding is minimal [4]. Fine-needle aspiration (FNA) cytology is not performed because it rarely provides enough information. A surgical incisional biopsy is also not recommended, since it put the peritoneal cavity at risk of contamination by sarcoma, alters plane of dissection, and may not provide diagnostic tissue. Sometimes ultrasound-guided endoscopic-biopsy can be performed [4].
Histological sampling is essential to establish the diagnosis, and plan proper treatment and management [4]. Well-differentiated liposarcomas are found to be a lobulated or round mass of macroscopic fatty components and thin septations. In contrast to dedifferentiated liposarcomas, variable enhanced densities are found within nodular formations associated with thicker septations and calcifications [1]. The latter show amplification of genes such as MDM2 and CDK4 confined to the chromosome region 12q13-15 [1].

Retropertitoneal liposarcomas don’t usually respond to chemotherapy. In contrast, Banvalot et al. showed a limited benefit in some types, where neoadjuvant radiotherapy followed by surgery was superior to surgery alone [6]. New studies nowadays are focusing on the new era of targeted genomic therapies [3].

Retropertitoneal Liposarcomas represent a big challenge to surgical resection, and the ultimate goal is achieving free margins with complete macroscopic R0/R1 resection [4]. This could be achieved by adequate preoperative planning and evaluation, a good estimation of organ, vessels, and nerve involvement, along with an assessment of extra-abdominal extension of the tumor [4]. Thus multiorgan resection is sometimes required [4]. Multiple subtypes may be found together with a hasty transition between them [1], but this should not be confused with multifocality which is considered a bad prognosis [4]. Resection is contraindicated when there’s metastasis, involvement of trunk vessel, or extensive spinal cord or bone involvement [4].

Follow-up imaging is directed by the final pathology, but in general, it is recommended to follow up every 3–6 months for the first 5 years, and then annually afterwards, as the risk of recurrence never plateaus [4]. Death is usually related to locoregional recurrence [6].

To describe the significant size of the tumor at the time of diagnosis, but there is no clear definition for this entity. By reviewing
the English literature, with possible content access, we found 52 cases published between 1998 and July 2020 under the label of giant retroperitoneal liposarcoma (Table 1). 44.2% were females whereas 55.8% of cases were males (Female-to-male ratio 0.79:1), with a mean age of presentation being 57 years old ± 14 years. Complete resection was achieved in 96.15% of cases, while R0 resection couldn’t be done in only 2 of the reported cases. The final diameter of the mass was reported in 47 cases, and it ranged from 17.5 cm to 80 cm with a mean of 42 cm, whereas mass weight was reported in 44 cases, ranging from 3.15–47 kg with a mean of 18.9 kg.

The histological repartition is summarised in Table 2. There was a dominance of the well-differentiated subtype, which constituted 50% of cases, followed by dedifferentiated liposarcoma with 19.2%. Post-surgery follow-up was reported in 42 cases, with a maximum reported follow-up of 108 months. Recurrence was reported in 21% of cases, of which 72% were reoperated.

Our patient has the longest reported follow up in the literature of 10 years, with 3 operations and 3 recurrences.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

The study type is exempt from ethical approval.

Consent

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 contribution

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Registration of research studies

N/A.

Guarantor

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