Surgical approach on recurrent retroperitoneal liposarcoma with involvement of surrounding organs – A Case Report

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Abstract

Background: Retroperitoneal sarcomas are neoplasms that occupy only 0.3 – 3% of all solid tumors. Liposarcomas are the most frequent soft tissue sarcomas in adults. Symptoms would only be detected if the liposarcoma infiltrates on the surrounding organs.

Case presentation: 58-year-old woman presented with complaints of progressive abdominal distension, loss of appetite, malaise, constipation and weight loss of about 7 kg. Abdominal CT scanning showed the presence of a retroperitoneal tumor that occupied almost the entire right part of abdominal cavity. 16 months earlier, the patient was operated in another surgical facility due to liposarcoma, derived from retroperitoneum (Histopathological finding: Poorly differentiated (G3) liposarcoma pleomorphic cell type). The patient was operated under the diagnosis of retroperitoneal liposarcoma.

Conclusions: Treatment of choice is radical surgical procedure including elimination of all the structures and organs involved by the tumour process - RO resection as a basic principle of surgical treatment.

Keywords: *giant retroperitoneal liposarcoma, dedifferentiated liposarcoma*

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Full Text

Introduction

Virchow first described liposarcoma in the 1860s. Liposarcoma (LPS) is a malignant tumor of mesenchymal origin. (1) Retroperitoneal liposarcoma is a rare tumor, with an incidence of 2.5 per million, accounting for 10-15% of soft tissue tumors. (2) The dimension and weight of liposarcoma are variable; those over 20 kg are called "giant liposarcoma". (3) Liposarcomas are one the most common of over 50 histologic subtypes of soft tissue sarcomas that are mostly resistant to chemotherapy. (4)

These tumors are classified into subtypes according to their pathological features including the amount of intracellular lipid, the quantity of mucoid lipid, and the differentiation of cells. There are three separate biologic groups of LPS encompassing five histologic subtypes. A fifth subtype (the so-called "mixed or combined liposarcoma"), which was still a separate entity in the 2002 WHO classification, has been removed from the most recent 2013 WHO classification, based on the consensus view that those rare cases probably represent examples of (variants of) dedifferentiated liposarcoma.

The recently updated World Health Organization (WHO) classification of soft tissue and bone tumors recognizes four major liposarcoma subtypes: (1) atypical lipomatous tumor/well-differentiated liposarcoma [which includes the adipocytic (or lipoma-like), sclerosing, inflammatory and spindle cell variants], (2) dedifferentiated liposarcoma, (3) myxoid liposarcoma, and (4) pleomorphic liposarcoma. (5,6)

There are three biologic groups of liposarcoma: well- and dedifferentiated liposarcoma (WD/DDLS), myxoid/round cell liposarcoma (M/RCLS) and pleomorphic liposarcoma. (7) Although well differentiated and dedifferentiated subtypes share common morphological features, dedifferentiated liposarcoma has much higher malignant potential with poor prognosis.

The anatomical distribution of liposarcoma appears to be partly related to the histologic type. Well-differentiated liposarcoma tends to occur in deep soft tissues of both the limbs and the retroperitoneum, and dedifferentiated liposarcoma occurs predominantly in the retroperitoneum. Although any liposarcoma subtype occasionally arises in the subcutis, involvement of the dermis appears to be exceedingly rare.

Case presentation

58-year-old patient admitted for elective operative treatment due to intra-

abdominal tumor formation that was verified on CT on the abdomen and small pelvis with contrast.

In the last few months she experienced weight in the stomach, loss of appetite, malaise, constipation, swelling of the right leg, movement difficulties and weight loss of about 7 kg. On a physical examination, a palpably accessible TU formation is recorded in the right half of the abdomen, which in the mesogastric region passes towards the anterior abdominal wall.

16 months earlier, the patient was operated in another surgical facility due to liposarcoma derived from retroperitoneum (Histopathological finding: Poorly differentiated (G3) liposarcoma pleomorphic cell type).

Abdominal CT scanning showed the presence of a retroperitoneal tumor that occupies almost the entire right part of abdominal cavity. The CT scan with contrast of the abdomen shows a large TU conglomerate originating from the right retroperitoneal space and it is in collision with the lower vena cava, and compresses the duodenum, stomach and transverse colon and high sited right dome of the diaphragm. Aorta and mesenteric vessels with small intestines and part of the colon are visualized in the left half of the abdomen ***(Figures 1, 2 and 3).***

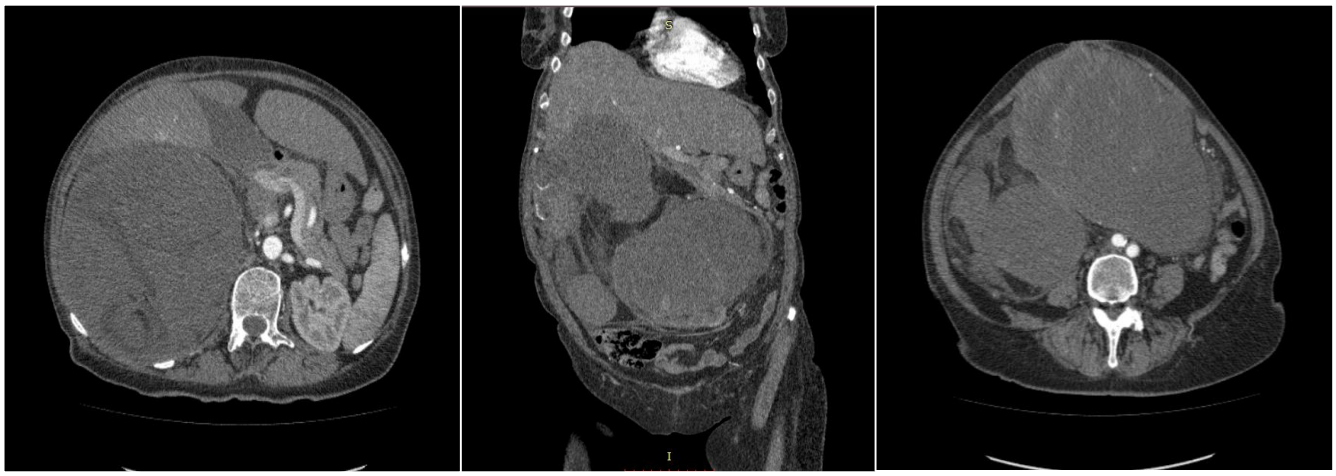

Figure 1,2,3: *Abdominal CT scanning showed the presence of a retroperitoneal tumor that occupies almost the entire right part of abdominal cavity. Large TU conglomerate originating from the right retroperitoneal space and it is in collision with the lower vena cava, and compresses the duodenum, stomach, transverse colon and high sited right dome of the diaphragm. Aorta and mesenteric vessels with small intestines and part of the colon are visualized in the left half of the abdomen.*

The right kidney cannot be located and defined at the ultrasonographic examination of the abdomen. From laboratory parameters: elevated values

of WBC 14.4, PLT 638 and C reactive protein - 196mg/L, the remaining laboratory parameters in order. Pre-operative J-J probe is placed in the left ureter, while in the right ureter the attempt to set the J-J probe was unsuccessful. The patient was operated under the diagnosis of retroperitoneal liposarcoma. A large conglomerate of tumor masses localized in the right half of the abdomen is recorded intraoperatively.

The tumor originates from the right hemiretroperitoneum and extends at a distance from the right diaphragmatic dome (which raises it in the right hemithorax and for which it is intimately attached) to the right iliac fossus where it lies on the right iliac vein. The upper half of the tumor is covered by the liver. The tumor collides with the right kidney, the right adrenal gland, the cava inferior vein, and compresses the duodenum, stomach and transverse colon. The aorta and mesenteric vessels with small intestines and part of the colon are swollen in the left half of the abdomen.

Block resection was made of the tumour together with the right kidney and the right adrenal gland, part of the right dome of diaphragm, cholecyst and partial resection of musculus ileopsoas from the right side. The specimen measured 36x22x14cm and weighted 7765 grams ***(Figures 4, 5, 6).***

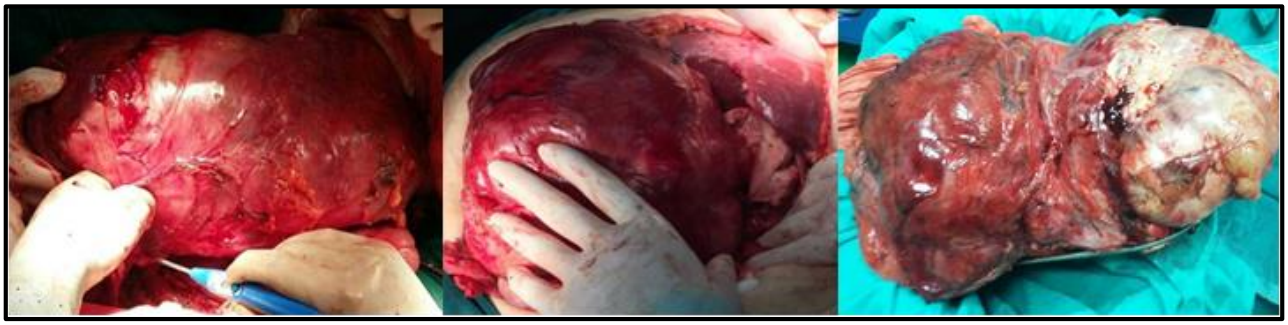

Figure 4: A large conglomerate of tumor masses localized in the right half of the abdomen.

Figure 5: The tumor originates from the right hemiretroperitoneum and extends at a distance from the right diaphragmatic dome to the right iliac fossus where it lies on the right iliac vein. The upper half of the tumor is covered by the liver. The tumor collides with the right kidney, the right adrenal gland, the cava inferior vein, and compresses the duodenum, stomach and transverse colon. The aorta and mesenteric vessels with small intestines and part of the colon are swollen in the left half of the abdomen.

Figure 6: The specimen measured 36x22x14cm and weighted 7765 grams.

The intersection of the tumor is encapsulated, without a macroscopically visible penetration of the capsule. On serial sections, the tumor tissue in the part is solid-colored, in part is necrotic, in

part it is haemorrhagic, and in part with the appearance of fat tissue.

Microscopic analysis showed a malignant mesenchymal neoplasm built from liposarcoma sites with variable degrees of differentiation from low to high degrees, bleeding and necrosis zones, as well as elongated cell lines in a reversed and trackable arrangement between which are large bizarre, single multidirectional cells, with the presence of mythological activity, that is, areas with features of fibrosarcoma.

Histopathological finding: Liposarcoma high grade (Dedifferentiated type) - a dedifferentiated liposuction containing a liposarcomate component and poorly differentiated fibrosarcoma. pTNM = G3 L0 VO NG3

Discussion

Liposarcoma is the most common soft tissue sarcoma, accounting for 10-20% of all cases. It commonly occurs between 40-60 years of age and has a 1:1 ratio between genders (8).

Based on its histological features, liposarcoma was initially classified into four subtypes: the well differentiated, myxoid, round cell and pleomorphic types. Evans introduced dedifferentiated liposarcoma later in 1979, so now there are five subtypes. (9)

The feature of dedifferentiated liposarcoma is the histological coexistence of well to poorly differentiated liposarcoma and non-lipomatous differentiated areas. (10) A dedifferentiated liposarcoma of the retroperitoneum is an extremely rare tumor. Dedifferentiated liposarcoma has a worse prognosis than other subtypes because it has a higher risk of local recurrence and distant metastasis. Retroperitoneal liposarcoma is usually asymptomatic until the liposarcoma is large enough to compress the surrounding organs. (11) Only complete excision provides hope for a cure, but this is often difficult because the margins are not grossly apparent, so a contiguous organ may have to be resected. For unresectable disease, retrospective studies have identified myxoid (round cell) and pleomorphic sarcomas to be relatively responsive to chemotherapy. (4)

In retroperitoneal liposarcoma, histological subtype, incomplete resection, contiguous organ resection, and older age are strongly associated with tumor-related mortality. For liposarcoma, it is necessary to customize the treatment strategy on a case-by-case. (12)

High histological grade is one of the most important negative prognostic factors in patients with retroperitoneal liposarcoma.

Generally, round cell, pleomorphic and dedifferentiated subtypes are regarded as high-grade; whilst well-differentiated

and myxoid liposarcoma are low-grade. (13) Well-differentiated liposarcomas may recur locally, but the metastatic potential is low, while pleomorphic liposarcomas have high metastatic potential, which may reduce the survival rate. Although well differentiated and dedifferentiated subtypes share common morphological features and genetic events, their biologic behaviors are quite different.

Well-differentiated liposarcoma often recur locally but have minimal metastatic potential, with a 5-year survival probability of 90%. (14) A dedifferentiated tumor tends to present more often as a recurrence, requires multi-organ resection more frequently, and has a shorter disease-free interval compared to those of well differentiated subtypes. Multiple factors, such as site and depth of origin, margin involvement after resection, and histologic grade affect survival rates for patients with liposarcoma. Factors associated with a poor survival prognosis include the dedifferentiated subtype, grade 2-3, stage II-III, size >20 cm, and involved surgical margins. Distant metastasis is more common with dedifferentiated, grade II-III, and the deep seated (retroperitoneal) tumor location. Distant metastasis relates to the tumor size. If the size of the tumor is less than 2.5 cm, the rate of metastasis at 5 years is approximately 3%. On the other hand, the rate of metastasis at 5 years is between 55 and 60% in cases of tumors larger than 20 cm. (15) Local recurrence remains the preliminary cause of mortality in retroperitoneal liposarcoma (16). Aggressive surgical approach over 80% of patients with dedifferentiated histology will recur locally and 30% will metastasize to distant sites within 3 years of diagnosis. (14)

In order to detect recurrence, a CT scan every 3 months for the first 2 years, every 6 months for 2–5 years and annually thereafter is generally recommended. Each recurrence should be included in a program of iterative resection as this is the only guarantee of prolonged survival. (17) The survival rate was improved in the patients who received a complete resection of their recurrent tumor compared with the patients who did not. Therefore, the gold standard treatment remains to be removal of the recurrence (18). Although controversy exists, adjuvant or neoadjuvant radiotherapy or chemotherapy do not have any survival benefits in retroperitoneal liposarcoma. In order to achieve the R0 resection, contiguous organ resection, including nephrectomy, is always performed when the intraoperative histological results are dedifferentiated, myxoid/round cell, pleomorphic or mixed-type. (15) Milone et al reported a 5 year survival of 85.7% in R0 resection versus 33.3% in R1 resection. (16) Because dediferentiated liposarcomas have high malignant potential with risk of recurrence and

distant metastasis, careful surveillance is warranted.

The resectability of the tumor and histopathologic grades are associated with disease prognosis. (19)

Local recurrence following complete resection of primary retroperitoneal liposarcoma is common with 50% of well-differentiated and 80% of dedifferentiated tumors recurring within 5 years, and most patients who die of retroperitoneal soft tissue sarcoma die from the efects of local recurrence (20).In this case there was no verification of signs for recurrence or distant metastatic changes in connection with liposarcoma during control and standard examinations after one year of the surgical treatment.

Conclusions

Complete surgical resection is the gold standard treatment, which might be curative. In many cases, combined resection of involved organs and vasculatures is required to achieve complete resection. Hemo and radiotherapy do not show usefulness in dedifferentiated type of liposarcoma versus other subtypes in which there are signs for an effect of certain hemo therapeutic means. In the cases of dedifferentiated liposarcoma, close follow up and careful surveillance are warranted for the detection of recurrence.

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