Recurrence of Giant Retroperitoneal Liposarcoma: A Case Report and Literature Review

***Ehab Sadeq El-Hussieny, [1]Maha Mohammed Saqnawi, [2]Banan Ali Najmi, [3]Nawaf Najim Alzahrani, [4]Sawsan Adil Khan**

*Surgical oncology consultant, King Abdullah medical city, Makah, KSA,

[1,2,3,4] King Abdullah medical city, Makah, KSA

Abstract

A giant liposarcoma derived from the retroperitoneum have rarely been reported. Here we will describe a case of male patient with giant retroperitoneal liposarcoma measuring about 34 X 22.5 x 35.7 cm that was occupying the left abdominal and pelvic cavities and displacing the surrounding structures. The tumor was treated by surgical resection with intra operative radiotherapy. The mass was histopathologically identified as malignant adipocytic tumors and showed recurrence after 10 months.

Introduction

Sarcoma can be mainly divided into liposarcomas, leiomyosarcomas and malignant fibrous histiocytomas. [1] Liposarcoma is a tumor of lipoblasts which is a rare mesenchymal neoplasm that involves deep soft tissues. [2] Retroperitoneal sarcoma is a rare cancer account about 15% of all soft tissue sarcomas. [3] Liposarcoma account about 40% of all sarcomas [4] and bout 30% of the retroperitoneal malignancy are liposarcoma. [5] Patients affected by this tumor usually have no symptoms. Symptoms develop only when the tumor grows large enough to exert a mass effect on surrounding structures. Diagnosis is made when the tumor is large and puts adjacent vital organs at risk. [6] Surgery can be challenging but it is the main stay of treatment along with adjuvant therapies. Complete resection of the tumor and a favorable histopathologic grade are positively associated with long term disease free survival. Compared with other retroperitoneal sarcoma subtypes, liposarcoma of this area shows a relatively better prognosis. Effects of chemotherapy and radiation therapy have also been reported. Compared with other types of tumors, the liposarcomas' biggest feature is a significant local recurrence rate and it has a higher recurrence rate compared with other liposarcomas in other parts of the human body. [7] Frequently it recurs within 2 years of the initial surgical resection. For the early detection of recurrent retroperitoneal liposarcomas, a shorter follow-up interval with CT or MRI would be helpful. [8]

Case report

We present a case of a 60-year-old male patient who was referred to the surgical clinic from peripheral hospital as a case of abdominal mass for investigation. The patient reported history of vague abdominal pain and distention distention for 7 months prior to his presentation associated with nausea, significant weight loss and loss of appetite. He had medical history of diabetes and hypertension, had no relevant surgical history.

On physical examination indicate a good performance status, abdominal examination revealed distended abdomen, a giant non tender mass occupying most of the abdomen mainly in the midline and left side of the abdomen, measuring about 30 x 20 cm oval in shape.
The laboratory examination showed hemoglobin 11.6 g/dL, leukocyte count 6.25 x 10^9/L, platelet count 243 x 10^9/L, serum creatinine 1.5 mg/dL, direct bilirubin 0.24 mg/dL, total bilirubin 0.078 mg/dL, AST 14 U/L, ALT 13 U/L, total protein 7.3 gm/dL, albumin 2.73 gm/dL and ALP 96 U/L.

Ultrasound KUB reported the right kidney is measuring 9.1 x 5.0 cm cortical thickness of 1.4 cm. The left kidney could not be visualized due to the large mass for further evaluation by cross-sectional imaging. The urinary bladder demonstrates no stones.
Computed tomography [CT; Fig. 1] demonstrate large left retroperitoneal mass occupying the left abdominal and pelvic cavities, displacing surrounding structures. Due to the presence of soft tissue and fat components, this can represent a primary sarcoma or liposarcoma, with possible invasion of or arising from left Psoas

muscle. No focal liver lesions or intrahepatic duct dilatation. Portal and hepatic veins are normally opacified. The gallbladder is unremarkable. No definite intrathoracic metastasis. No aggressive bone lesions. No definite involvement of the lumbar vertebral bodies with primary left retroperitoneal mass.

Magnetic resonance imaging [MRI; Fig. 2] showed left large retroperitoneal mass measuring a 34 X 22.5 x 35.7 cm, in keeping with the patient diagnosis of liposarcoma and is associated with mass effect on the adjacent bowel loops and is seen pushed to the right side. The left kidney and the left adrenal glands are pushed to the right side.
Imaging guided true cut biopsy done, the histopathological examination revealed Malignant spindle cell neoplasm, probably liposarcoma (well differentiated, sclerosing subtype).

The patient underwent surgical resection, finding was huge retroperitoneal mass measuring about 55x55 cm and weighted 18 Kg [Fig. 3], the mass was found encasing the left kidney, left ureter, left gonadal vessel and left spermatic cord, was attached to the left side of vertebra, the left colon and spleen. The left colon, abdominal aorta and IVC found displaced to the right side, the stomach and the pancreas were found compressed and shifted superiorly by the tumor due to mass effect. All Small and large bowel was shifted to the right side. En-block resection of the mass involving the left kidney, left ureter, left gonadal vessels, left spermatic cord and. Intra-Operative radiotherapy was done given 13 Gy.

The mass was histopathologically identified as malignant adipocytic tumors, dedifferentiated liposarcoma. Grade 3.
The patient recovered well and was discharged 1 Week following the surgery. After discharge he received radical doses of radiation 45 Gy / 4 weeks. And the patient refused trial of palliative Doxorubicin and Ifosfamide and he lost follow up.
10 months following the surgery, recurrence was observed in computed tomography which showed 13.5 x 21.3 x 25 cm left retroperitoneal soft tissue mass suggestive of tumor recurrence, inseparable from major abdominal structure [Fig. 4].

The patient is currently under palliation.

Discussion

Liposarcomas are uncommon malignant mesenchymal tumors with adipocytic differentiation. [9]
It classified in to four principal subtypes: Well-differentiated (WD), Dedifferentiated (DD), Myxoid (MD) and Pleomorphic (PL) liposarcomas. The Well-differentiated liposarcoma (WDLS) is the most common subtype about (40%) of liposarcomas, it grows slowly and generally does not metastasis. It is associated with local recurrence and insensitivity to radiotherapy and chemotherapy. Dedifferentiated liposarcoma (DDLS) represents focal progression of well-differentiated disease into a more aggressive form, with lower risk of distant metastases but higher risk of local recurrence, it accounts about (10%) of all liposarcomas. [10] Myxoid liposarcoma (MLS) is the second most common type of liposarcoma account about (20%), it tends to grow more slowly. It has a metastatic potential and tend to show good prognosis. [11] Pleomorphic liposarcoma (PLS) is extremely rare (5%) of liposarcomas. It accounts for fewer than 5% of all liposarcomas. It has a quicker metastatic potential and tend to show poor prognosis. [10] Liposarcomas can occur in different anatomic regions and can be found throughout the body, most commonly in the extremities, head and neck, trunk, retroperitoneum and mesentery. [12]

Retroperitoneal liposarcomas worldwide incidence varies between 0.07% and 0.2% of all retroperitoneal tumours. Retroperitoneal liposarcoma has a peak incidence in mid-fifties, with an equal sex distribution. The retroperitoneum has no bony boundaries; therefore, no immediate symptoms were highlighted, and it usually diagnosed due to mass effect of the large tumor, such as early satiety or abdominal distension. [13]
Although the gold standard for diagnosis remains biopsy, imaging with computed tomography (CT) or magnetic resonance imaging (MRI) currently are widely accepted diagnostic tool. The presence of macroscopic fat on imaging suggests the presence of LPS. [14]

The dedifferentiated subtype found to be the most reported subtype in the literature of giant retroperitoneal liposarcomas about (75%) of all DDLS and about (25%) found in the extremities. DDLS can arise de novo or as a recurrence of WDLS. [15]

The most important favorable prognostic factor is the complete resection of the tumor with negative margins

and the histologic subtype. Because of the ineffectiveness of current chemotherapy and the requirement of high radiation doses, aggressive surgical excision remains the gold standard treatment even for the localized recurrences of retroperitoneal liposarcoma, however adjuvant chemotherapy and radiation also remain options. [16]

No enough randomized controlled trials were published to evaluate the response of the retroperitoneal sarcomas to the neoadjuvant and adjuvant chemotherapy and radiotherapy, however many clinicians briefer to use adjuvant radiotherapy in patients with positive margins. Doxorubicin is an anthracycline based chemotherapy have been used as a neoadjuvant therapy over the years, but showed no survival benefit, so the routine use of it has been discontinued. Also, the use of adjuvant chemotherapy showed no benefit in the survival and local recurrence rates. [18] As reported in a retrospective study conducted among 174 with retroperitoneal liposarcoma who received administration of perioperative radiotherapy was found to be associated with better local control. [19] As concluded in another retrospective study among 908 patients with retroperitoneal liposarcoma who underwent surgical resection with perioperative radiotherapy, intraoperative radiotherapy generated equivalent outcomes compared to external beam radiation. Patients with liposarcoma may benefit from combination of intraoperative radiotherapy with adjuvant external beam radiation. [20] Following the NCCN recommendations for radiotherapy postoperative radiotherapy should be given to patients with high-grade tumors, extremely large tumors and those with close margins in patient with R0 resection. Postoperative radiotherapy should be given if neoadjuvant therapy was not given for patient with R1 resection. [21]

Among all subtypes, WDLS had the best prognosis while DDLS and PLS had the worst. The 5-year survival rate of the WDLS reaching up to 90% while the SSLS and PLS reaching below 75%. [16] According to a previously conducted study series, R0 resection increases survival rate from 16.7% to 58% with a median survival of 103 months for R0 and 18 months for R1. [22]

In general sarcomas of the retroperitoneum are characterized by a high rate of local recurrence. [23] The retroperitoneal liposarcomas are noted to have a 3-fold higher risk of local recurrence compared with other sarcomas. [24] The overall rate of local recurrence of WDLS is 50%, while DDLS is 80% within five years of a complete surgical resection. [25] Local tumor control tend to improve with intraoperative radiotherapy while it does not affect the overall survival, this was shown in a retrospective study conducted among 25 patients with retroperitoneal sarcoma in which 11 patients received intraoperative radiotherapy. [23]

Conclusion

The retroperitoneal liposarcoma is uncommon type of malignancy. The gold standard for diagnosis remains biopsy, meanwhile imagings are widely accepted diagnostic tools. The complete resection of the tumor with negative margins considered the mean stay of treatment. Liposarcoma is usually associated with a high of recurrence rate even with grossly complete resection, that indicate a long-term and close frequent follow-up.

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Ethics approval and consent to participate

The approval of the current study has been granted by the medical committee of research ethics in King Abdulla Medical City.

Consent for publication

Written informed consent was obtained from the patient for publication of this study.