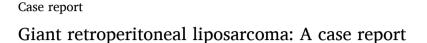
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ABSTRACT

Introduction and importance: Retroperitoneal liposarcomas are rare malignancy. They can grow usually asymptomatic until large enough to compress the surrounding organ. Giant retroperitoneal liposarcoma with diameter over 30 cm and weight over 20 kg is extremely rare. There has been limited report of giant retroperitoneal liposarcoma.

Case presentation: A 34-year old woman complained about intermittent abdominal discomfort and progressive abdominal distension for last 2 years. There was history of weight loss for last 3 months. CT scan with contrast showed giant right abdominal mass that expanded to the pelvis ($30.4 \times 28 \times 34 \times 29$ cm), oppressed surrounding organs and displaced the intestine to the left side with no visualization of normal right kidney structure. Complete resection of this retroperitoneal tumor was performed without combined resection of the surrounding organ. The biopsy of tumor showed a well differentiated liposarcoma. We diagnosed this patient with giant retroperitoneal liposarcoma. The postoperative course was uneventful and the patient was discharged on the 3th postoperative day. Last follow up, 3 months after surgical resection, patient had no complaints and there was no recurrence of this retroperitoneal liposarcoma.

Clinical discussion: Complete resection is the predominant treatment of retroperitoneal liposarcoma to avoid recurrence. Successful complete resection of retroperitoneal liposarcoma may increase the 5-year survival rate from 16.7 to 58 %. However, complete resection is a challenge, particularly in the well-differentiated subtype, for the reason that the margins are not easily distinguishable.

Conclusion: Giant retroperitoneal liposarcoma is an extremely rare tumor with high rate of recurrence, depends on some factors such as the histological type and grade, the metastasis, and also completeness of tumor resection. In this case we performed complete resection without combined resection of the surrounding organ. Furthermore, we will continue to observe our patient closely for recurrence.

1. Introduction

Soft tissue sarcomas are very rare tumor. They account for <1% of all malignant tumors in adults with an estimated incidence of 4-5/100,000/year in Europe. About 10–15 % of adult soft tissue sarcomas are located in the retroperitoneal. There are many histological subtypes of soft tissue sarcomas and liposarcoma is the most common variant and accounts for 20 % of all soft tissue sarcomas [1,2]. Liposarcoma most commonly occurs in the extremities (52 %), followed by the retroperitonum (19 %) [3].

Retroperitoneal liposarcoma is usually asymptomatic until the liposarcoma is large enough to compress the surrounding organs. It is often misdiagnosed due to its rarity and absence of symptoms [3]. The dimensions and weight of liposarcomas are variable; those over 20 kg are called 'giant liposarcomas' and are extremely rare [1]. The prognosis of retroperitoneal liposarcoma is poor if it compare to the other subtypes of retroperitoneal sarcomas. Only complete excision provides a hope of a cure. This is often difficult, especially in well differentiated subtypes because the margins are not grossly apparent thus often necessitating contiguous organ resection [4].

Oh et all reported the case report of giant retroperitoneal liposarcoma that received organ-preserving surgical removal. They reported that follow up CT revealed a locally recurrent tumor at postoperative month 16 [1]. While Lee et all reported the case series of 21 patients of retroperitoneal liposarcoma with the major histological subtype was well differentiated liposarcoma (61.9 %). Only 6 patients (28.6 %) received tumor resection without combined resection of adjacent organ. They reported that 10 patients (47.6 %) had no any

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recurrence of retroperitoneal liposarcoma. Median disease-free survival of all patients was 19 months and the overall 3 and 5 years survival rate was 87 % and 49 % respectively [4].

We report a giant retroperitoneal liposarcoma with diameter 28×32 cm and weight 21 kg. The mass oppressed surrounding organs is successfully removed with surrounding organs preserved. This case report has been reported in line with the SCARE criteria [5].

2. Case presentation

A 34-year old woman complained about intermittent abdominal discomfort and progressive abdominal distension for last 2 years. There was history of weight loss for last 3 months. Suggestively, there was no any other chronic disease in past history and there was no history of the patient's family about the similar tumor. Physical examination indicated the distention of abdomen and found a diffuse tough mass with unclear margins in the abdominal area with size 30×34 cm (Fig. 1). There was no significant finding in the laboratory testing. CT scan with contrast showed giant right abdominal mass that expanded to the pelvis ($30.4 \times 28 \times 34 \times 29$ cm), compressed surrounding organs and displaced the intestine to the left side with no visualization of normal right kidney structure (Fig. 2). Therefore, it was decided that surgery was necessary.

During surgery, it was determined that the mass came from retroperitoneal fat. The mass compressed surrounding organ but none of them was invaded. The patient received complete resection by an urologist of this retroperitoneal tumor without combined resection of the surrounding organ. The diameter of this retroperitoneal liposarcoma was 28×32 cm and the weight was 21 kg. The postoperative course was uneventful and the patient was discharged on the 3th postoperative day. A week after surgery, patient came to urology clinic and no problems were found. The mass pathologically identified as well differentiated liposarcoma. Last follow up, 3 months after surgeical resection, the patient had no complaints and there was no recurrence of retroperitoneal liposarcoma (Fig. 3).

3. Clinical discussion

Retroperitoneal liposarcoma commonly occur in patients aged 40–60 years, and men and women are equally affected, although some large retrospective series suggest a small surplus of female patients. The dimensions and weight of liposarcomas are variable; those over 20 kg are called 'giant liposarcomas' and are extremely rare [1,6]. The size of retroperitoneal liposarcoma in this patient is 28×32 cm and its weight is 21 kg. It can occur because the retroperitoneal is a deep, expandable space without many bony boundaries. So retroperitoneal space can

allow liposarcoma to grow without compressing the vital organ. When clinical symptoms do present like abdominal discomfort, weight loss and lump abdomen, it means that the retroperitoneal liposarcoma usually grow to very large in size which can oppress or invade the surrounding organs [2,6].

Liposarcoma is the most frequent histological type of retroperitoneal sarcoma, corresponding to 41 % of these tumors [6]. The retroperitoneal liposarcomas are generally neoplasms with a low or intermediate grade of malignancy. The occurrence of hematogenous metastasis is a rare finding at the time of diagnosis; the lung represents the main site of distant metastases. Liposarcomas can divide into 4 types: (1) dedifferentiated, (2) pleomorphic, (3) well differentiated, (4) myxoid/round cell. The dedifferentiated and pleomorphic type are neoplasms with high grade of malignancy accompanied by remarkable biological aggressiveness and with metastatic potential while well differentiated and myxoid/round cell fors are tumors with a low-moderate grade of malignancy [6,7]. In this patient we found that the type of retroperitoneal liposarcoma was well differentiated, it associated with a more favorable prognosis which has 5 years survival rate between 83 % - 90 % [1].

Complete resection is the predominant treatment of retroperitoneal liposarcoma. The use of neoadjuvant or adjuvant chemotherapy and/or radiotherapy is controversial given the low sensitivity of these tumors [3,8]. Jones *et al* reported myxoid liposarcoma had a statistically significant higher respond rate for chemotherapy compared to other type of liposarcoma, but they found poor respond of well differentiated liposarcoma [9]. Ballo *et al* reported no benefits in the use of radiotherapy for retroperitoneal liposarcoma [10].

The principle of the surgery is to resect the tumor and any invaded organs without any residue of the liposarcoma or membrane remaining. Successful complete resection of retroperitoneal liposarcoma may increase the 5-year survival rate from 16.7 to 58 % [3,11]. However, complete resection is a challenge, particularly in the well-differentiated subtype, for the reason that the margins are not easily distinguishable. Combined resection is occasionally required to achieve macroscopic clearance; the kidney is the most commonly removed organ, followed by the colon. Local recurrence remains the preliminary cause of mortality in retroperitoneal liposarcoma. 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. The purpose of the secondary operation is to remove the tumor, in addition to relieving the compression of vital organ. When radical surgery is not possible, palliative resection is advisable. 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 [3].



Fig. 1. Picture of patient with abdominal mass.

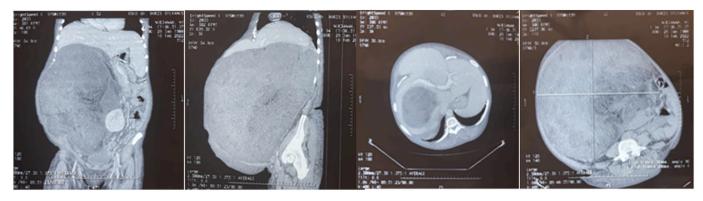


Fig. 2. Picture of patient CT scan show giant right abdominal mass that expanded to the pelvis and compressed surrounding organ.

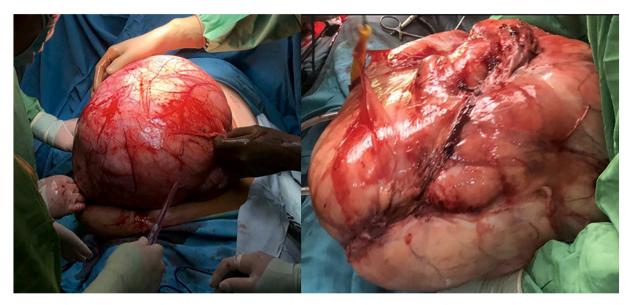


Fig. 3. Picture of retroperitoneal liposarcoma during surgery and after complete resection.

4. Conclusion

Giant retroperitoneal liposarcoma is an extremely rare tumor with high rate of recurrence depend of some factors such as the histological type and grade, the metastasis, and also completeness of tumor resection. In this case we performed complete resection without combined resection of the surrounding organ. Furthermore, we will continue to observe our patient closely for recurrence.

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None.

Ethical approval

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. The requirement for ethical approval from our institution was waived due to the nature of case reports.

Consent

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Registration of research studies

N/a.

Guarantor

Yosua Hardja.

Provenance and peer review

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CRediT authorship contribution statement

Yusuf Rachman conceived the idea, designed the study, collected the patient data, wrote the original draft and edited final manuscript.

Yosua Hardja confirmed the diagnosis, performed the surgery, evaluated and lead the case management, supervised the project, review final manuscript.

Declaration of competing interest

None.

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