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Giant retroperitoneal dedifferentiated liposarcoma

INTRODUCTION: *Liposarcoma is the most frequent type of retroperitoneal sarcomas. Dedifferentiated liposarcoma is the least common subtype and is an extremely rare tumor.*
CASE REPORT: *We present the case of a 53-year-old male who was referred with a giant retroperitoneal mass. The patients' mass was deemed unresectable by the previous institution and received chemotherapy with no benefit. We macroscopically removed the 38x32 cm mass with right nefrectomy. Pathological examination revealed dedifferentiated liposarcoma.*
CONCLUSION: *Surgery is the gold standart in the treatment of retroperitoneal sarcomas. Giant masses present a challenge for the surgeon with possible major vascular injuries and multiorgan resections. Therefore it is important for these patients to be referred for surgery without delay.*

KEY WORDS: Dedifferentiated liposarcoma, Liposarcoma, Retroperitoneal sarcoma

Introduction

Liposarcomas are neoplasms of mesodermic origin derived from adipose tissue, accounting for 10-14% of all soft tissue sarcomas. Approximately 85% of these tumors are malignant. It has a slow expansive growth and a deep localization. There is an adjacent organ compromise in upto 80% of cases[1].
This tumor is classified into well differentiated, myxoid, round cell, pleomorphic and dedifferentiated types. Dedifferentiated is the least common subtype and usually arises from a well-differentiated liposarcoma [2]. Dedifferentiated sarcoma refers to a condition in which well and poorly differentiated liposarcoma and non-lipomatous sarcoma coexist in one tumor. Thus, histological diagnosis can be difficult.
Surgery is the gold standart for treatment. It requires an aggressive approach with possible multiple resections.
We are reporting a case of giant retroperitoneal dedifferentiated liposarcoma.

Case Report

53-year-old male was referred to our clinic with a giant retroperitoneal mass. The patient applied to a rural state hospital with a palpabl abdominal mass about 6 months ago. Computerized Tomography (CT) revealed a 18 cm heterogeneous retroperitoneal mass. Biopsy was performed from the mass and it was not diagnostic. Medical team decided tumor was unresectable and pati-

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ent recieved Ifosfamide/Mesna/Adriamycin chemotheraphy. Tumor progressed under chemotherapy and later sent to our clinic for further evaluation.

Initial examination revealed a distanded abdomen and respiratory difficulty due to intrabdominal mass. Abdominal CT revealed a giant retroperitoneal mass with an approximate size of 40x30 cm. Tumor had clear margins except for a possibly compromised right kidney (Fig. 1). Surgical exploration was decided upon. Surgery was performed with the patient in supine position and a midline incision was made from xyphoid to the pubis. Right kidney was compromised as expected. Tumor was macroscopically removed with right nefrectomy. Tumor was measured as 38x32 cm (Fig. 2).

Microscopically the tumor was composed of areas of atypical lipomatous neoplasm/ well differentiated liposarcoma and fibrosarcoma like dedifferentiated component (Figs. 3, 4). We noticed a sharp demarcation between the

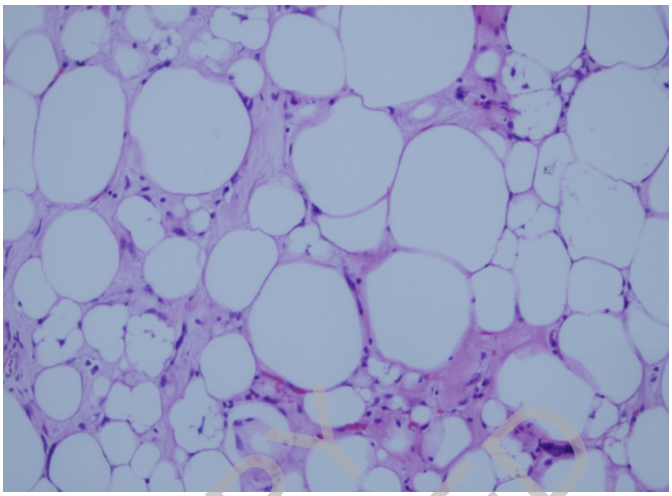

Fig. 3: Well-differentiated liposarcoma area (H&E, x400).

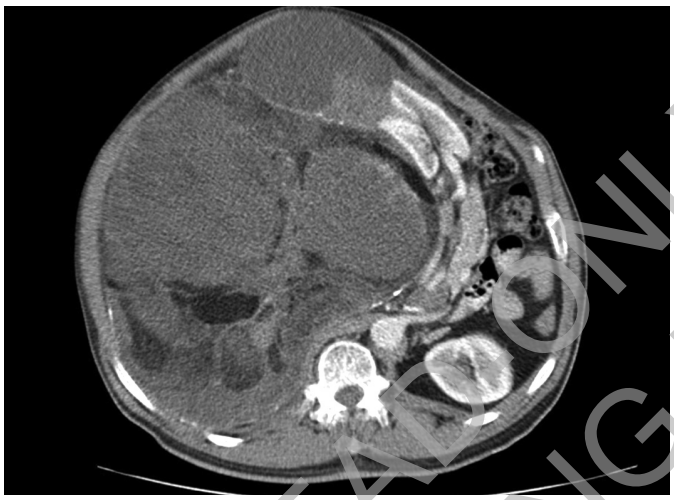

Fig. 1: Preoperative CT imaging of the mass.

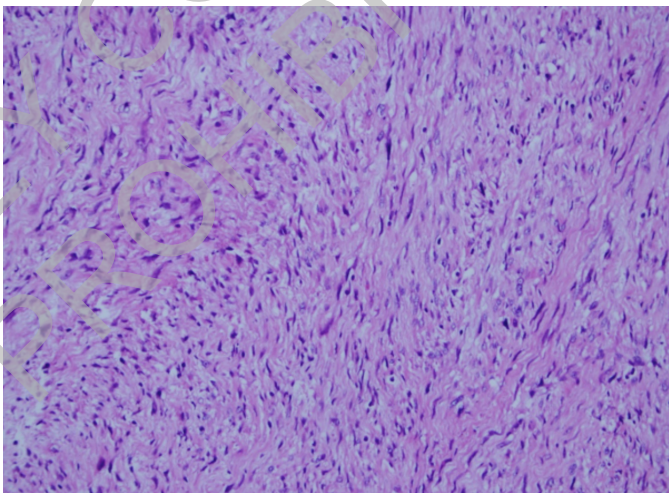

Fig. 4: Fibrosarcomatous area (H&E, x200).

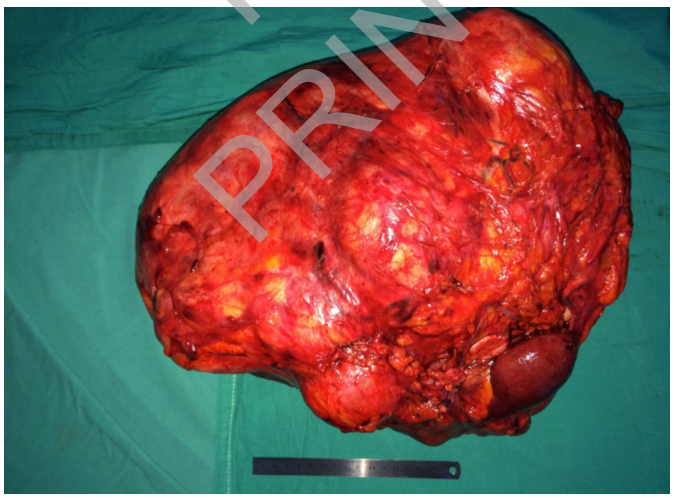

Fig. 2: Macroscopically resected tumor (Compared with a 20 cm ruler).

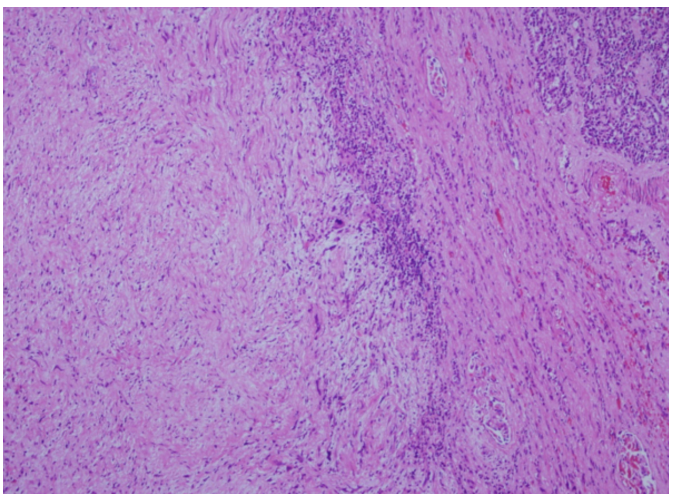

Fig. 5: The non-lipogenic zone infiltrated kidney (H&E, x100).

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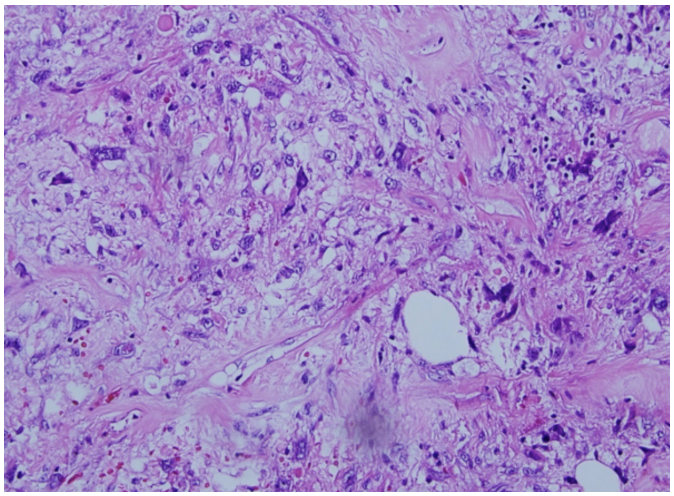

Fig. 6: Pleomorphic liposarcoma like areas (H&E, x400).

two. The dedifferentiated areas have the appearance of fibrosarcoma. The non-lipogenic zone infiltrated kidney (Fig. 5). There was also pleomorphic liposarcoma like areas (Fig. 6) including giant lipoblasts like cells with hyperchromatic nucleus and multivacuolated cytoplasm. There were many eosinophilic cytoplasmic globules.
Ultimately, patient was decided to be followed without adjuvan chemoradiotherapy. Postoperative Magnetic Resonans Imaging did not reveal any leftover tumor.

Discussion

Retroperitoneal liposarcomas are rare tumors that accounts for less than 0.2% of all malignant tumors. Dedifferentiated subtype is even more so, making up approximately 20% of all liposarcomas. It has been reported that 20% of the tumors are >10 cm at the time of diagnosis [1]. Yet few liposarcomas are large enough to be considered as giant[3-8].
The tumor was initially considered unresectable by the previous institution and chemotherapy was decided upon. Different chemotherapy regimens based on ifosfomide, mesna, doxorubicin, dacarbazine and paclitaxel have been described. However these are usually used for palliation and at the setting of recurrent disease. There is no reported survival benefit [7]. This patient did not benefit from chemotherapy, instead progressed under it.
Large tumors present a challenge for the surgeon with possible major vascular injuries and multiorgan resections. McGrath et al [8] demonstrated that complete resection can be carried out in 70% of cases and about 50% of these cases require multiorgan resection [9]. Therefore these patients should be experienced centers. This patient was diagnosed with a 18 cm tumor, however was finally operated when the tumor was 38 cm. Which was a significantly more risky operation.
Radiotherapy was not considered in this case because of the possible gastrointestinal morbidity associated with the radiation of a wide field in the abdomen. RT is usually accepted as a complementary treatment for palliation in incomplete resections and unresectable tumors[10]. Liposarcoma is regarded as more radiosensitive than the other mesodermic tumors. However, there is conflicting data on the benefit of radiotherapy on survival at the setting of macroscopically complete resections [10,11].

Conclusion

Surgery is the gold standart in the treatment of retroperitoneal sarcomas. Giant masses present a challenge for the surgeon with possible major vascular injuries and multiorgan resections. Therefore it is important for these patients to be referred for surgery without delay.

Riassunto

Il liposarcoma è il tipo più frequente di sarcoma retroperitoneale, ma la forma sdifferenziata rappresenta il più comune sottotipo e dunque è un tumore estremamente raro.
Viene presentato il caso di un uomo di 53 anni ricoverato per la presenza di una grande massa retroperitoneale, giudicata non resecabilein in altra sede e sottoposta senza effetto ad un trattamento chenioterapico. Da noi la massa è stata asportata (dimesnioni 38x32 cm) contestualmente alla nefrectomia destra. L'esame istopatologico ha dimostrato trattarsi di un liposarcoma sdifferenziato.
Si può concludere che la chirurgia rappresenta il miglior tipo di trattamento per i sarcomi retroperitoneali, anche se le masse giganti rappresentano una sfida per il chirurgo, con la possibilità di incorrere in danni vascolari maggiori e la necessità di resezioni multiorgano. Ne consegue la necessità di far ricorso alla chirurgia senza ritardi.

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