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Case Report

Giant retroperitoneal dedifferentiated liposarcoma - A rare case report

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ABSTRACT

Retroperitoneal liposarcomas are uncommon malignancy accounting for 0.15% of all malignancies. The overall survival and prognosis is predominantly based on the total completeness of resection i.e. margin status and the histopathologic grade of the tumour.
The retroperitoneal sarcomas usually present late in the disease course and often develops local invasion of the vital structures at the time of clinical presentation which makes resection with clear margins difficult and challenging. Here we present a case of Giant dedifferentiated liposarcoma arising from retroperitoneum.

1. Introduction

Retroperitoneal sarcomas are rare malignant tumour and constitute upto 0.15% of all malignancy.[1] Those liposarcomas with weight measuring more than 20 kg are termed as 'giant liposarcomas' and are extremely rare. Dedifferentiated liposarcomas contains well-differentiated liposarcomatous areas in addition with, or developing into high-grade sarcomas which might not be lipogenic in nature.[2] These tumours were initially described in 1979. Their prognosis is directly related to the completeness of tumour resection and their five-year mortality rate reaches upto 30%. The ability of this particular type of liposarcoma for local invasion and tumour recurrence has been highly described. The incidence of metastatic spread in dedifferentiated liposarcoma is found to occur up to 18% of the time,[3] which is comparatively less common than other subtypes presenting in adulthood. The retroperitoneal sarcomas usually present late in the disease course and often develops local invasion of the vital structures at the time of clinical presentation. The 5-year survival rate of dedifferentiated liposarcoma is found to be around 23% which is much less when compared to that of well-differentiated liposarcoma which is around 80%. This tumour usually presents as mass per abdomen around 80% of the time with compressive symptoms either causing pain (37%) or lower limb neurologic symptoms (42%).[4]

2. Case History

A 53-year-old male presented to our surgical oncology department with complaints of abdominal distension for 3 months. Computed tomography of the abdomen demonstrated a large Heterogeneous tumour in the retroperitoneum arising from right side encircling right ureter. Image guided biopsy revealed dark spindle cell nuclei with stroma showing areas of collagenization with necrosis and haemorrhage. IHC showed scattered nuclear positivity for MDM2 which suggested a possibility of liposarcoma. Patient was proceeded with exploratory laparotomy which revealed a bulky lesion of size 65*60*20 cm with multilobulated appearance with attachment with right kidney and ureter. Adhesion of the tumour from the duodenum and IVC released, and we proceeded

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with en bloc removal of the tumour together with right nephrectomy. Cut surface showed homogeneous grey white and yellow glistening area with intact capsule.(Figures 3, 4 and 5) The postoperative course was uneventful. Histopathology revealed a mixed cytology with few areas showing mature adipocytes admixed with lipoblast suggestive of well differentiated type and few areas showing sheets and cluster of spindle cells with bizarre nuclei and multinucleated tumour giant cell suggestive of undifferentiated pleomorphic type while few showing vesicular nuclei and prominent nucleoli suggesting round cell type.(Figures 1 and 2) Diagnosis of dedifferentiated liposarcoma was concluded. Pseudocapsule was not invaded by the tumour. No tumour invasion was seen in kidney. Postoperative events were uneventful.

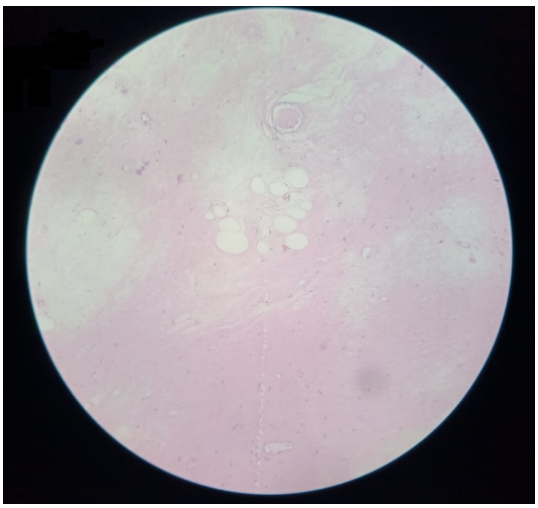

Fig. 1: Histopathological images of the specimen

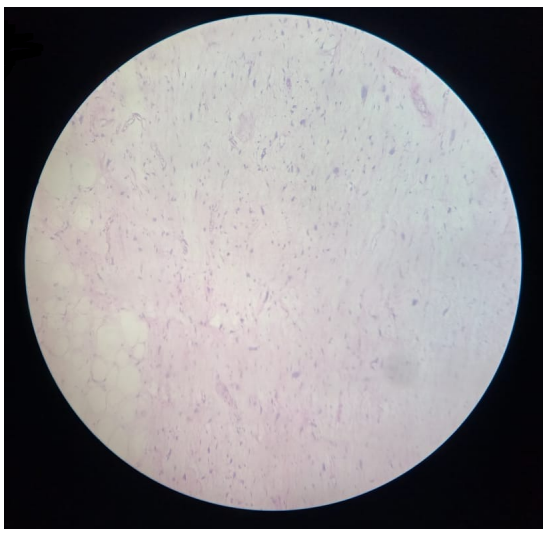

Fig. 2: Histopathological images of the specimen

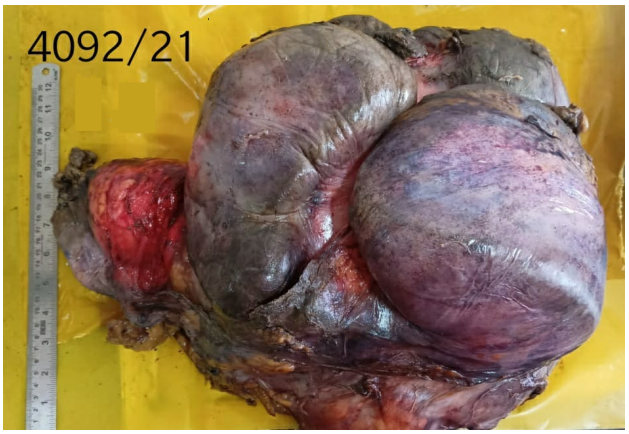

Fig. 3: Gross images of the specimen

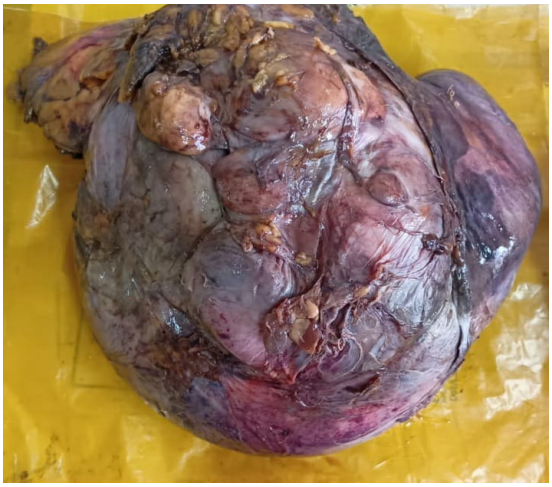

Fig. 4: Gross images of the specimen

Fig. 5: Cut surface of the specimen revealing homogenous yellowish white glistening areas

3. Discussion

Retroperitoneal liposarcomas are uncommon malignancy accounting for 0.15% of all malignancies.[5] Liposarcomas are usually classified into four types which are well-differentiated, dedifferentiated, myxoid or round cell, and pleomorphic.[6] The overall survival and prognosis is predominantly based on the total completeness of resection i.e. margin status and the histopathologic grade of the tumour (high grade versus low grade), with the best outcomes found in complete resection with negative margin of a low-grade tumour.[7] In spite of complete surgical resection with negative margin, this peculiar type of liposarcoma i.e. dedifferentiated type will have local recurrence in 80% of cases and has a four times higher risk of local recurrence when compared with other histological types of liposarcoma . The incidence of metastasis is around 18% with most common site being liver and lung. Surgery is the primary modality of treatment for retroperitoneal liposarcoma. The ability to completely resect the tumour is the most important predictor of local recurrence and overall survival.[8,9] Due to its slow indolent course and mild vague symptoms and signs, they usually present as a large tumour compressing other structures at the time of diagnosis. The primary imaging modality of choice is CT abdomen; however, in certain situations where tumour involvement of unresectable structure is visualised in CT, MRI scans might be useful in delineating further information. In some cases, angiogram may be useful. CT chest must be done in all cases as a part of workup to rule out metastatic lung involvement when approaching these patients with curative intent.

Vague abdominal discomfort and abdominal mass are the presenting complaint in 60–80% of cases with only about 6% of patients presenting without symptoms (i.e., found incidentally at laparotomy performed for other reasons). Other less common presenting symptoms are weight loss, fever, anorexia, genitourinary complaints, and bowel obstruction. The paucity of symptoms allows these malignancies to attain considerable size before presentation, making adjuvant treatment with tumoricidal doses of radiation hazardous and likely to cause significant morbidity to adjacent vital structures. In one large series, the presenting size was over 10 cm in 71% of the cases. Attempts at intraoperative radiotherapy (IORT) combined with external beam radiotherapy have demonstrated no survival benefit when compared to standard external radiotherapy alone. Chemotherapy in tumours with a significant mitotic rate also fails to show any improvement in overall survival.[10] Overall survival rates of 43–55% at 5 years are typically reported. Complete resection is possible in 60% of cases and often involves removal of adjacent organs such as kidney, ureter, and large bowel. More complex cases may involve en-block resection of gallbladder, psoas muscle, small bowel, spleen, pancreas, and major vascular structures. In selected cases of unresectable tumours, incomplete resection can increase survival and provide palliation of symptoms over biopsy alone.

4. Source of Funding

None.

5. Conflict of Interest

None.

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