CASE REPORT OPEN ACCESS

Kidney Sparing Huge Retroperitoneal Liposarcoma

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ABSTRACT

Retroperitoneal Liposarcoma is a rare malignant tumor of mesenchymal origin and is the most common type of soft tissue sarcoma with a rate of 3-4 individuals for every million people. It usually appears between 50-70 years of age, and the proportion between the sexes are equal. Due to their retroperitoneal location they are much more difficult to diagnose and often came into notice as incidental findings when they became very large or start to invade adjacent organs. The Gold Standard treatment of Retroperitoneal Liposarcoma is resection as per the European Society for Medical Oncology guideline (ESMO). Chemotherapy and radiotherapy can be offered as palliative treatment because of the extremely massive size of primary tumors rendering them inoperable as well as in high-grade cases. In this case study, a case of 40 years old male was reported, who underwent surgical resection of retroperitoneal liposarcoma. The complete resection of the mass was successful and the kidney was spared in the procedure.

Keywords: Retroperitoneal Liposarcoma; Multimodality Treatment; Chemotherapy; Radiotherapy.

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INTRODUCTION

Retroperitoneal Liposarcoma is rare with no obvious cause only a few factors like genetic alterations, exposure to radiation, or chemical substances have a profound risk of developing retroperitoneal Liposarcoma[1]. Most cases appear in patients in the 5th-7th decades of life while the male-to-female proportion is equivalent[2]. The diagnostic investigation of choice is a contrast-enhanced CT scan and MRI of the abdomen and pelvis and a multidisciplinary approach is compulsory in all cases including pathologists, radiologists, surgeons, radiation therapists, and medical oncologists[3]. The primary treatment of choice is surgical excision, whereas wide resection followed by radiation therapy is the standard treatment in high-grade deep lesions >5 cm[2,3]. Retroperitoneal liposarcoma seldom metastasizes to the Lungs and Liver through a hematogenous course[1].

CASE PRESENTATION

40 years old male inhabitant of Karachi complain of fever on and off for the last 3- 4 months for which conservative management was initiated. Various investigations were carried out however no conclusion was made. Besides fever, he had no other complaints and was doing his routine activity. On performing ultrasound an incidental mass was found

in the left lumbar region while other biochemical parameters were within normal limits.

The patient consulted a tertiary care center and after history and clinical assessments routine investigations were performed. On performing an ultrasound whole abdomen, a heterogenous yet essentially hypoechoic enormous solid nodule was seen close to the lower pole of the kidney estimating about 10.2 x 8.2 x 8.0 cm with minimal peripheral vascularity. Because of the large solid mass and its location, a CT scan whole abdomen and pelvis (with contrast) was performed which uncovered a well-defined mass estimated at 10x9.5x8 cm in the inferior aspect of the left kidney which was isodense with no evidence of calcification and hemorrhagic changes, another well-defined non-enhancing thin-walled cystic lesion measuring 7.1x6.2x2.7cm in the posterior aspect of left kidney within perinephric space was also observed in Figure 1.

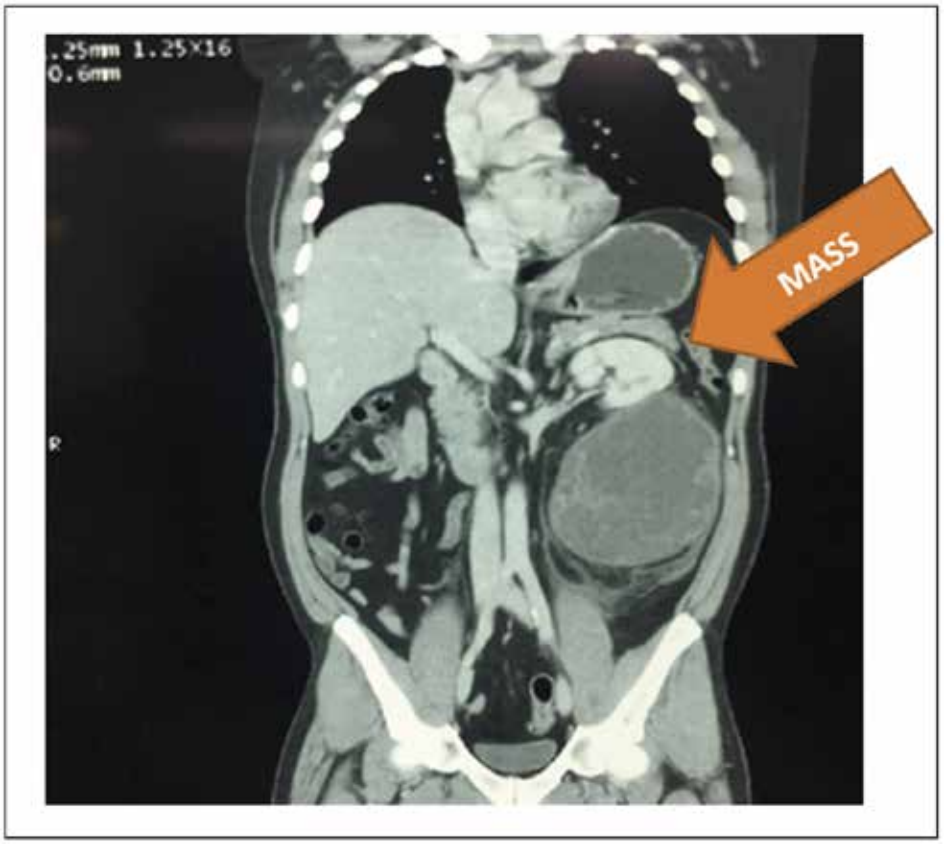

Figure 1: CT whole abdomen with contrast showing a well-defined mass measuring 10x9.5x8 cm.

On the premise of CT SCAN findings, an ultrasound-guided fine needle biopsy was likewise performed which shows the possibility of dedifferentiated liposarcoma and there was no evidence of well-differentiated liposarcoma. After a discussion of the case in a multidisciplinary tumor board meeting plan of careful extraction of retroperitoneal mass was made alongside the excision of the kidney. The flank approach was used to access the mass and during the surgical procedure, it was revealed that the mass was firmly clung to the external surface of the kidney and encompassed surrounding viscera which were isolated and the kidney was carefully spared in the procedure. The mass was then sent to a histopathology lab that confirmed dedifferentiated liposarcoma with central chondroid and osteoblastic differentiation. The size of the mass was 27.4x14.1x12.5cm shown in Figure 2. The patient had an uneventful recovery and was discharged on the fifth postoperative day. On the initial (1st follow-up) after 1 week the patient stayed liberated from pain and his general condition improved. The case was and by talking about with a medical oncologist and the thought of chemotherapy as adjuvant treatment was dropped anyway however the patient was advised to follow up after 3 months with a CT abdomen and pelvis with contrast.

After a 3-month CT abdomen and pelvis with contrast was performed this revealed a poorly circumscribed heterogeneously dense retroperitoneal mass in the left perinephric space measuring about 3 x 3 x 3cm and the left kidney was pushed

superiorly. Possibility includes small residual/ recurrent mass or post-op collection with surrounding post-op changes in the perinephric area. A few sub-centimeter retroperitoneal nodes were also noted. Based on these findings patient was referred to an oncologist and after the consultation was advised PET SCAN which manifested increased soft tissue density, and heterogeneous distribution with modest FDG avidity in the left perinephric region and may reflect post-surgical changes, although residual disease can't be entirely excluded.

Once again, the case was discussed in a multidisciplinary tumor board meeting and the plan of chemotherapy and re-exploration was dropped the patient was advised to follow up after 6 months with CT Abdomen and pelvis with contrast.

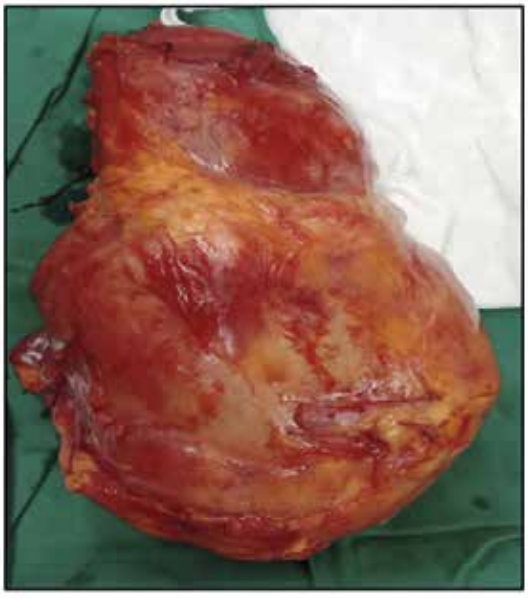

Figure 2: The excised mass was 27.4x14.1x12.5cm weighing approximately 3kg.

DISCUSSION

Retroperitoneal Liposarcoma is exceptionally uncommon and is viewed as the most widely recognized variation of soft tissue sarcoma, with a rate of 3-4 individuals for every million people. It usually appears between 50-70 years of age, and the proportion between the sexes are equal with no racial predominance[4]. They may happen at any anatomical location where fat is available, particularly within the deeper soft tissues, in the extremities, between 12-40% occur in the retroperitoneum and about 35% emerge from the perirenal fat[1].

There is no obvious reason behind Retroperitoneal liposarcoma except for some inclining elements like genetic alterations, multiple exposures to radiation or chemical substances, repeated computed tomography (CT)-scanning, and particularly high-dose ionizing radiation as used in external beam radiation therapy (EBRT) is associated with a higher risk of developing retroperitoneal liposarcoma. Exposures to certain toxic chemical agents for example arsenic, asbestos, androgenic-anabolic steroids, and dioxins are also listed in predisposing factors[1].

Intra-abdominal large space permits liposarcoma to expand without compressing the surrounding viscera, bringing about uncommon early conclusions and this is the only reason they remain silent and no symptoms appear at all. At the point when clinical manifestations do appear, the retroperitoneal liposarcoma has generally become enormous and starts invading encompassing organs[5]. In general, these tumors may present with pain in the abdomen accompanied by anorexia and an increase in abdominal girth however the most trademark sign is an abdominal mass without pain which is present in nearly 78% of the cases[2,6].

Investigations required to discriminate from other soft tissue tumors are sonography, CT scan MRI and Biopsy however diagnostic investigation of choice is a contrast-enhanced CT scan and MRI of the abdomen and pelvis. As indicated by ESMO guidelines following a fitting imaging assessment, the standard way to diagnose retroperitoneal liposarcoma incorporates the use of multiple core needle biopsies using >14-16 G needles but a biopsy can bring about tumor seeding if there is an occurrence of

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giant retroperitoneal liposarcoma[1,7].

Liposarcoma appears as a large space-occupying lesion that dislodges or compresses the surrounding organs with a negative attenuation coefficient and sometimes internal septations on CT scan while under MRI, they are reflected as a mass with a thick peripheral ring and linear septae with internal nodules and preserved contiguous structures[3]. Histologically Liposarcoma is divided into 5 subtypes: (a) Well differentiated, (b) Myxoid, (c) round cell, (d) Pleomorphic and (e) dedifferentiated[8,9].

Usually, round cell, pleomorphic and dedifferentiated subtypes are observed to be high grade whereas well-differentiated and myxoid subtypes are considered low grade with a higher overall survival rate compared to other subtypes. It should be remembered that the most significant negative prognostic factor in patients with retroperitoneal liposarcoma is the high histological grade which may diminish the overall survival rate[5]. A multidisciplinary team consisting of pathologists, radiologists, surgeons, radiation therapists, and medical oncologists is required to treat Retroperitoneal liposarcoma[3].

The Gold Standard treatment of retroperitoneal liposarcoma is complete careful excision without limitation of size. However, local recurrence is frequent and occurs in two-thirds of patients and this is usually a sign of inadequate resection whereas wide resection followed by radiation therapy is standard treatment in high-grade deep lesions >5 cm. Complete resection can be achieved in up to 70% of cases while in up to 50% of these cases multi-organ resection is essential which mostly includes the kidney (30%) [1,10].

Due to the high frequency of local recurrence after surgical excision, Adjuvant radiotherapy is routinely advised regardless of complete resection. Radiotherapy has commonly been utilized in the treatment of not completely resected liposarcoma, recurrence at the primary site, huge inoperable primary tumors, and for palliative purposes in case of metastasis. In some cases of huge liposarcoma, neo-adjuvant radiotherapy can be given which makes inoperable liposarcoma technically resectable[4].

Radiotherapy is usually advised for 4.5-6 weeks and the dose remains 5000-6000 RADS, but to convey such a high dose to the retroperitoneum is itself challenging and considered to be unsafe and difficult. Well-differentiated myxoid type of liposarcoma responds very well to radiotherapy in comparison to other histological subtypes making them more radiosensitive. Recurrence of liposarcoma makes them less radiosensitive than primary tumors[4].

Additionally, there might be some role of chemotherapy as myxoid liposarcoma was seen as the most chemo-sensitive histological sort, while the well-differentiated and dedifferentiated sort is less responsive. Multimodality treatment which includes both chemotherapy and radiotherapy is considered the best if there should arise an occurrence of high-grade liposarcoma[3]. Indeed, even with the complete evacuation of liposarcoma, the overall prognosis stays poor. The 5-year survival rate of the well-differentiated subtype is 90% dedifferentiated 75% and Myxoid/round cells at 60-90% and pleomorphic at 30-50% [5].

The prime reason for mortality in retroperitoneal liposarcoma is local recurrence. The overall survival rate is much better in the patients who got complete resection of their tumor in contrast to those who did not. Metastases from Liposarcoma may happen at any place in the body, yet are generally common in the lungs and liver. In this way, the highest quality level treatment stays to be the complete evacuation of the mass. In case of recurrence, the prime trouble of the surgery is that the anatomical relationship would be increasingly mind-boggling owing to post-surgical adhesions and this brings about expanded trouble during the surgery.[5]

Follow-up of a patient is mandatory where a CT scan is performed with an interval of 3 months for the first 2 years and dropped to every 6 months for the next 3 years followed by once a year to look for recurrence of the disease[5]. Most reported liposarcoma is less than 1 kg in weight but few authors have reported gigantic masses weighing 10 Kgs 18Kgs, 28kgs and among them highest 42 kg has also been reported[10]. The case we encountered was huge measuring 27.4x14.1x12.5cm and weighing roughly 3Kgs whereas on histopathology dedifferentiated liposarcoma with central chondroid and osteoblastic differentiation was found. The case was discussed in a multidisciplinary tumor board meeting and a unanimous decision was made to avoid further treatment and patient was advised for regular follow-up after every 6 months with CT abdomen and pelvis with contrast.

CONCLUSION

Retroperitoneal liposarcoma is an uncommon disease having a greater incidence of recurrence. Gold standard treatment remains complete resection. However, the combined resection of neighboring organs from time to time is essential.

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CONFLICT OF INTEREST

The authors declared no conflict of interest.

PATIENT CONSENT

Consent was taken from the patient before writing this case report and after the consent, the work on this case report was initiated.

AUTHORS' CONTRIBUTION

OKS was involved in the writing of this case report. MSM being the consultant supervised the whole study and helped in the final review and correction before the submission of this case report. WF helped in the data collection.

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