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Case report

Huge retroperitoneal liposarcoma encasing right kidney: A case report from Nepal

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ABSTRACT

Introduction and importance: Retroperitoneal liposarcoma is a rare entity originating from adipocytes. Before showing any symptoms, it can grow hugely and envelop nearby structures. Surgery is the optimum course of treatment, however, well-differentiated liposarcomas make it challenging to discern between malignant and normal adipocytes during surgery.

Case presentation: We report a case of a 62-year-old male referred to evaluate his abdominal distention presented for four years. Recently he had developed loin pain for six months. A computerized tomography scan showed extensive tumor encompassing the right kidney and ureter, colon, and duodenum, however, exploratory laparotomy revealed free colon and duodenum. Complete resection of the mass with a right nephrectomy was performed. Subsequently, a histopathological assessment of the resected specimen confirmed the diagnosis of well-differentiated liposarcoma. Adjuvant chemotherapy was initiated as the tumor was a high-risk sarcoma but local recurrence was observed after 2 years despite surgery and chemotherapy.

Clinical discussion: Imaging modalities are the mainstay of preoperative diagnosis. Preferably, surgical resection with a tumor-free margin is recommended to avoid tumor recurrence which remains the primary challenge. This, along with the grade of the tumor, multifocal disease, and invasion of adjacent structure dictate the prognosis of the disease. Adjuvant chemotherapy and radiotherapy are not regarded as standard therapies for resectable retroperitoneal liposarcoma, although further research is still needed to determine their value in the case of high-risk sarcoma.

Conclusion: Retroperitoneal liposarcoma has the potential to present as huge asymptomatic masses which with an added predilection for recurrence poses a huge challenge to any surgeon.

1. Introduction

Sarcoma also known as Soft Tissue Sarcoma (STS) are malignant tumors that arise from mesenchymal tissue. There are >100 different histologic subtypes out of which liposarcoma are most common which accounts for 20 % [1,2]. Liposarcoma appears to arise from adipocyte (fat cell) and is most common in extremities (52 %) and retroperitoneum (13 %) [3]. Retroperitoneal Liposarcoma is clinically silent until they are large enough to compress the adjacent structure [4]. Although biopsy is the gold standard for diagnosis, imaging is accepted as a modality for diagnosis and staging as well as the determination of preoperative unresectability [5]. Surgery is a cornerstone in treating retroperitoneal liposarcoma with or without resecting an adjacent structure [6]. Herein, we present a case of huge retroperitoneal liposarcoma encasing the entire right kidney and adherent to the adjacent structure. Written informed consent has been taken from patients for the publication of this case report. This work has been reported in line with SCARE 2020 criteria [7].

2. Case presentation

A 62-year-old male was referred to the outpatient department of General Surgery at Tribhuvan University Teaching Hospital for further evaluation of abdominal distention that had been present for four years and right loin pain that had been present for the previous six months. He was previously treated for pulmonary tuberculosis and has comorbid

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conditions like hypertension and diabetes mellitus. All of the vital signs were stable during the examination, and the system reviews were normal. The abdominal examination revealed a distended abdomen and a mass that was firm, irregular, non-tender, and did not move with respiration. It was extended from the right subcostal margin to the right iliac fossa, measuring 22 cm, and crossed the midline. Baseline laboratory investigations including the renal function tests were normal. The contrast-enhanced computed tomography (CECT) of the abdomen and pelvis demonstrated a huge 30 × 28 × 21 cm, fat attenuating heterogenous mass with septation and few nodularities on the right side of the abdominopelvic cavity. The mass was encasing and displacing the right kidney and ureter, while merely displacing ascending colon including the appendix, hepatic flexure, transverse colon, and 2nd and 3rd part of the duodenum to the left side (Fig. 1). Patient underwent trucut biopsy which revealed an atypical lipomatous tumor. Furthermore, a diethylenetriamine pentaacetate (DTPA) renogram was performed to assess renal function, revealing the bilateral kidney's normal functioning (differential function: Left kidney-51 % and Right kidney-49 %).

The exact origin of the tumor was unclear, so exploratory laparotomy was performed which revealed an extensive tumor extending superiorly up to the liver and inferiorly up to the pelvis (Fig. 2). Furthermore, the tumor entirely encased the right kidney and ureter, but was only juxtaposed to the duodenum, colon, and inferior vena cava (IVC).During surgery, the tumor was freed from the intestine and IVC, however, it could not be separated from the kidney. As a result, the tumor was removed, followed by a right nephrectomy and DJ stenting of the left ureter.

On gross examination, the tumor measured 30 × 20 cm including the right kidney, and weighed approximately 10 kg (Fig. 3). On histopathological examination of the resected specimen, atypical lipoblast cells were found which was consistent with well-differentiated liposarcoma, which infiltrated only up to perinephric fat (Fig. 4).

His postoperative stay was uneventful and was discharged on his ninth day of hospital stay with a scheduled follow-up six monthly for initial two years. He received 4 cycles of adjuvant treatment (in each cycle: intravenous (i.v) Adriamycin 30 mg from day 1–3 and i.v Ifosfamide 2 g + Mesna 200 mg from day 1–4) which was associated with systemic toxicity. However, he was inconsistent in his follow-up, and a local recurrence was discovered two years later. Follow-up CECT abdomen and pelvis revealed a 7.3 × 7.1 × 6.8 cm enhancing tumor in the right abdominopelvic cavity displacing and abutting the adjacent bowel loop and urinary bladder (Fig. 5). For the local recurrence, a second surgery was performed which involved removing of the tumor, leaving bowel and urinary bladder intact. The patient was discharged

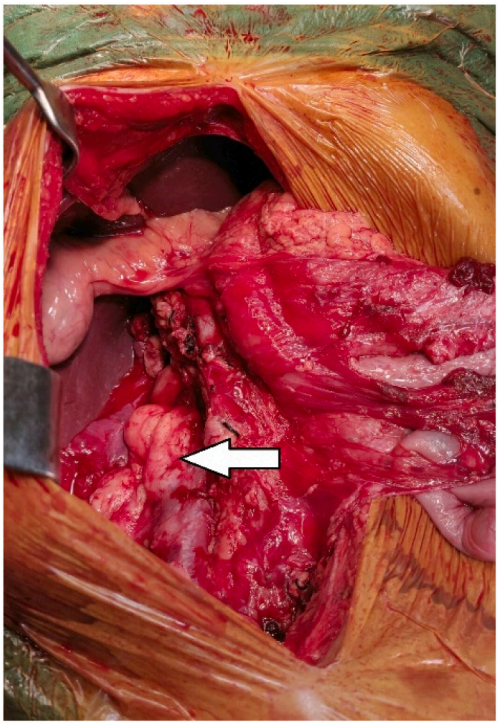

Fig. 2. Intra-operative view showing tumor in -situ (white arrow).

after 7 days of hospital stay but refused chemotherapy because of systemic toxicity in the previous regime, however, he has been doing well till now.

3. Discussion

Overall retroperitoneal tumor represents 1 % of all malignancies and liposarcoma is the commonest to occur in retroperitoneum (41 %) [2]. Retroperitoneal liposarcoma is usually present in the mid-fifties but can occur at a wide range of ages [8]. According to WHO, it has four histological subtypes: 1. Well-differentiated 2. Dedifferentiated, 3. Myxoid/Round cell, and 4. Pleomorphic. A well-differentiated subtype is the commonest sarcoma arising in the retroperitoneum [9].

More than its rareness, its presentation makes the task of the physician more difficult. Either they remain asymptomatic (80 %) or

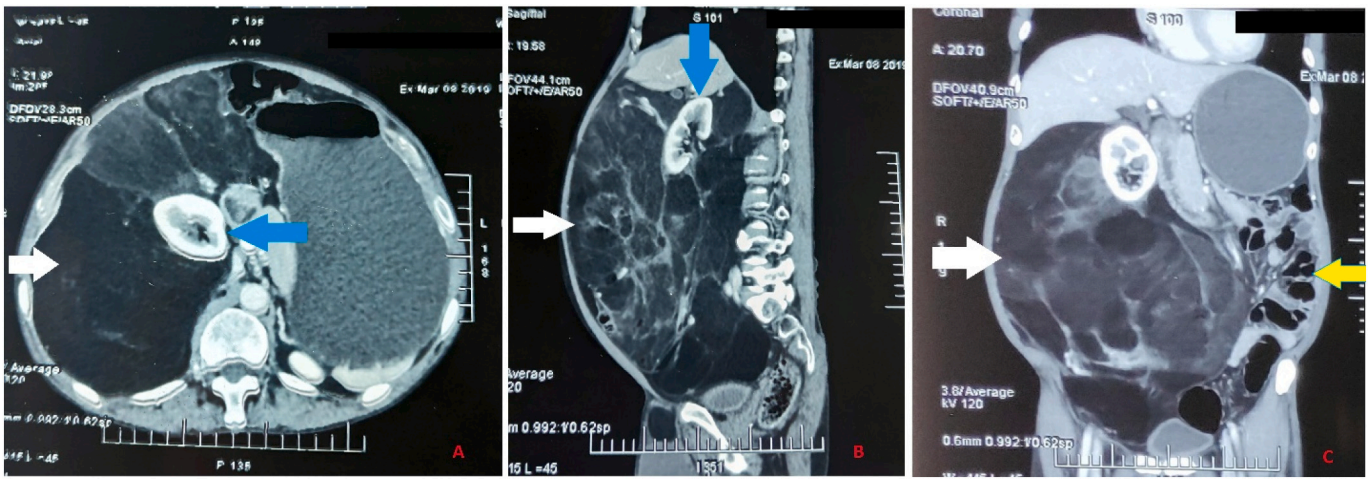

Fig. 1. CECT showing huge retroperitoneal liposarcoma (white arrow) with high-fat attenuation and septation. The tumor is encasing the right kidney (blue arrow) and displaces the viscera (yellow arrow). (A-axial, B-sagittal, C-coronal view). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

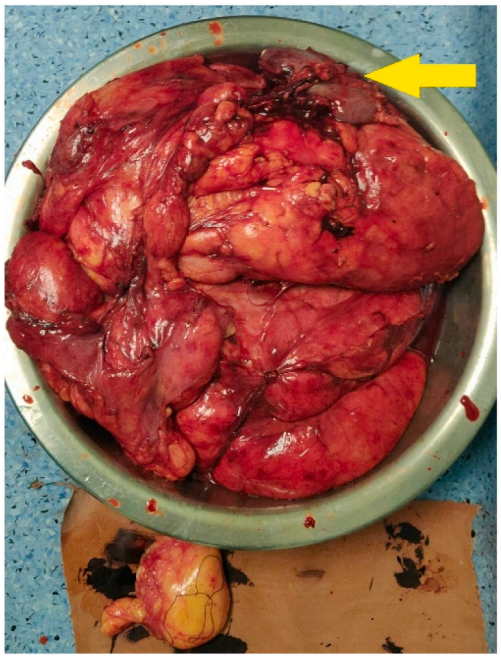

Fig. 3. Gross picture showing huge tumor including kidney (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

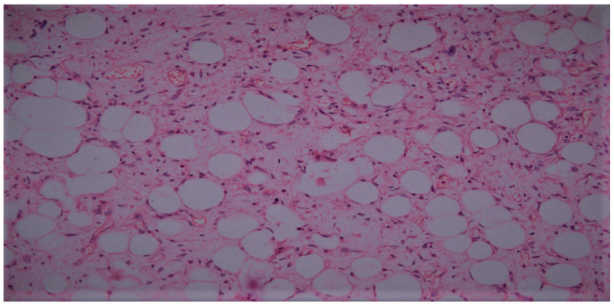

Fig. 4. Hematoxylin and eosin stain (magnification ×100) showing atypical adipocytes with eccentric nuclei.

present with non-specific symptoms such as vague abdominal pain and abdominal mass. In addition, neurological, musculoskeletal, and urinary/bowel symptoms are also common, resulting from compression of nearby structures, because most retroperitoneal sarcomas reach >10 cm at diagnosis [8].

Imaging modalities are the appropriate mainstays of presurgical diagnosis [4,5]. Uniquely, in cases of liposarcoma imaging may even help differentiate the cell types. CECT scans are the most commonly used modality but MRI is debated as an equally efficacious technique but lacks large-scale comparisons [2]. Despite being the gold standard for diagnosis, biopsy is not routinely recommended. Only those patients with surgically difficult tumors, hematogenous spread, and those being considered for preoperative radiotherapy/chemotherapy are recommended a percutaneous core needle biopsy, which should not be delayed due to the risk of tumor seedling [8].

Surgical resection remains the only potentially curative therapy for retroperitoneal liposarcoma. Most clinicians suggest R0 resection (complete resection with microscopically negative margin), although R1 resection (microscopically positive margin) is also accepted at the cost of a high recurrence rate. As compared to extremity STS, R0 resection is very difficult to achieve in retroperitoneal liposarcoma due to anatomic constraints [6]. Furthermore, it is a real challenge in the case of well-differentiated liposarcoma because of the difficulty in distinguishing atypical adipocytes from normal. Nevertheless, complete R0 resection should be the goal during surgery and to achieve this sometimes en block resection along with adjacent structures is needed, of which kidney being the commonest [6]. Because the tumor was widespread and could not be freed from the kidney, a resection of the right kidney was required to accomplish R0 resection in our case.

The multidisciplinary treatment approach is an emerging concept in the treatment of STS including retroperitoneal liposarcoma, which combines surgical resection with radiotherapy (RT) and chemotherapy [10]. Preoperative RT and chemotherapy can be considered in cases when total resection is not possible or there is high-grade sarcoma, however, preoperative radiation is not currently advised for resectable retroperitoneal liposarcoma. A recent randomized trial (EORTC-62092, STRASS) failed to demonstrate the advantage of recurrence-free survival of radiotherapy plus surgery over surgery alone [11]. Regarding chemotherapy, neoadjuvant and adjuvant are the two different approaches available. The benefit of the neoadjuvant approach is still insufficient and a new study has been started (STRASS 2) to explore this gap [12]. In the case of adjuvant chemotherapy, it is not recommended as standard therapy for resectable retroperitoneal liposarcoma [13]. However, for high-risk sarcoma (deep location, size >5 cm, high-grade

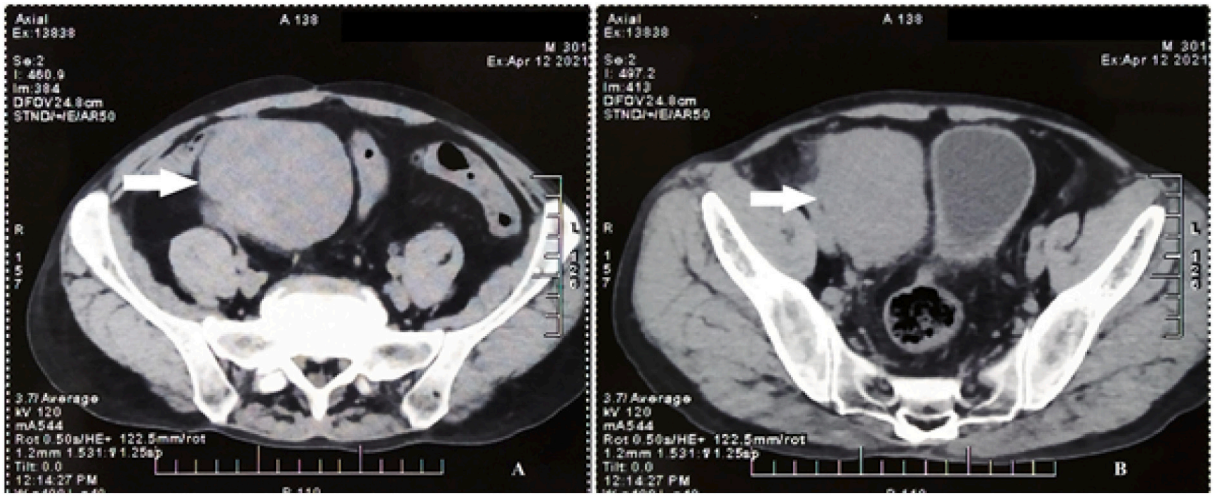

Fig. 5. CECT of abdomen and pelvis showing heterogeneously enhancing soft tissue lesion (white arrow) in the right side of abdominopelvic cavity abutting and displacing adjacent bowel loops and urinary bladder.

sarcoma) which has high chances of recurrence, anthracycline-based adjuvant chemotherapy can still be considered a reasonable option. The systemic meta-analysis done by Pervaiz et al. (2008) showed that the adjuvant approach can reduce local and distant recurrence. In addition, survival was significantly improved with the addition of ifosfamide to doxorubicin [17]. Our patient also underwent anthracycline-based adjuvant chemotherapy because the tumor satisfied the criteria of high risk (size 30 × 20 cm) sarcoma.

In terms of survival and recurrence (local/distant), the major prognostic factors include tumor-free margin, histologic grade, presence of multifocal disease, and invasion of an adjacent structure [8,14]. Our case had well-differentiated liposarcoma, which has a good prognostic value and has a 5-year survival of about 90 %. However, because the tumor was huge and adhered to an adjacent structure, a local recurrence was observed after 2 years despite surgery and 4 cycles of chemotherapy. Despite being considered a high risk when larger than 5 cm, size as such is not a prognostic factor for retroperitoneal liposarcoma as compared to truncal liposarcoma when discussed separately [15]. Even though we considered size when devising our treatment plan, it demands more light on an exploration of better prognostic evidence for retroperitoneal liposarcoma. In fact, all the prognostic factors should be considered while deciding the treatment plan and for convenience Zhuang et al. developed a nomogram that can predict 1-, 2-, and 5-year recurrence-free survival in Asian patients with retroperitoneal liposarcoma after surgery with a concordance index of 0.695 [16].

4. Conclusion

Given the size of the retroperitoneum, retroperitoneal sarcoma has the potential to enlarge before becoming apparent and symptomatic. Surgery is the mainstay of treatment and has high survival benefits if carried out in a specialized center with an interdisciplinary team because many times complete resection requires removal of the adjacent structure as well. Due to its notoriously recurring nature, the treatment should be shifted to a multimodal treatment approach and require lifelong vigilant surveillance.

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Ethical approval

None.

Consent

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.

CRediT authorship contribution statement

AS and BT collected data, prepared the original manuscript, reviewed, and edited the manuscript. AR, RSB, and NK reviewed and edited the manuscript. AS, BT, AR, NK, and RSB reviewed and edited the manuscript and were in charge of the case.

Registration of research studies

Not applicable.

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Guarantor

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Declaration of competing interest

All the authors declare that they have no conflict of interest.

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