Case Report

Liposarcoma excision and removal with right total nephrectomy in a large retroperitoneal perinephric liposarcoma involving the right kidney: A case report

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ABSTRACT

An uncommon tumor, retroperitoneal liposarcoma (LPS) occurs in 2.5 out of every million people. Due to the lack of distinct clinical manifestations, early diagnosis is challenging. LPS is a mesenchymal tumor that is malignant and mostly develops into adipose tissue and is a common soft tissue tumor that affects people in their middle years. The thigh, gluteal region, retroperitoneum, leg, and shoulder area are among the areas that are frequently affected. Here, we report the case of a 72-year-old female who had a history of abdominal distension from the past 6 months, occasional retention of urine from the past 3–4 months, and constipation on and off from the past 1 year. The patient was evaluated and diagnosed with a case of right retroperitoneal LPS and treated by surgical resection. To sum up, retroperitoneal LPS is an uncommon condition involving other organs, though occasionally combined resection of other organs is also required.

Key words: Kidney, Perinephric fat, Retroperitoneal liposarcoma, Surgical resection of retroperitoneal liposarcoma

Retroperitoneal liposarcoma (LPS) is a rare type of cancer that occurs in the fatty tissue behind the abdomen. LPS is a commonly reported tumor produced from adipocytic differentiated primitive mesenchymal cells, making up only around 10% of all soft tissue sarcomas and 15% of all sarcomas [1]. Its incidence peaks in the age range of 50–60 years [1]. LPS has been documented in the abdomen in regions such stomach, descending mesocolon, and esophagus, despite the fact that it mostly affects the deep soft tissues of the extremities [1]. With an estimated incidence rate of 0.5/100,000 of the population, retroperitoneal LPS accounts for 12–40% of all LPSs [2]. In some cases, it can affect the kidney and perinephric fat. The majority of tumors are palpable masses; therefore, the clinical presentation varies depending on the tumor's location and compressing the surrounding organs [3-8]. Large space of the retroperitoneum allows for the growth of retroperitoneal LPS. Any area with fat can develop LPSs; about 30% of the tumors start in the retroperitoneal cavity [9], and 35% come from the perirenal fat. Clinically, LPS presents as a painless, gradually growing mass; hence, when identified, over half of the retroperitoneal LPSs have a diameter >20 cm [10]. Five separate histological subtypes of soft-tissue LPSs have been identified by the World Health Organization: Well-differentiated, dedifferentiated, myxoid, pleomorphic, and mixed type. Depending on their histological subtype and grade, retroperitoneal sarcomas can have different clinical courses [11,12]. Complete surgical excision is the preferred course of treatment. According to Stoeckle *et al*., adjuvant radiation therapy for a resected well-differentiated retroperitoneal LPS does not now improve survival [13].

Here, we report the case of retroperitoneal LPS found on the right side of the abdomen originating from the right kidney of a 72-year-old female, which is a rare location.

CASE REPORT

A 72-year-old female presented with a history of abdominal distension from the past 6-month complaints of occasional retention of urine for the past 3–4 months and constipation on and off from the past 1 year. The patient had a history of frequent regurgitation of gastric contents post meals and abdominal pain for the past 2–3 months. A comprehensive medical history was obtained from the individual. There was no record of identified co-morbidities or other distinct symptoms associated with the hepatic or gynecological conditions.

Upon examination, the vitals of the patient are as follows: Blood pressure – 140/80 mmHg, pulse – 78 bpm, SpO_2 – 98%,

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temp – 98.6 F, and respiratory rate – 18/min. A thorough systemic examination of the patient was carried out, which suggested acromegaly with clubbing of nails, and prominent features of anemia. Upon local examination, the abdomen was found distended from the xiphisternum to the pubic symphysis vertically and laterally midway between the midaxillary and midclavicular line. The swelling was smooth in consistency with well-defined upper margins and deep margins were not palpable, it was mobile to some extent on bimanual pushing, not on simple palpation, the fluid thrill was not found, and the tympanic note was replaced by the dullness of whole abdomen, bowel sounds present 4–5/min. Cough impulse was absent and on leg rising, test swelling disappeared suggesting intra-abdominal origin.

Ultrasound whole abdomen and pelvis of the patient was advised which suggested retroperitoneal mass lesions. For further evaluation, a computed tomography (CT) was done which suggested a large encapsulated predominantly fat-density mass lesion involving the right side of the retroperitoneum also involving the peritoneal cavity displacing the right kidney inferomedially and bowl loops toward the left side of the abdomen. Lesion measures 22 × 20 × 27 cm (TR × AP × CC). The lesion also displaces the inferior vena cava toward the left. The right adrenal is not separately delineated. Diagnosed as a huge retroperitoneal LPS (Fig. 1a-c).

The condition was explained to the patient, high-risk consent was taken, packed red blood cells were arranged for transfusion, pre-esthetic evaluation was done, physician fitness and cardiac fitness were taken, and the patient was prepared for surgical resection of the lesion. The complete resection of the LPS was advised under high spinal anesthesia with sedation and was executed. Under high spinal anesthesia with sedation, the patient was laid down in a supine position with the abdomen uncovered from the xiphisternum till mid-thighs, and parts painted and draped and, a vertical midline incision from the pubic symphysis to the xiphisternum was created and deepened to open all layers of the abdomen (Fig. 2a-c). After retracting the abdominal muscles, fascia, and peritoneum, a big sarcoma was visibly displacing coils of intestines and stomach to the left side, adhesions were lysed and separated gently with bipolar cautery securing other organs. The duodenum, right ascending and transverse colon, and left kidney were adhered with the swelling which were separated with caution and secured. This LPS was seen originating as perinephric fat from the right kidney, arising as such a huge size, that it engulfed the right kidney and adrenals in it thus altering their morphology, finally right total nephrectomy was done by ligating the right ureter and right renal vessels first to prevent any future recurrences. The inferior vena cava was also adhered to the mass and separated gently. After complete adhesiolysis, the LPS was taken out of the abdomen, and the whole abdomen was explored for any secondary swelling and bleeding. Complete hemostasis was achieved an intra-abdominal drain was placed, and the abdomen was closed in layers (Fig. 3).

The weight of the retroperitoneal LPS was measured and was found 8456 g approximately and was sent for histopathological examination (Fig. 4a). Histological examination confirmed the diagnosis of pleomorphic retroperitoneal sarcoma (Fig. 4b). Following the procedure, she recovered without any problems or recurrence.

DISCUSSION

Retroperitoneal tumors are a highly diverse category of neoplasms with 85% of them being malignant. Of retroperitoneal masses, LPSs make up 45–55% [14]. In the age range of 54–65 years, male-to-female ratio is equal [11]. Eighty percent of the time, retroperitoneal sarcomas appear as an asymptomatic abdominal tumor. In addition, symptoms linked to local invasion or mass effect may include discomfort, gastrointestinal blockage, early satiety, and weight loss. Moreover, the lower extremities are referred for neurologic and muscular-skeletal disorders [15].

Five kinds of LPS are commonly recognized: Pleomorphic LPS, myxoid LPS, dedifferentiated LPS (DLPS), well-differentiated LPS (WDLPS), and mixed LPS. WDLPS make up 40–45% of all retroperitoneal LPS, making it the most prevalent kind [2,16]. WDLPS and DLPS frequently exhibit gene amplification in the 12q12–21 and 10p11–14 chromosomal regions; DLPS is also linked to 6q23 and 1p32 [17,18]. Furthermore, a 10% chance exists for WDLPS to change into DLPS, a more invasive LPS subtype [17]. Primary retroperitoneal LPS usually originates in the perirenal fat [1]. When it comes to pre-surgical imaging diagnosis, CT and magnetic resonance imaging (MRI) are thought to be the most suitable modalities. CT and MRI can be used to discriminate between different kinds. More than 75% of WDLPS often consists of adipose tissue, with tiny interior nodules, and

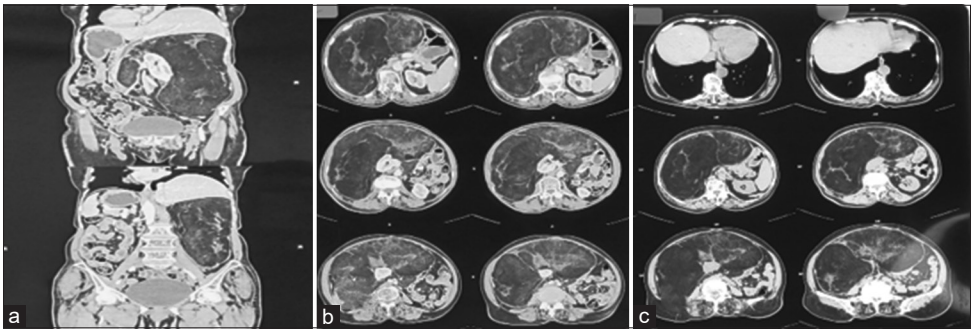

Figure 1: Coronal and axial computed tomography films signifying large encapsulated right liposarcoma involving the left kidney, displacing coils of the intestine, and other organs to the left side of the abdomen

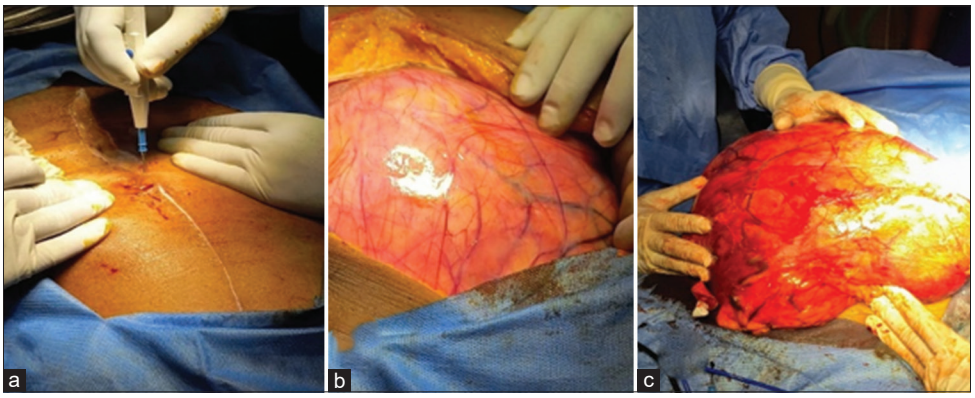

Figure 2: Displays the steps of the procedure, which include making a midline skin incision and separating the rectus to visualize the abdomen

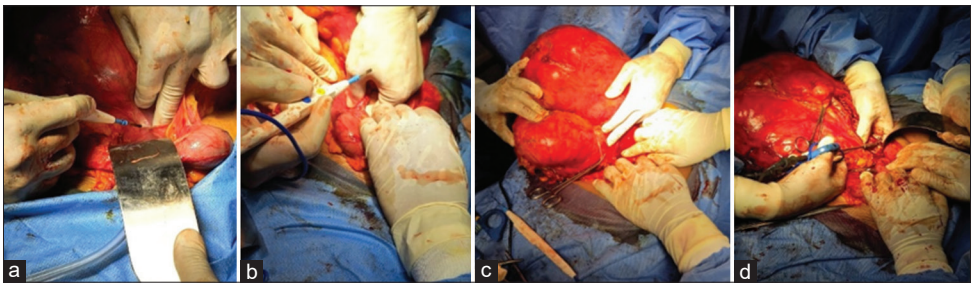

Figure 3: (a and b) show separating the transverse colon from the adhesions of liposarcoma; (c) shows identifying ureter and renal vessels; and (d) shows ligation of the right renal vessels and ureter, and subsequently right nephrectomy

Figure 4: (a) The post-operative specimen with (b) histopathology report

septations thicker than 2 mm. Soft-tissue attenuation on CT can be used to identify these nodular regions. On T2–W1 MRI, septations and nodular regions in WDLPS have a hyperintense nature, setting this subtype apart from the other LPS types [1].

The best prognosis is for WDLPS, which accounts for about 30% of cases. Approximately 50% of all LPSs are of the myxoid form, which is the most common type. It progresses less favorably since it frequently returns at an early age. The prognosis is poorest for pleomorphic, round cells, and undifferentiated kinds [15]. For patients with resectable, localized retroperitoneal sarcomas, surgery with negative margins both grossly and microscopically is the best course of treatment. *En bloc* excision of surrounding viscera is frequently necessary for complete surgical resection. The most common organ to be removed (36%) was the kidney, which was followed by segmental resections of the pancreas, spleen, and large intestine [19,20]. The mainstay of treatment for LPS remains surgery, even if the extent of resection necessary is still up for debate. Regardless of pathogenic subtypes, a macroscopically negative margin has historically been sufficient for treatment [21]. For the purpose of predicting the prognosis, including local recurrence, distant recurrence, and death, the histological subtype is crucial. According to earlier cohort studies, WDLPS has the lowest chance of local and distant recurrence, while DLPS has the highest risk [22]. This case as per histopathological reports suggested WDLPS. Apart from the extended surgery method, other strategies to improve the prognosis for lung cancer patients include chemotherapy and radiotherapy. While neighboring organs are also radiosensitive, retroperitoneal LPS is not. The time and kind of radiation utilized for retroperitoneal LPS are important since an excess of radiation can harm nearby radiosensitive organs including the liver and kidney [2].

CONCLUSION

The body of research highlights that the treatment of retroperitoneal sarcomas involves total tumor excision, both

grossly and microscopically, to minimize local recurrence, which does not impact overall survival. This is paired with surveillance to identify metastases or recurrence early on. Follow-up care for the patient should include routine physical examinations and imaging tests, such as CT scans and chest X-rays. Our patient is doing well and is still going to follow-up for surveillance.

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