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A Case of Giant Dedifferentiated Retroperitoneal Liposarcoma in a 49 year-old Filipina

Jude Immanuel P. Mascariñas, MD; Vanessa Maris C. Cariño, MD; Francisco F. Firmalo III, MD, FPCS, FPUA and Glenn P. Villanueva, MD, FPCS, FPSGS
Department of Surgery, Ospital ng Maynila Medical Center, Manila, Philippines

Liposarcoma is a malignant mesenchymal neoplasm composed of adipose tissue with varying degrees of atypia. While generally indolent, some tumors have the potential to grow enormously particularly if located in the retroperitoneum. We report a 49-year-old female generally asymptomatic except for a 1-year history of gradual abdominal enlargement. Contrast-Enhanced Computed Tomography (CT) of the abdomen showed a large retroperitoneal mass with characteristic features consistent with liposarcoma. On laparotomy, the mass was encapsulated with good plane of dissection, great vessels were preserved, however the right kidney and suprarenal gland were undetachable from the primary tumor hence was removed *en bloc* with the liposarcoma. The post-operative course was uneventful with excellent outcome after 6 months of follow-up. Final histopathologic diagnosis revealed low-grade, dedifferentiated liposarcoma, which has favorable prognosis following radical surgery.

Key words: Liposarcoma, sarcoma, renal veins, laparotomy, nephrectomy

Introduction

Soft-tissue sarcomas represent less than 1% of all malignancies, and only 10% to 15% of these tumors are located in the retroperitoneum[1,2]. Liposarcoma is the most common soft tissue sarcoma in adults, with peak incidence occurring between the ages of 40 and 60 years[1]. Retroperitoneal liposarcomas usually grow indolently and frequently may reach a very large size.

Dedifferentiated liposarcoma represents 10% of liposarcomas and accounts for the vast majority of retroperitoneal malignant spindle cell neoplasms[3].

Dedifferentiated Liposarcoma (DDL) is characterized as a typically non-lipogenic sarcoma compared to Well-Differentiated Liposarcoma/Atypical Lipomatous Tumor (WDL/ALT). Dedifferentiation occurs in up to 10% of WDL/ALT cases. The incidence of DDL is less than 0.1 per 1,000,000 each year[2].

This case is being reported because of the limited data available for local experience with Giant Dedifferentiated Retroperitoneal Liposarcomas.

The Case

A 49-year-old female with no previous comorbidities presented with a 1-year history of a gradually enlarging abdomen and a palpable right hemi-abdominal mass. She had occasional non-radiating low back pain, pain scale 6-7/10. There were no complaints of early satiety nor changes in bowel movement. There was no lower extremity swelling, and no documented weight loss when she came to this institution.

On physical exam, she had a globular abdomen with normoactive bowel sounds. There was a palpable 12 cm x 15 cm well-circumscribed, movable, non-tender mass in the right hemiabdomen. Contrast enhanced CT scan revealed a large, mildly lobulated, well-circumscribed, heterogeneously enhancing mass of mixed attenuation (mostly cystic with solid and fat components) centered in the right suprarenal and posterior para renal regions (Figure 1). Its aggregate measurements were about 13.9cm x 14.9cm x 21.9cm (AP x W x CC).

An exploratory laparotomy was performed through a midline incision. Intraoperatively, there was an encapsulated retroperitoneal mass. A Cattell-Braasch maneuver was performed to achieve complete access of the mass which had a good plane of dissection, allowing for preservation of the great vessels. However, the right kidney and right suprarenal gland were undetachable from the primary tumor. These structures were removed *en bloc*

with the mass without any intra-operative complications (Figure 2). The gross specimen measured 35 cm x 30 cm x 18 cm and weighed 22 kg. It was well-encapsulated, multilobulated, with soft to hard areas.

The histopathology report revealed: Dedifferentiated Liposarcoma with Neural Features. It was Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) Grade 2, 35.0 cm in widest dimension. Lymphovascular space invasion is not observed. Mitotic count: 0-9 Mitosis/10 High power fields. No necrosis seen. positive for tumor involvement: Hilar fat, Right kidney. AJCC Pathologic Staging: PT4NxMx (Figure 3). Post-operative diagnosis was Dedifferentiated Retroperitoneal Liposarcoma, Right Stage IIIB (T4NxMx).

The patient was discharged on the third post-operative day without any immediate post-operative complications. Preoperative and post-operative serum creatinine remained normal. The patient was asymptomatic at follow-up 6 months post-operatively. There was no recurrence of the mass appreciated on physical exam. She was advised to undergo repeat imaging for complete assessment but was lost to follow-up.

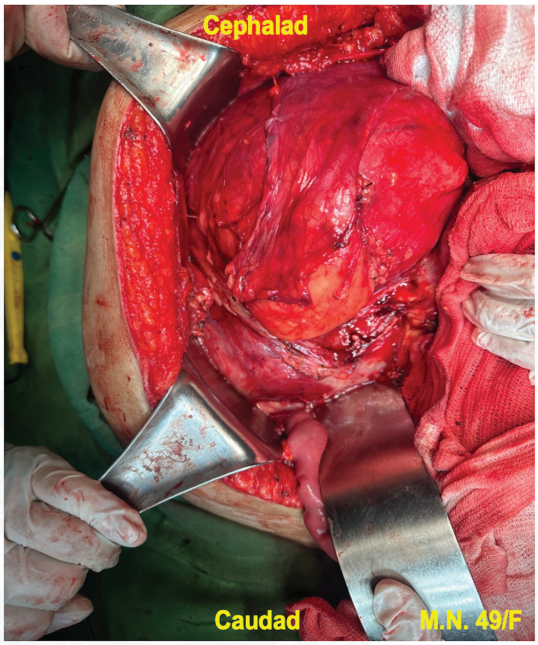

Figure 2. Retroperitoneal mass with fat and soft tissue component occupying almost the entire abdominal cavity enveloping the right kidney; Ospital ng Maynila Medical Center, 2025.

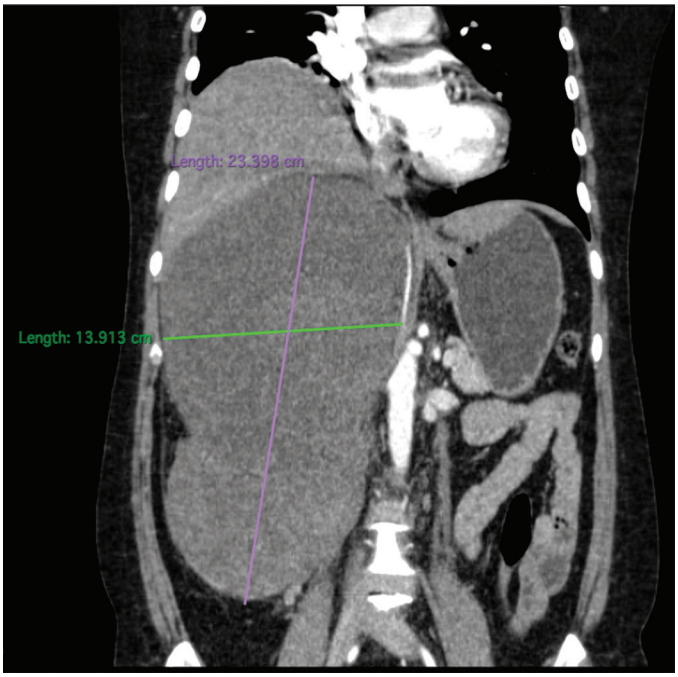

Figure 1. Retroperitoneal mass measuring 23.3 cm x 13.9 cm on contrast-enhanced whole abdominal CT scan; Ospital ng Maynila Medical Center, 2025.

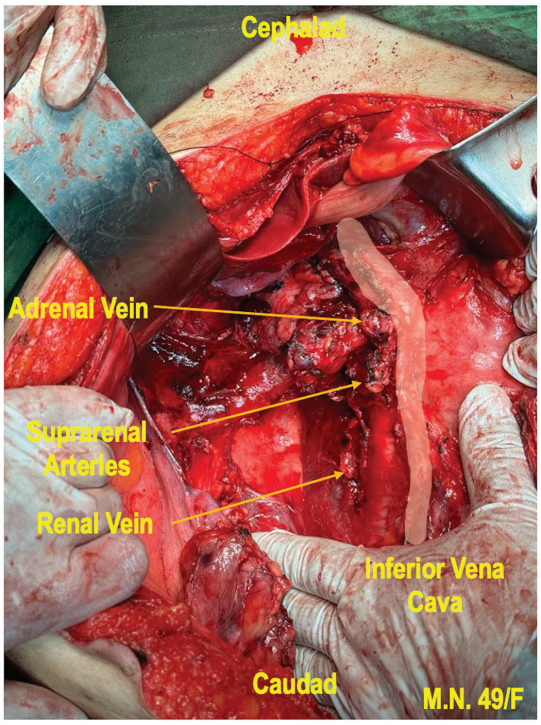

Figure 3. Retroperitoneum and great vessels identified after specimen removal; Ospital ng Maynila Medical Center, 2025.

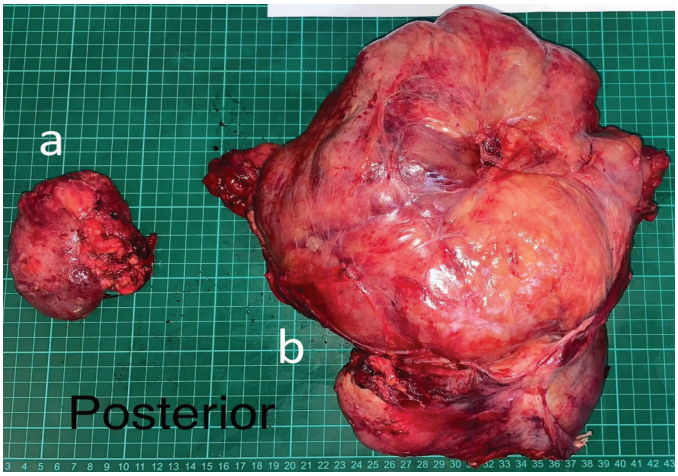

Figure 4. Gross specimens. A. Anterior view, B. Posterior view. Legend: (a) Right Kidney, 8 cm x 10 cm x 5 cm; (b) Right Retroperitoneal Mass, 28 cm x 29 cm x 20 cm; Ospital ng Maynila Medical Center, 2025.

Discussion

Dedifferentiated retroperitoneal liposarcoma (DDLPS) is a rare soft tissue sarcoma, comprising about 20–30% of all liposarcomas and 1 per 100,000 population annually[1]. Among retroperitoneal sarcomas (RPS), DDLPS represents about 40–45%, with a median age of diagnosis around 55 years, slightly more common in males[3,4]. The dedifferentiated variant typically arises from a preexisting well-differentiated liposarcoma (WDLPS). According to Makni et al.[5], giant retroperitoneal liposarcomas are tumors with a diameter of 30 cm or more, or tumor weight of 20 kg or more.

The gold standard for evaluating retroperitoneal liposarcomas is a contrast-enhanced CT scan. Liposarcomas present as large heterogeneous mass with fatty and non-fatty components[6]. The presence of calcifications and thick septa suggests dedifferentiation (Sn 83-90%, Sp 67-80%). In patients with contraindications to a CT scan, with fat-suppressed MRI sequences are also a useful diagnostic tool (Sn 74%, Sp 88%) revealing non-lipomatous solid areas with heterogeneous enhancement[7]. While there are no specific serological biomarkers for (DDLPS), molecular studies such as amplification of MDM2 and CDK4 are diagnostic of the disease[8,9].

Complete surgical resection with (R0) negative margins remains the cornerstone of treatment[2]. Given the infiltrative nature of DDLPS, en bloc resection of nearby organs (kidney, colon, pancreas, or spleen) may be required. Common post-operative complications include surgical site infection, post-operative ileus, renal dysfunction, and intra-abdominal hemorrhage or abscess formation.

The 5-year overall survival reportedly ranges from 50-60%[9], with a median disease-free survival of 24-36 month[10]. Local recurrence rate has been reported at 40-60% within 5 years post-operatively. The lungs and liver are the most common sites of distant metastasis occurring in 10-20% of cases[11]. In addition, lower age, primary surgery, absence of synchronous metastasis, absence of local relapse and treatment with experimental therapy as statistically significant favorable prognostic factors[12].

The role of adjuvant chemotherapy or radiotherapy remains debated, especially for low-grade DDLPS. The study by Swallow et al. revealed no significant overall survival and disease-free survival benefit in treating DDLPS[2]. Follow-up with repeat CT scans are usually done every 3-6 months for the first 3 years to monitor recurrence.

Conclusions

Retroperitoneal liposarcomas are rare, locally aggressive tumors with an indolent course. They cause mass-effect symptoms when they reach extreme dimensions. Complete surgical resection with negative margins is the mainstay of treatment and provides the best chance of attaining disease-free survival.

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