



# A Large Dedifferentiated Retroperitoneal Liposarcoma Extended to the Testis: A Rare Case Report

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## Abstract

Liposarcomas are neoplasms of mesodermal origin, they represent less than 1 % of all malignant tumors and 1 to 2 % of urogenital lesions. Primary retroperitoneal liposarcomas extending into the inguinal canal are rare. We present the case of a large retroperitoneal liposarcoma invading the left testicle and its spermatic cord. It was diagnosed by imaging as a large mass which compresses the surrounding abdominal structures with probable communication to the inguinal canal. A surgical intervention consisting of en bloc resection of the tumor and the left testicle with its cord was performed by 2 routes, intercostal and inguinal. The histological study showed dedifferentiated liposarcoma which is a rare entity with a high rate of malignancy and a poor prognosis. The treatment of choice is wide surgical resection with clean margins because chemotherapy and radiotherapy are less sensitive according to the literature. The postoperative follow-up was simple during 6 months of follow-up.

Received: Jan 30, 2023

Accepted: Feb 23, 2023

Published Online: Feb 28, 2023

Journal: Annals of Urology Research & Practice

Publisher: MedDocs Publishers LLC

Online edition: <http://meddocsonline.org/>

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## Introduction

Retroperitoneal liposarcomas (RPLPS) are rare mesenchymal tumors. They account for 7.5 to 25 % of all soft tissue sarcomas and 1 to 2 % of urogenital lesions [1]. Liposarcoma most often develops from the soft tissue of the extremities. However, retroperitoneal location of this tumor is rare. Extension of RPLPS into the inguinal region with scrotal involvement is uncommon [2]. Given the large size of the tumor and its invasion into adjacent organs, complete resection with negative margins constitutes a surgical challenge [3]. We report a rare case of large retroperitoneal liposarcoma involving the testis and its spermatic cord.

## Case Report

A 58-year-old man, with a history of chronic smoking who presented with an abdominal mass, progressing gradually for three months and left scrotal swelling. He was in good general condition with no urinary or transit disorders. The physical examination showed a large palpable firm mass occupying the entire left flank, painless and giving lumbar contact.

An inguino-scrotal MRI showed thickening of the scrotal envelopes and a large fatty mass in the left flank and iliac fossa on the sections of the abdominal floor. A thoraco-abdomino-pelvic CT showed the presence of a voluminous retroperitoneal

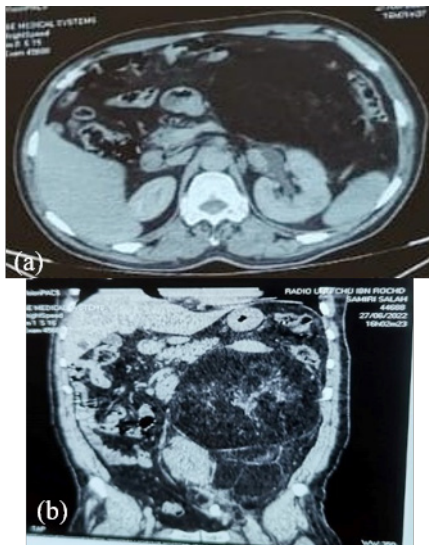


**Cite this article:** Habonimana P, Niyonkuru E, Nisabwe S, Mazti A, Moataz A, et al. A Large Dedifferentiated Retroperitoneal Liposarcoma Extended to the Testis: A Rare Case Report. *Ann Urol Res.* 2023; 1(1): 1001.

mass of the left flank of fatty density measuring 20 x 18 x 21 cm, seeming to communicate with the left inguinal canal, and responsible for a minimal left pyelocaliceal dilation. These radiological features suggested a liposarcoma of the retroperitoneum (**Figure 1**).

The patient underwent surgery under general anesthesia in two stages: via an intercostal incision (between the 10th and 11th ribs) and left inguinal via the upper route. Resection of the retroperitoneal tumor was difficult because it was large and invaded the adjacent structures. The mass drove the peritoneum forward, the spleen upwards and the renal compartment backwards. Its extraction required the opening of the peritoneum and the realization of an extended detachment up to the left colic angle and the release of the left ureter up to its crossing with the iliac vessels. The release of the lower pole of the mass revealed its extension through the left inguinal orifice with the spermatic cord hard on palpation. The intervention continued with the monobloc extraction of the retroperitoneal mass which infiltrates the spermatic cord with extension as far as the left testicle, completed by the excision of the small masses at the expense of the left kidney, left mesocolon and bladder (**Figure 2**).

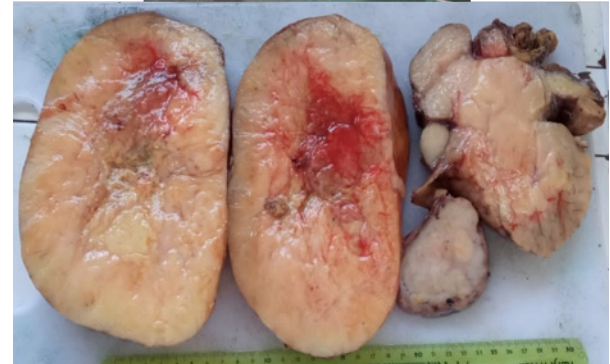
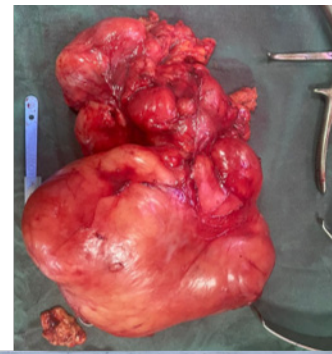
The resected specimen have been submitted to the pathological examination. The macroscopic examination of the specimen revealed an encapsulated mass 4440 g in weight, measuring 30 x 25 x 8 cm, firm, yellow white, fatty with hemorrhagic and necrotic areas. The histological examination concluded a dedifferentiated retroperitoneal liposarcoma involving the testis and spermatic cord. The postoperative course was uneventful (**Figure 3**).



**Figure 1:** Axial (a) and sagittal (b) sections of an abdominal-pelvic CT scan showing a retroperitoneal liposarcoma compressing the surrounding structures and responsible for left pyelocaliceal dilation.

### Discussion

The description of Liposarcoma (LPS) is by far the oldest, under the term “myxoma lipomatode” by Virchow [4]. However, a perplexity still lies in its pathogenesis; the role of trauma, ionizing radiation, an oncogenic virus, certain chemicals and the occurrence of a liposarcoma in a lipoma remain incriminated [1]. LPS is a malignant soft tissue tumor of mesenchymal origin, developed from immature lipocytes at different stages of differentiation. It represents 14 to 18 % of all malignant soft tissue tumors [5].



**Figure 2:** Macroscopic appearance of a voluminous dedifferentiated liposarcoma, retro-peritoneal of 30 cm well encapsulated, firm with whitish areas, necrotic, slightly hemorrhagic and 20 cm hard testicle with unidentifiable spermatic cord lumen.



**Figure 3:** Illustrative image of wound healing with redon drains in place on postoperative day 7.

The disease commonly occurs in age groups of fifty with a slight male predominance [5,6]. Our patient was 58-year-old and was in the range of age as reported in the literature. The slow increase in tumor size and the compliance of the retroperitoneal space explain the pauci-symptomatic character and the large volume of the tumor at the time of diagnosis (up to 40 kg) [7]. The mass effect of the tumor or even the invasion of adjacent organs is expressed over time by abdominal pain or heaviness in 50 to 80 % and the perception of an abdominal and/or pelvic mass in 70 to 80 % cases. The associated symptoms, urologic or digestifs are rare [6]. In our case, the major symptom was an abdominal mass and a scrotal swelling with no other associated symptoms.

Scrotal involvement is more common with inguinal LPS than with RPLPS because the latter less often extends into the inguinal canal. Rhu et al. [2] in their comparative clinicopathologic study on LPS (RPLPS vs inguino-scrotal LPS) showed the clinical similarity of two those locations and the challenge to determine the accurate origin of the tumor (either inguinal or retroperitoneal). In all RPLPS, they found only 3.6 % with an extension into the inguinal canal. Inguino-scrotal invasion of retroperitoneal LPS is rarely reported in the literature. In our patient, the RPLPS extended through the left inguinal orifice and invaded the spermatic cord and the testis. Therefore, it is essential to research for probable tumors hidden in the retroperitoneal space when

inguinal masses are suspected to be the sarcomas. The inguinoscrotal LPS can be isolated in this area without extending into the retroperitoneal space and vice versa [2]. CT and MRI are the mainstay of diagnosis aid for these tumors and specify the relationship with adjacent organs [6]. Imaging makes it possible to link the mass to the retroperitoneum, to carry out a diagnosis approach, to carry out the locoregional and remote extension assessment and to plan the operative strategy. The imaging performed in our patient revealed a large retroperitoneal mass pushing back the nearby structures, communicating with the inguinal canal and a scrotal location in favor of liposarcoma.

Pathological examination is the gold standard for diagnosis of LPS. It specifies the type, grade, level of invasion and guides therapeutic strategies. The World Health Organization WHO, in its 5th edition 2020 of classification of soft tissue and bone tumors, classifies LPS into 4 subtypes, which can be compiled into 3 groups: myxoid LPS (56.2 %), atypical LPS (well-differentiated LPS 21.9 %, dedifferentiated LPS 6.8 %) and pleomorphic LPS (17.8 %) [8,9]. Atypical LPS are characterized by an amplification of the MDM2 gene and CDK4, myxoid LPS by a rearrangement of the DDIT3 gene and finally pleomorphic LPS are components with a complex genome [8]. The DDLS have a strong propensity for locoregional versus distant recurrence despite advanced means of diagnosis, macroscopically complete surgical excision and possibly additional treatment. The local recurrence rate is roughly around 50 % [3]. The presence of metastases is rare at the time of diagnosis. Among all histological subtypes, DDLS is high grade, faster growing and far metastatic [10]. The rate of metastases systemic is evaluated between 5 % and 29.7 % [11].

Surgical resection remains the mainstay of treatment for LPS and local recurrences [1,2]. In the case of giant retroperitoneal liposarcoma, complete resection of the tumor and removal of invaded adjacent organs is the gold standard of the treatment. The average rate of resectability in large series is around 70 % with complete resections in half of the cases [3]. RPLPS is a difficult cancer to treat. In case of high-risk disease and positive surgical margins, chemotherapy is considered despite the fact that LPS are less chemo sensitive [6,10]. The use of a multimodal treatment, combining chemotherapy and radiotherapy to improve recurrence rates and prognosis, is of limited effect to date. According to Keita MM et al, the rate of local relapse and metastasis is 13 % vs 14 %, 37 % vs 29 % and 50 % vs 34 % respectively after 1 year, 3 years and 5 years of appropriate surgical intervention [1].

### Conclusion

Extension of retroperitoneal liposarcoma through the inguinal canal is an extremely rare situation. Its clinical revelation is late because of the compliance of the retroperitoneal space. Abdominal computed tomography and/or magnetic resonance imaging aid in diagnosis; however, pathological examination remains the cornerstone of diagnosis. Despite appropriate means of treatment, the rate of local recurrences remains high. It is therefore important to follow the patient in order to quickly identify any sign of relapse or metastasis.

### Consent

The patient freely consented to the publication of the case.

### Conflict of Interest Statement

None declared.

### Acknowledgments

At the end of this work, our thanks go to all the people who have actively participated in its writing, including histology staff for identification of histological type.

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