

## **Atypical Retroperitoneal Liposarcoma Mimicking Retroperitoneal Teratoma: A Case Report**

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### **Abstract**

Liposarcomas are malignant neoplasms deriving from the fatty tissue and are known to be the most common soft-tissue sarcomas diagnosed in adults. Dedifferentiated ones are uncommon, with poor prognosis and higher local recurrence rate. The preoperative diagnosis can be easy in the typical forms and is challenging in the atypical ones. Here we describe an atypical case of dedifferentiated retroperitoneal liposarcoma in a 37 years old male patient, with radiologic findings highly suggestive of teratoma with several components including calcifications, cystic areas and a solid component. The tumor was entirely removed through midline incision. As radiologic diagnosis was challenging for radiologists, it was the same for anatomopathologists. To the best of our knowledge, about thirty cases with similar radiologic findings were reported in the literature.

**Keywords:** *Dedifferentiated Liposarcoma; Retroperitoneum; Surgery*

### **Introduction**

Liposarcomas are malignant neoplasms deriving from the fatty tissue and are known to be the most common soft-tissue sarcomas diagnosed in adults (10%) [1], though 10 - 15% of all liposarcomas arise in the retroperitoneal space [2].

Retroperitoneal liposarcomas are rare malignant tumors with an aggressive disease course and high recurrence rate. It commonly occurs in patients with 40 - 60 years-old with a 1/1 ratio between male and female [3].

Its tardive discovery is caused by shortage of the clinical signs and its deep retroperitoneal localization. The preoperative diagnosis can be easy in the typical forms and is challenging in the atypical ones. Surgical resection remains the mainstay of therapy and is required for definitive diagnosis [4].

This report illustrates a dedifferentiated retroperitoneal liposarcoma (DDL) mimicking a retroperitoneal teratoma, presenting as an abdominal mass in a 37-year old patient. The diagnosis was made on an operative specimen after an immunohistochemical study. A review of the current clinical literature on this topic supports our management of this case.

### **Case Report**

A 37-year-old male presented with a 9-months history of left abdominal pain and a palpable abdominal mass growing rapidly. There was no associated bowel or urinary symptoms. Physical examination revealed a firm, non-tender mass, about 20 cm in diameter, occupying the left lower quadrant and left flank.

Biological tests, including serum alpha-fetoprotein (AFP), CEA and CA 19-9 were unremarkable. Routine blood test and urine analysis findings were also within normal limits. Plain abdominal films showed calcifications projecting on the left laterovertebral space (Figure 1). An abdominal and pelvic computed tomography scan was performed revealing a 12\*15\*13 cm mass in the left retroperitoneum with several components including arcuate calcifications, cystic areas and a solid component which enhanced after bolus injection of iodinated contrast media (Figure 2). No colonoscopic study was performed in our case.

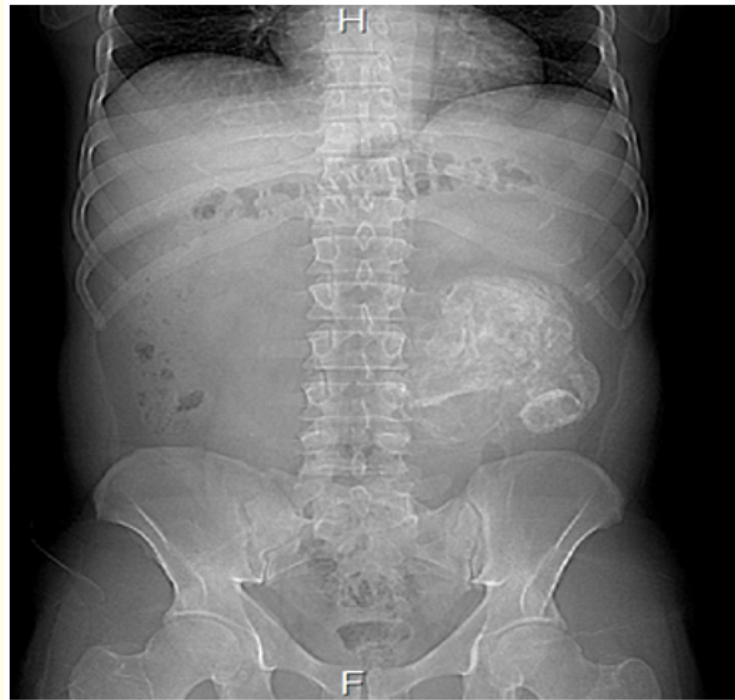


Figure 1

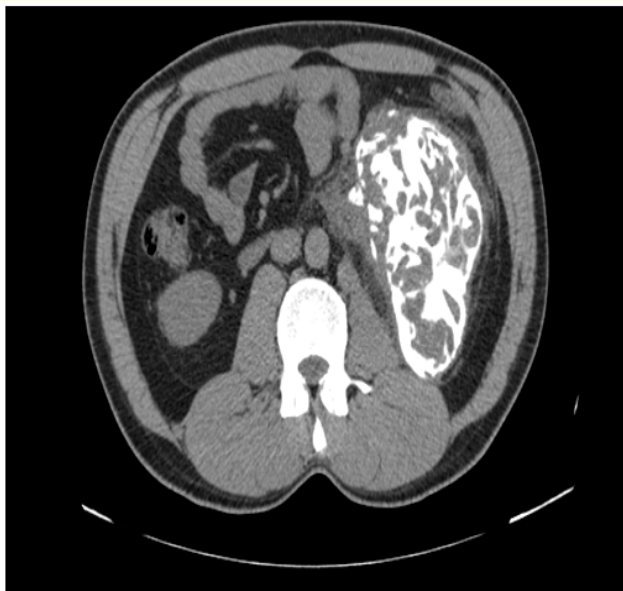
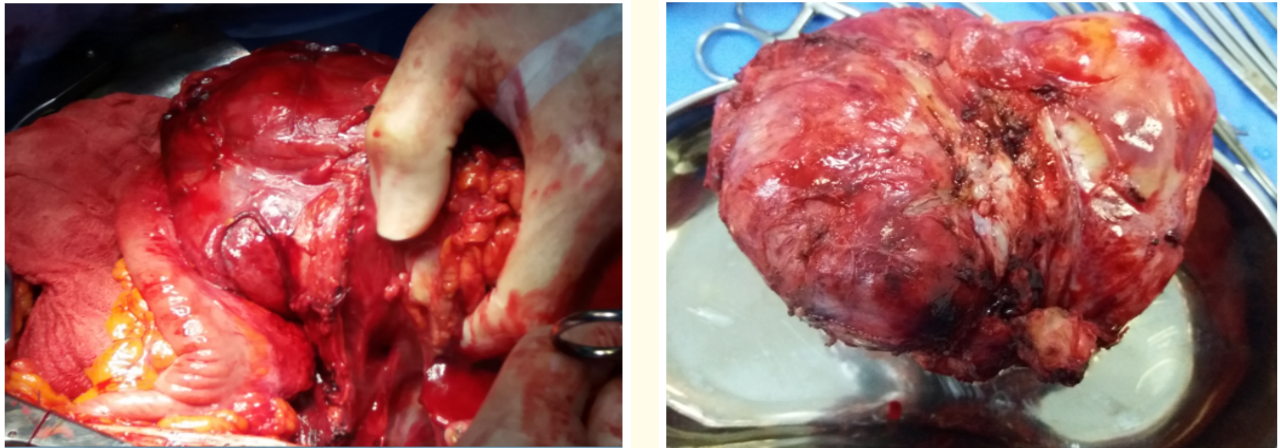


Figure 2

Provisional diagnosis of primitive retroperitoneal teratoma was made and an explorative laparotomy was performed through a mid-line incision, revealing a large retroperitoneal tumor displacing the left colon anteriorly and medially and the left kidney upperly and medially. There were dense fibrous adhesions of the mass with the psoas muscle. The retroperitoneal dissection was tedious and difficult. The tumor was excised entirely after left colonic detachment (Figure 3).



**Figure 3**

The postoperative course was uneventful, and the patient was discharged from the hospital 5 days later.

Histopathological examination of the surgical specimen showed a de-differentiated liposarcoma with a meningotheial-like component and a bone differentiation. An immunohistochemical study was conducted and the tumor was immunoreactive for MDM2 and CDK4. Thus, a diagnosis of retroperitoneal DDL with high-grade osteosarcomatous transformation was confirmed. The tumor was classified as T2a N0 M0 according to the AJCC /UICC staging. No recurrence was detected at the 2-year follow-up.

### **Discussion**

Retroperitoneal liposarcoma (RPLS) is a rare tumor, with an incidence of 2.5 per million. It is the most common primary retroperitoneal malignant soft tissue tumor ranging from 20 % to 45 % [5] followed by leiomyosarcoma (17 - 29%), malignant fibrous histiocytoma (7 - 17%), malignant peripheral nerve sheath tumor (8%), rhabdomyosarcoma (7%), synovial sarcoma (2%), and Ewing sarcoma (2%) [3]. According to the 2002 World Health Organization histologic classification of soft tissue tumors it is divided into five subtypes, including well differentiated, myxoid, round cell, pleomorphic, and dedifferentiated liposarcomas [6]. The dedifferentiated tumors are characterized by local aggressiveness with high metastatic potential. They have been associated with a 6-fold increased risk for death and a 15-fold increased risk for distant recurrence compared with well-differentiated tumors [7].

Recently many studies demonstrated that the use of 18- fluorodeoxyglucose positron emission tomography FDG PET/CT can be viewed as a surrogate method for grading liposarcomas, given the strong correlation between the standardized uptake value (SUV) and tumor grade. Well-differentiated liposarcomas showed a significantly lower SUV than myxoid/round cell and pleomorphic subtypes [8]. In addition to that FDG PET/CT is highly sensitive and reasonably specific for the detection of early relapse in liposarcoma [9].

The peak incidence of liposarcoma occurs from 40 to 60 years of age with a 1/1 ratio between male and female [3]. Retroperitoneal liposarcoma often are asymptomatic and incidentally diagnosed. As tumor mass increases it may produce a wide range of signs and symptoms due to the compression and infiltration of the surrounding organs: causing back or abdominal pain, gastrointestinal symptoms (nausea, vomiting, and constipation) or lower extremity edema [10].

Physical examination may detect a midline or paramedian abdominal mass with limited mobility or only abdominal tenderness. Both CT and MRI may provide an accurate assessment of the characteristics of soft tissue sarcomas and the involvement of adjacent structures in the typical forms. The diagnosis of DDL can be confound with other retroperitoneal tumors, such as retroperitoneal teratoma, since we can identify various components, including bone, soft-tissue density structures, adipose tissue, and sebaceous and serous-type fluids, but the presence of macroscopic fat usually leads to the presumptive diagnosis of liposarcoma [6].

Magnetic resonance imaging should be considered to be the best method for the diagnosis of retroperitoneal tumors, for showing the precise location of the tumor and its anatomic relationships with adjacent structures which provide better preoperative planning and increased likelihood of complete removal of the tumor [11]. In our case the tumor was considered to be resectable without the necessity to practice an MRI imaging. Because definitive diagnosis is only achieved after histologic evaluation of the specimen, surgical resection is overriding for both diagnosis and treatment.

Although challenging, is still currently the primary method of locoregional control and the most important prognostic factor. Complete resection is feasible in 80% to 88% of cases. Resection of adjacent structures is required in 57% to 77% of cases to achieve complete resection of the tumor [1,4].

Liposarcomas are among the most radiosensitive of the sarcomas and adjuvant radiotherapy has been occasionally used in RPLS [12]. However, substantial morbidity is associated in this setting because of the proximity of dose-limiting structures, such as the small intestine, spleen and bone marrow.

The role of chemotherapy for soft tissue sarcomas of the retroperitoneum remains controversial. Only two drugs, doxorubicin and ifosfamide, have demonstrated a relatively consistent single-agent activity, with response rates ranging from 10% to 25% [13].

Successful treatment of recurrent RPLS using CT-guided radiofrequency ablation has been recently reported and appears to be feasible therapeutic approach for patients with previous surgery [14].

### Conclusion

Retroperitoneal liposarcoma is a rare tumor and exhibits considerable histological heterogeneity. Despite the development of computed tomography and MRI imaging the diagnosis of the dedifferentiated forms of retroperitoneal liposarcoma still challenging requiring an anatomopathological examination with immunohistochemical study.

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