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*Case report*

# Asymptomatic dedifferentiated liposarcoma mimicking renal cell carcinoma—A rare case report and review of literature

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**Abstract**
*Introduction:* Soft tissue sarcomas are rare and account for one percent of all cancers globally. Approximately 15% of these tumours are found in the retroperitoneum and have a peak incidence at around 40 to 60 years of age.
*Observation:* We report on a extremely rare case of asymptomatic perinephric dedifferentiated sclerosing liposarcoma which appeared to mimic cystic renal cell carcinoma on clinical and radiological presentation. The patient underwent a right partial nephrectomy and remains tumour free at six months follow up.



## Introduction

Soft tissue sarcomas (STS) are rare and account 1 pecent of all cancers globally [1]. Liposarcoma is the second most common soft tissue sarcoma in adults after fibrous histiocytoma. Of all sarcomas, the majority occur outside of the retroperitoneum. Only 10–20% of sarcomas occur in the retroperitoneum, of which 35% arise in the perinephric fat. The peak incidence is in the 5th decade of life with a distinct male predominance, but no racial predominance [2]. It is

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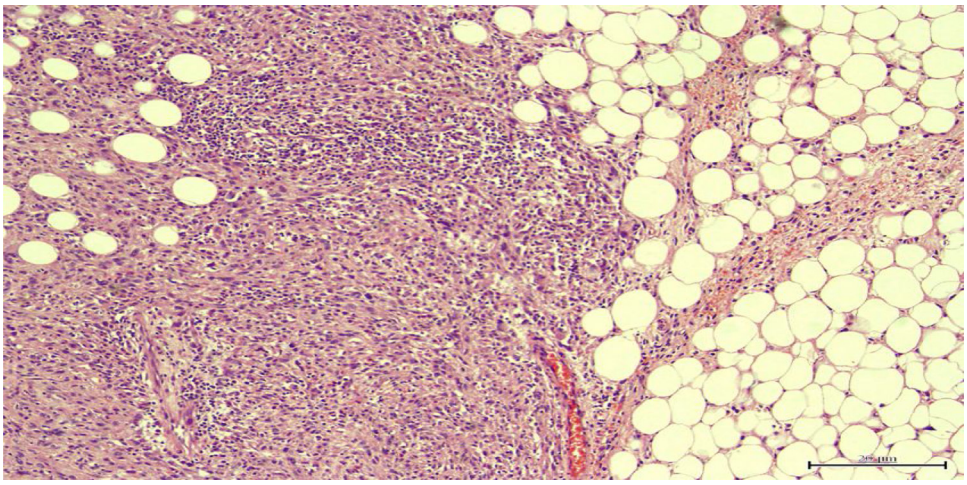

**Figure 1** Abrupt transition between well differentiated sclerosing liposarcoma and dedifferentiated liposarcoma with myoid features (H&E X200).

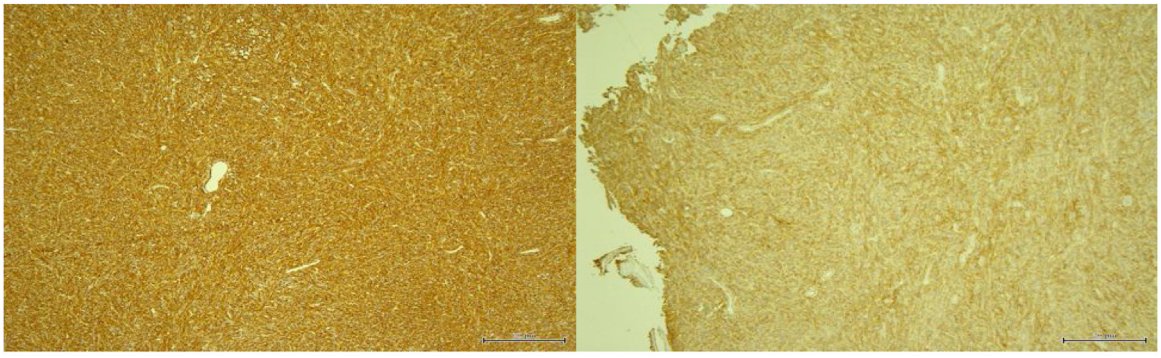


**Figure 2** **Immunohistochemical profile of dedifferentiated liposarcoma**. (A) Smooth muscle actin positive. (B) CD99 positive (H&E X200).

a malignant tumour of mesenchymal origin in which the bulk of the tumour differentiates into adipose tissue. There are no proven associated conditions related to liposarcoma.

## Case report

An 86 year old man was referred to Urology team with an asymptomatic renal mass found incidentally following a CT scan. The patient had a background history of malignant melanoma on the planter surface of the right foot which was treated with local excision two years previously. On routine follow up, a chest x-ray showed a soft tissue shadow in the right lower lobe which was confirmed as a small nodule on CT scan of chest. This was thought to be a metastatic deposit originating either from the primary melanoma or from potential renal cell carcinoma given the cystic nature of renal mass. The patient was also under regular follow up for raised PSA of 9.2 μg/L. His other co-morbidities included hypertension and bilateral cataracts which had been previously treated successfully. The patient's physical examination was normal and routine blood tests including full blood count, creatinine and urine analysis were unremarkable.

As per MDT recommendation, the patient underwent diagnostic laparoscopy which showed a large right sided renal mass at the lower pole. The operative team then proceeded to perform an open partial nephrectomy. The tumour measured 140 mm × 90 mm × 30 mm. The histology confirmed that it is a well-differentiated liposarcoma present around the nodular areas of dedifferentiated liposarcoma with positive margins indicating an incomplete resection. The immuno-histochemical profiling of the tumour cells demonstrated a strong diffuse positive reaction with smooth muscle actin (SMA) and a focal weak positive reaction with CD99. Negative reactions are seen with AE/3, S100, HMB45, Melan A, Cytokeratin 7, Cytokeratin 20, desmin and inhibin. The latter profile is important in excluding metastatic melanoma and metastatic prostatic carcinoma given the patient's past medical history (Figs. 1 and 2).

The patient made a good recovery and no chemotherapy or radiotherapy was given. At present, the patient remained well and tumour free at six months with no evidence of tumour recurrence on CT scan.

## Discussion

Liposarcomas are one of the most common soft tissue sarcomas and frequently occur in the extremities and the retroperitoneum of adults. Liposarcomas are histologically defined as being tumours composed of lipoblasts. They are currently classified into five groups: myxoid liposarcoma, well differentiated liposarcomas, round cell (poorly differentiated myxoid liposarcomas), pleomorphic liposarcomas and dedifferentiated liposarcomas. Among these, myxoid liposarcomas are the most common type, found in approximately 50% of cases, followed by well differentiated with 25% of cases.

The clinical characteristics are closely related to the histological type.

Although recurrence is common in deep seated liposarcomas of all types, well differentiated and myxoid liposarcomas have a good prognosis and their rates of metastasis are low compared to the other types of liposarcomas.

Our case is unique as it is an asymptomatic dedifferentiated sclerosing liposarcoma which has presented as a cystic renal cell carcinoma. The mode of presentation is highly unusual with only one other case in our knowledge reported in the world literature of a liposarcoma mimicking a cystic renal cell carcinoma. We could not find any association or link with metastatic melanoma or with prostate cancer although initially it was suspected in this case. The current literature demonstrates that cutaneous melanoma can metastasize to the retroperitoneum, however, there is no known association between cutaneous melanoma and the formation of dedifferentiated liposarcoma. Dedifferentiated liposarcomas are considered to be a high-grade form of well-differentiated liposarcoma which is more aggressive and has greater potential for metastasis [3,4]. It can also occur as a primary tumour or arise from recurrence after resection of well differentiated liposarcomas [5].

Liposarcomas located retroperitoneally most commonly present late and metastasize to nearby major vessels and organs. Typical presentations are abdominal discomfort (60%–70%) with or without a palpable abdominal mass (70%–80%) [6] and occasionally genitourinary or gastrointestinal symptoms from extensive compression from a large mass on adjacent viscera [7].

## Treatment and prognosis

Complete resection with wide excision to include a margin of normal tissue offers the best chance of cure however, at times, this can only be achieved at the expense of important organs or structures. Some authors reported that radiation and chemotherapy are helpful for both primary and metastatic lesions, however, the efficacy of this type of treatment is still controversial [8–10]. Some centre's recommend neo-adjuvent chemotherapy [11], however, the use of this treatment modality requires more studies to ascertain the long-term benefit with regards to morbidity and mortality. In our case we did not commence any adjuvant treatment as per MDT recommendations. In cases of locoregional recurrence, re-resection may give good results.

The overall, five year survival rate has been shown to be 36–58% and is highly dependent on tumour size, grade, extent of previous resection and extent of surrounding tissue invasion [12]. Post-operative clinical follow up and imaging must continue up-to and beyond 5 years due to the continued risk of long-term relapse [13].

## Conclusion

Asymptomatic perinephric dedifferentiated sclerosing liposarcoma which mimics renal cell carcinoma is an extremely rare and aggressive tumour. Aggressive radical total excision of tumour with healthy tissue margins is the gold standard of treatment. Other treatment modalities are controversial and less evidence based, however, a multidisciplinary approach is paramount to treat the patient effectively.

## Consent

Patient consent had been obtained verbally. All efforts were made to remove possible identifiers from the text.

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## Conflict of interests

No conflict of interests.

## Authors' Contributions

Mohamed El Howairis: writing manuscript.

Rohit Malliwal: data acquisition.

Shiv Bhanot: supervision and manuscript editing.

Noor Buchholz: Writing, editing, supervision, submission.

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