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Case report

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

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KEYWORDS
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Abstract

Introduction: Soft tissue sarcomas are rare and account for one percent of all cancers globally. Approximately 15% of these tumors are found in the retroperitoneum and have a peak incidence at around 40–60 years of age.

Observation: We report on an extremely rare case of asymptomatic perinephric de-differentiated sclerosing liposarcoma mimicking a cystic renal cell carcinoma on clinical and radiological presentation. Difficulties in diagnosis and therapy of this rare tumor are discussed in the light of the available literature.

Introduction

Soft tissue sarcomas are rare and account for 1% of all cancers globally [1]. Liposarcoma (LS) is the second most common soft tissue sarcoma in adults after fibrous histiocytoma. The majority of sarcomas occur outside the retroperitoneum. Only 10–20% of sarcomas occur within, with 35% arising 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 a malignant tumor of mesenchy-

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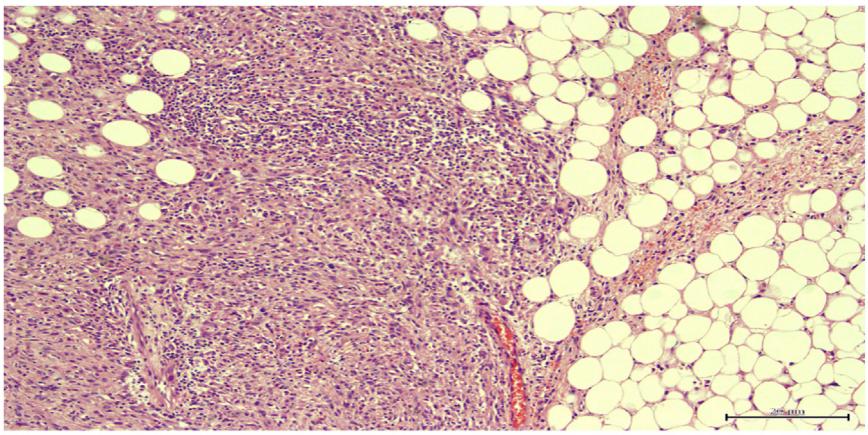

Figure 1 Abrupt transition between well differentiated sclerosing and de-differentiated liposarcoma with myxoid features (H&E ×200).

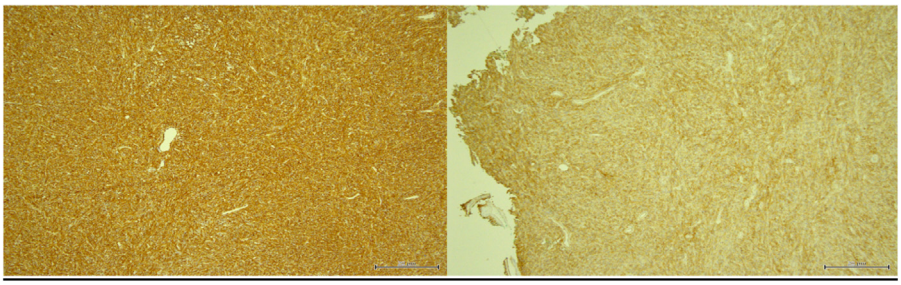

Figure 2 Immunohistochemical profile of dedifferentiated liposarcoma.
Left: smooth muscle actin positive, right: CD99 positive (H&E ×200).

mal origin with the bulk of the tumor differentiating into adipose tissue.

Case report

An 86 year old man was referred to urology with an incidental asymptomatic cystic renal mass on CT for follow up of a plantar malignant melanoma locally excised two years previously. A small nodule in the right lower lung lobe was thought to be a metastasis either from the melanoma or from the newly discovered renal mass. The patient was also followed up for a raised PSA of 9.2 ug/L. The patient's physical examination was normal and routine blood tests unremarkable.

After discussion in the multidisciplinary team meeting (MDT), an atypical cystic renal cell carcinoma (RCC) was suspected and the patient underwent diagnostic laparoscopy confirming a large right lower pole renal mass. With regard to his age, close serial observation of the tumor or surgical resection were offered to the patient. The patient elected for partial nephrectomy. An open partial nephrectomy was performed which was able to preserve at least 50% of the kidney as the mass had a significant exophytic component.

Histology confirmed a well-differentiated LS around nodular areas of a de-differentiated LS (Fig. 1) measuring 140 mm × 90 mm × 30 mm. Immuno-histochemical profiling demonstrated a strong diffuse positive reaction with smooth muscle actin (SMA) and a focal weak positive reaction with CD99 (Fig. 2). Negative reactions are seen with AE/3, S100, HMB45, Melan A, cytokeratin 7, cytokeratin 20, desmin and inhibin. Thus malignant melanoma and prostatic adenocarcinoma could be excluded.

The patient recovered without complications. MDT recommended no immediate adjuvant therapy. The patient remains under regular surveillance and is tumor-free at six months post-operatively.

Discussion

Liposarcomas are common soft tissue sarcomas and frequently occur in the extremities and the retroperitoneum of adults. LS are histologically composed of lipoblasts. They are classified into five groups:

- Myxoid
- Well differentiated
- Round cell (poorly differentiated myxoid)
- Pleomorphic
- De-differentiated.

Myxoid type is the most common with 50%, followed by well differentiated with 25%. The clinical characteristics are closely related to the histological type.

Although recurrence is common in deep seated LS of all types, well differentiated and myxoid LS have a good prognosis with a low rate of metastases compared to other types.

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Our case is unique as it is an asymptomatic de-differentiated sclerosing LS which presented as a cystic RCC. This presentation is highly unusual with only one other case to our knowledge reported in the literature. De-differentiated LS are considered a high-grade form of a well-differentiated liposarcoma, more aggressive and with greater potential for metastases [3,4]. It can occur as a primary tumor, or arise from recurrence after resection of a well differentiated LS [5].

Retroperitoneal LS typically present late and metastasize to nearby major vessels and organs. They present with abdominal discomfort (60–70%), palpable abdominal mass (70–80%) [6], and occasionally with genitourinary or gastrointestinal symptoms from extensive compression of adjacent viscera [7].

Complete surgical resection including a margin of normal tissue offers the best chance of cure. Sometimes, this can only be achieved at the expense of important organs or structures. Radiation- and chemotherapy (adjuvant and neo-adjuvant) have been reported as useful for both, primary and metastatic lesions, however their efficacy is controversially discussed [8–11]. The overall 5 years-survival rate is 36–58% and depends on tumor 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].

De-differentiated sclerosing LS mimicking RCC is an extremely rare and aggressive tumor. Radical excision with wide healthy tissue margins is the gold standard of treatment. Other treatment modalities are controversial and lack an evidence-base.

Consent

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

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Authors' contribution

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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