

**Case report:****Dedifferentiated liposarcoma of the perinephric fat**

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Received:23<sup>rd</sup> July 2008; Accepted:15<sup>th</sup> Nov 2008.**ABSTRACT**

A 55 years old female with dedifferentiated liposarcoma is described in which the diagnosis was unsuspected clinically and diagnosed histopathologically. The clinical and pathological features are briefly reviewed.

**الخلاصة:**

وصف لحالة سيدة عمرها ٥٥ سنة مصابة بلحمانية دهنية في النسيج الدهني حول الكلية وغير المتوقعة سريرياً وشخصت بالفحص النسيجي وروجعت الحالة السريرية والباثولوجية باختصار.

**R**etroperitoneal sarcoma constitutes 10-15% of all soft tissue sarcoma in adults, the most common of which is liposarcoma whether well differentiated or dedifferentiated<sup>(1)</sup>. Twelve to forty percent of liposarcoma in the body present in the retroperitoneum, 35% of which arise from the perinephric fat<sup>(2)</sup>. Dedifferentiated liposarcoma was first described by Evans *et al*<sup>(3,4)</sup> who used to describe focal poorly differentiated areas within well-differentiated lesions<sup>(4)</sup>. These features may resemble malignant fibrous histiocytoma, or other undifferentiated components<sup>(4)</sup>. These dedifferentiated features are described mainly in lipomatous tumors<sup>(5)</sup>. Dedifferentiated liposarcomas occur in adults with highest incidence in the sixth and seventh decades of life and have an equal sex predilection<sup>(6)</sup>.

We report this case, because it is rare and may be misdiagnosed with other sarcomas and to the best of our knowledge it is the first case to be reported in our locality.

**Case report:**

**Clinical presentation:** A 55 years old female presented with a right hypochondrial swelling, associated with vague pain and constitutional

symptoms. Imaging study revealed abdominal mass.

Diagnostic laparotomy through right paramedian incision, revealed retroperitoneal mass related to the right kidney. Radical nephrectomy was done.

**Gross examination:** Right nephrectomy specimen with perinephric fat weighed 800g. Cut section revealed kidney tissue measuring 11x5.5x4cm, surrounded by multiple masses embedded in perinephric fat. The masses are ranging in size from 2.5- 8cm. They are firm, solid, whitish-yellowish in color with distinct gelatinous areas. The masses are attached to the capsule of the kidney but easily detached from its surface ( Fig.1).

**Microscopically:** Hematoxiline and eosin stained sections show cellular tumor composed of a well differentiated liposarcoma in the form of lipoblast proliferation (Fig. 2 ) and the presence of plexiform blood vessels (Fig.3,4) intermingled with dedifferentiated component in the form of whorling arrangement of spindle cells areas resemble MFH (Fig.5) and giant cells with eosinophilic cytoplasm (Fig.6) . Another striking feature is the presence of extensive lymphoid follicular infiltrate (Fig.7).



Figure (1): Tumor mass attach to the kidney tissue.

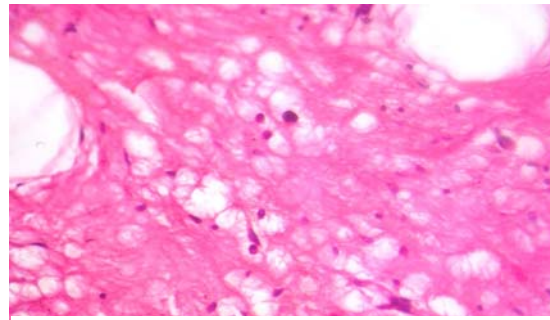


Figure (2): Liposarcoma with lipoblast proliferation, original magnification x100

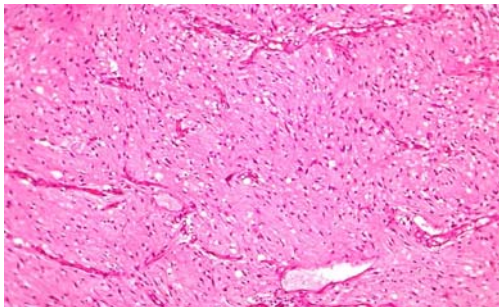


Figure (3) Liposarcoma with plexiform blood vessels, original magnificationx100

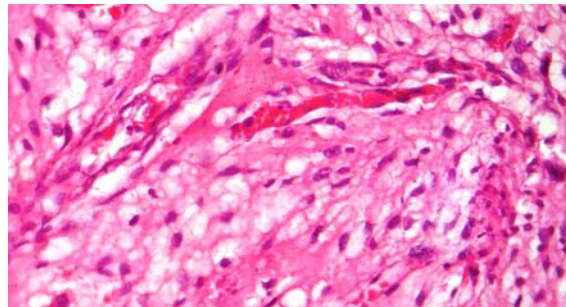


Figure (4): Liposarcoma with plexiform blood vessels, original magnificationx400

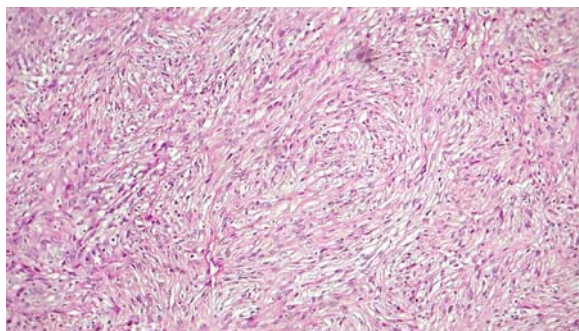


Figure (5): Liposarcoma with whorled appearance, original magnificationx100

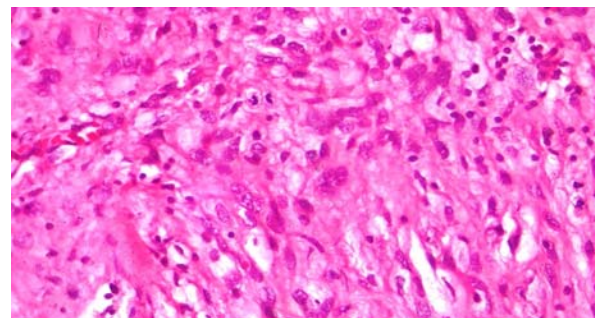


Figure (6): Liposarcoma with giant cell, original magnificationx400

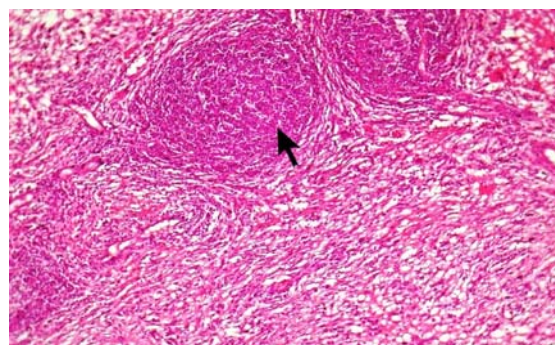


Figure (7) : Liposarcoma with lymphoid follicular infiltrate, original magnificationx100

## Discussion:

Dedifferentiated liposarcoma occurs in adults with no sex difference<sup>(7)</sup> but a study done by Qiang<sup>(2)</sup> observed a slight male predominance; the patient of present case is a female of 55years.

The case had the dedifferentiated component of liposarcoma, which arose de novo from the perinephric fat. However secondary dedifferentiated component can occur in a recurrent well differentiated liposarcoma<sup>(8)</sup>. Nephrectomy was done in 38% of patients<sup>(9)</sup>.

Retroperitoneal liposarcomas are usually large tumors, and can be single or multiple nodules. The proportion of well differentiated and dedifferentiated components are variable<sup>(6)</sup>. Therefore extensive sampling is recommended to avoid missing any component. The sampling should involve both the adipose and non-adipose areas; the former is important in the diagnosis<sup>(6)</sup>.

On the other hand any retroperitoneal tumor diagnosed as malignant fibrous histiocytoma or fibrosarcoma should be extensively sampled in order to demonstrate well differentiated component of liposarcoma<sup>(8)</sup>.

The majority of dedifferentiated liposarcomas commonly display high-grade areas resembling malignant fibrous histiocytoma or high-grade fibrosarcoma whereas minority shows low-grade areas resembling fibromatosis or well-differentiated fibrosarcoma<sup>(10)</sup>. They may show other forms of dedifferentiation including osteosarcoma, leiomyosarcoma, or even rhabdomyosarcoma. In the present case, the dedifferentiated liposarcoma displays variable histological pattern, including pattern of malignant fibrous histiocytoma and fibrosarcoma. Extensive lymphocytic infiltration is also observed and proved to be reactive in nature<sup>(11)</sup>. In addition to the morphologic characteristic of dedifferentiated liposarcoma, immunohistochemical marker can be of help in confirming such tumor from other<sup>(4)</sup>. MDM2, a recently described marker was reportedly useful for distinguishing DDLS from their histological mimics<sup>(4,12)</sup>, that is

unfortunately not available in our departments.

Dedifferentiated liposarcomas represent aggressive variants of liposarcomas<sup>(8)</sup>. Each morphological element of these heterogeneous tumors may manifest completely different biology. The overall biological behavior of dedifferentiated liposarcomas is likely dictated by the most aggressive element, which typically resides in the non-lipomatous portion of the tumor<sup>(8)</sup>. The prognosis is unrelated to the grade or extent but is related to mitotic activity of the dedifferentiated area<sup>(10)</sup>.

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