

GIANT RETROPERITONEAL LIPOSARCOMA

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ABSTRACT

Introduction: We report a case of a huge well-differentiated liposarcoma with dedifferentiated areas contributing to the patient's unfavorable outcome.

Case Report: A 77 year old woman presented with a 8-month history of progressive abdominal enlargement accompanied by diffuse abdominal discomfort. A firm mass was noted on the right quadrants of her abdomen. Computed tomography revealed 2 large masses displacing intra-abdominal organs. The masses were removed surgically and the histopathologic examination of the specimens showed a well-differentiated liposarcoma with dedifferentiated areas. Twenty six months after this surgery, the patient underwent another surgery to remove a dedifferentiated liposarcoma and died 2 years later.

Discussion: The outcome of a liposarcoma is influenced by its histological subtype. Well-differentiated liposarcomas are low-grade tumors that usually do not have an aggressive behavior, but may recur locally. However, our patient had a huge tumor (> 20 cm) with dedifferentiated areas, which are associated with a poor prognosis.

Key words: tumor; retroperitoneal neoplasm; sarcoma; liposarcoma
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INTRODUCTION

Liposarcoma is a rare mesenchymal tumor that occurs more frequently in the soft tissues of the extremities between the fifth and seventh decades (1). However, other sites such as the retroperitoneum and the abdominal cavity may be affected (2). Retroperitoneal liposarcoma may achieve a considerable size before diagnosis, thanks to its clinical presentation characterized by symptoms which are usually late and nonspecific (1). We report a case of a huge well-differentiated liposarcoma with dedifferentiated areas contributing to the patient's unfavorable outcome.

CASE REPORT

A 77-year-old woman presented with a 8-month history of progressive abdominal enlargement

accompanied by diffuse abdominal discomfort. During general examination, a firm mass without tenderness was noted on the right upper and lower quadrants of the abdomen. Computed tomography (CT) scan of the abdomen revealed 2 large masses (Figure-1).

At laparotomy, a huge retroperitoneal tumor consisting of 2 masses was completely removed (Figure-2). A multilobulated mass involving the right kidney and the right adrenal gland weighed 3400 g and measured 37.5 x 11 x 8 cm. The histopathologic examination of the surgical specimens showed a well-differentiated sclerosing liposarcoma. There was no evidence of renal and adrenal tumor infiltration. Some areas revealed dedifferentiation with a histological pattern compatible with fibrous histiocytoma.

Twenty six months after this surgery, the patient underwent another surgery to totally remove

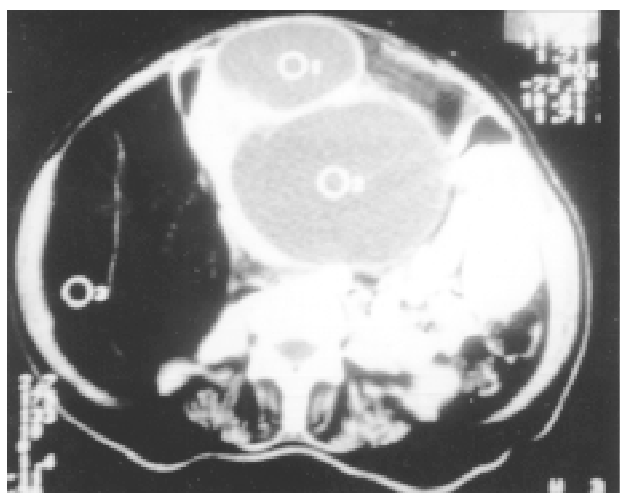


Figure 1 - CT scan revealing the giant liposarcoma composed of two masses displacing the abdominal viscera.

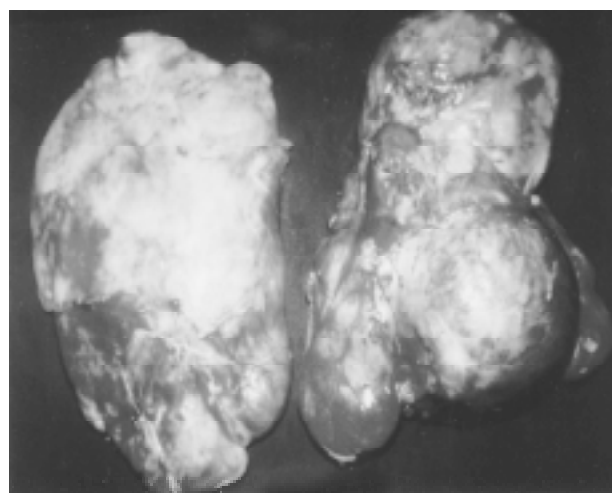


Figure 2 - Macroscopic aspect of the well-differentiated liposarcoma masses.

a dedifferentiated liposarcoma, at L3-L4 level adherent to the inferior vena cava, characterized by spindle cells arranged in fascicles in a schwannoma-like pattern. It weighed 980 g and measured 18 x 13 x 8 cm. Immunohistochemistry study was negative for S-100, CD68 and muscle-specific actin.

The patient died 2 years later by disease progression. Cachexia was evident and although no metastases were observed, another retroperitoneal mass was revealed at CT. At that point, the patient refused any additional surgery.

DISCUSSION

Liposarcomas are classified according to 3 histological types: well-differentiated, myxoid, and pleomorphic (2). Among all liposarcomas, the myxoid type is the most common, accounting for 60% of cases, while well-differentiated liposarcoma occurs in 25% and pleomorphic in 10% (2).

The outcome of liposarcomas is influenced by their histological subtype (3). Pleomorphic liposarcomas are highly aggressive with significant rates of recurrence and metastasis, while well-differentiated liposarcomas are low-grade tumors that usually do not have an aggressive behavior, but may recur locally (1,3).

Dedifferentiated areas and large tumors (> 20 cm) have been associated with poor prognosis (2,3). Dedifferentiation is defined as the presence of non-lipogenic high grade areas within the well-differentiated liposarcoma (2). Dedifferentiation is rare, occurring in 15% of the well-differentiated liposarcomas. In most cases, it recurs in the primary tumor in most cases, but may also develop later, usually after liposarcoma recurrence (2).

Divergent differentiation in schwannomas, osteosarcomas, leiomyosarcomas and rhabdomyosarcomas is possible (2). In our case, the histological presentation, at the moment of recurrence, was a well-known phenomenon called dedifferentiation. Immunohistochemistry study, using an antibody panel for identification of specific antigens related to different sarcomas was performed to make the differential diagnosis. All were negative excluding leiomyosarcoma, fibrous histiocytoma and schwannoma. The other retroperitoneal mass, possibly another recurrence, will not be discussed since its histological subtype was not diagnosed.

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