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A case report

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Rare giant dedifferentiated liposarcoma with osteosarcomatous component. A case report

We report a rare case of giant retroperitoneal dedifferentiated liposarcoma with osteosarcomatous component. The tumor was removed in a 63-year-old male that not showed any symptom. The histopathological examination highlighted a first differentiated neoplastic component and a second dedifferentiated neoplastic component where prevailed areas of osteosarcoma. To our knowledge only fourteen similar cases have been reported in the literature.

KEY WORDS: Dedifferentiated liposarcoma, Osteosarcoma

Introduction

Liposarcomas represent one of the most common soft tissue sarcoma. The annual incidence is about 2.7 new cases/ 1 million person/ year [1]. However , they are one of the most common soft tissue sarcomas (15% in adults)[2], and the most frequent histological type of retroperitoneal sarcoma, corresponding to 41% of these tumors[3,4]. They tend to grow at the extremities and in the retroperitoneum, where they may reach considerable size [2]. The peak of incidence is between 50-70 years. They are usually asymptomatic and rarely metastasize. Surgery is the only possible therapy with curative intent.

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Case report

A 63-year old male was referred to our Unit of General Surgery and Organ Transplantation of University Hospital of Parma for an incidental finding of massive retroperitoneal lesion. His surgical history included cholecystectomy and resection of malignant pleural mesothelioma that was performed 1 year ago by the Unit of Thoracic Surgery of our hospital. The patient had no symptoms referable to the mass and reported no weight loss or change in bowel habits. History included hypertension and type 2 diabetes mellitus. During physical examination an immobile solid tumor was palpated on the left half of his abdomen. Although blood investigations results were normal, contrast enhanced Computed Tomography and Magnetic Resonance of abdomen revealed "…presence of bulky lesion occupying almost fully the half left of abdomen (124x166x220 mm in size), that extends from the kidney's hilum to the ipsilateral iliac fossa and comes to the anterior abdominal wall, displacing the bowel loops contralateral. This lesion is composed of different components; the prevalent, with adipose thype signal (150x100x170 mm), capsulated,

occupies the central portion. Are also present: a calcified component (74x72mm) that comes close to the aorta; an additional component with cystic signal (72x59mm) and a solid component (67x32mm) with contrast enhancement. The left kidney is moved and rotated and the ipsilateral ureter is anterior to the lesion, in close contact with it. The left colon is displaced to the right. All these features most likely suggest a malignant tumor, probably a liposarcoma (Figg. 1, 2).

Preoperative investigations did not show any contraindication (Table I), therefore, on 18.09.2013, we proceeded to the remove of the retroperitoneal mass (after placing an ureteral stent) (Figg. 3, 4). The postoperative course was uneventful and the patient left our clinic on 25.09.2013.

Histopathological examination was indicative of dedifferentiated liposarcoma. In particular, the microscopic description highlighted two components: a first differentiated neoplastic component like usual type and sclerosing liposarcoma; a second dedifferentiated neoplastic component where prevail areas of osteosarcoma, infiammatory type pleomorphic sarcom, and fusocellular sarcoma.

At ambulatory visit at 3 months from surgical procedure the patient shows a good state of health.

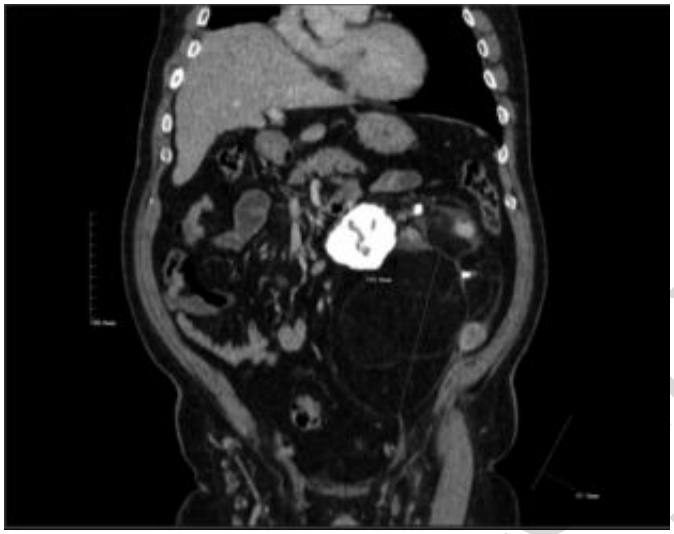

Fig. 1: Abdominal computed tomography (CT) reveals a giant tumor containing calcification.

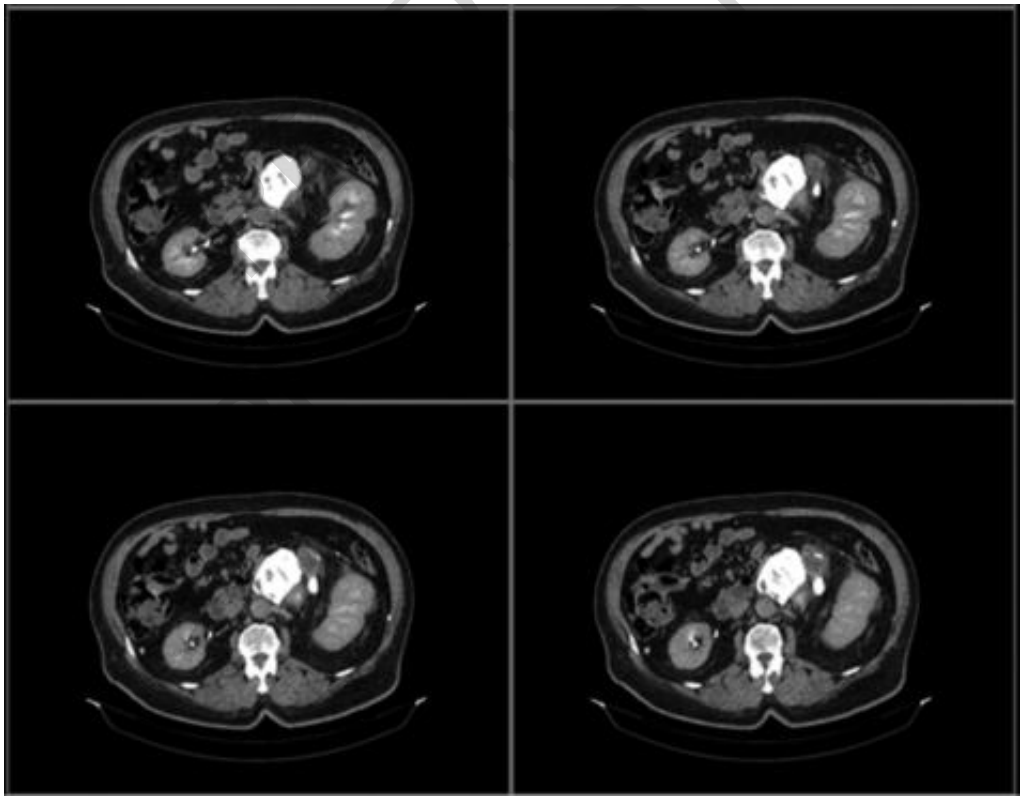

Fig. 2: Axial CT scan hows retroperitoneal mass containing ossified component.

TABLE I - *Preoperative investigations.*

WBC	7.50	x 10^3/uL
RBC	5.01	x10^6/uL
Hb	14.4	g/dL
Ht	42.3	%
MCV	84.4	fL
MCH	28.7	pg
MCHC	34.0	g/dL
RDW/SD	38.3	fL
RDW/CV	12.6	%
PLT	229	x10^3/uL
PDW	13.2	fL
MPV	10.7	um^3
Total bilirubin	0.5	mg/dL
Direct bilirubin	0.2	mg/dL
Total protein	6.9	g/dL
Albumin	4.3	g/dL
Gamma-GT	37	U/L
Alkaline phosphatase	65	U/L
AST (GOT)	17	U/L
ALT (GPT)	17	U/L
Pt	0.97	INR
aPtt	24.9	seconds
Creatinine	0.7	mg/dL
Iron	77	ug/dL
Ferritin	114	ng/mL

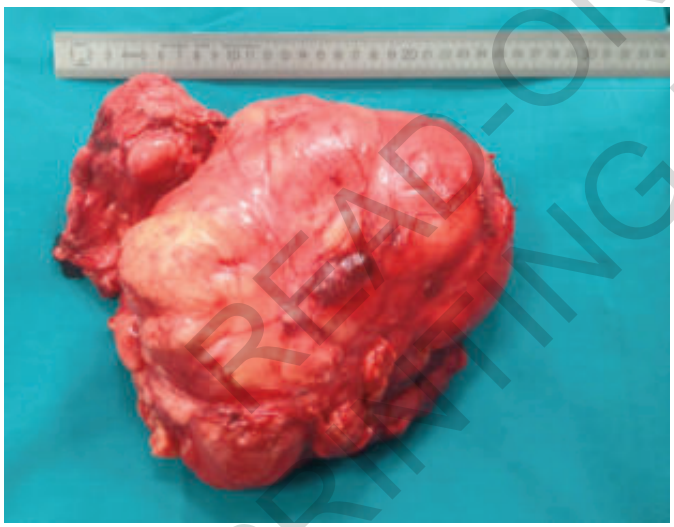

Fig. 3: Macroscopic findings of the tumor. The longest diameter is 26 cm.

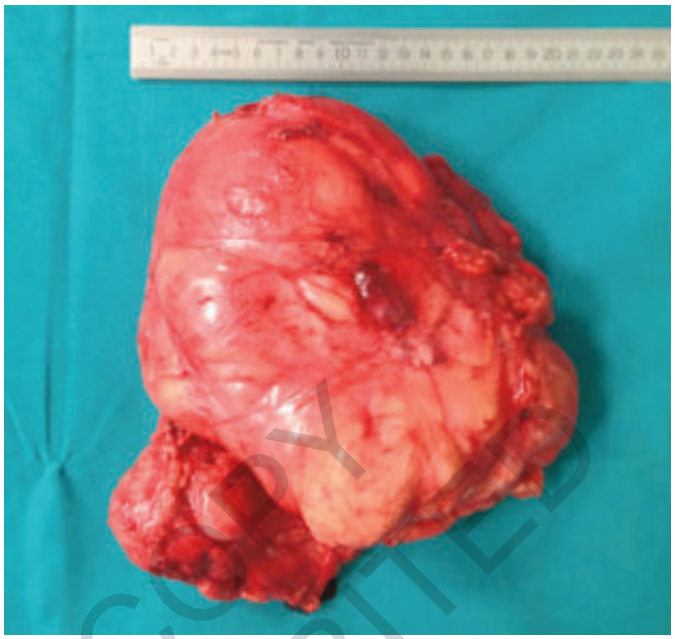

Fig. 4: Macroscopic findings of the tumor. The shortest diameter is 22 cm.

Discussion

Soft-tissue sarcomas represent an heterogeneous group of rare neoplasms that develop from the embryonic mesoderm. Among these, Liposarcoma is the most frequent type that develops in retroperitoneum and the most common symptom of his presence is abdominal distension. Many patients are asymptomatic until the tumor reaches large dimension, because intra-abdominal space is ample and neoplasms can grow without compromising gastrointestinal, urinary or vascular structures. In fact even if the diameter of the tumor is more than 20 cm, only a few patient are symptomatic. This limits the possibility to perform a curative resection. The literature shows that the prognosis for patients with retroperitoneal sarcoma is poor, with a 36% to 58% overall 5-year survival rate [5]. The natural history of these tumors is also characterized by locoregional recurrence that represents an important cause of death, since only 28% of patients don't show any recurrence within the first 5 years [5,6]. Chemotherapy is non effective and radiotherapy is limited by toxicity to adjacent intraabdominal structures [7]. The only potentially curative treatment is represented by the complete surgical resection [3,5,8-11]. From an histological point of view, according to the World health Organization (WHO) classification of soft-tissue sarcomas published in 2002, we can divide liposarcomas into 5 subgroups: well-differentiated liposarcoma, dedifferentiated liposarcoma, mixoid liposarcoma, round cell liposarcoma and mixed-type liposarcoma[12].

Dedifferantieted liposarcoma is an entity classified by Evans in 1979 like a sarcoma histologically characterized by the coexistence of well-differentiated liposarcoma and poorly differentiated non lipogenic sarcoma [13].

Dedifferentiation occurs in up to 10% of well differentiated liposarcoma, and can be histologically demonstrated by the transition to a nonlipogenic sarcoma. About 90% of dedifferentiated liposarcomas arise ex

novo, while in 10% of cases they appear as recurrence, both are characterized by an high risk of recurrence after surgery. Dedifferentiated areas may have different histological characteristics: most of the time they resembles malignant fibrous histiocytoma like high grade pleomorphic sarcoma or an intermediate- high grade myxofibrosarcoma. On the other hand other differentiations, like rhabdomyosarcoma,leiomyosarcoma and in particular osteosarcoma, are rare: in literature are reported in less than 5% of dedifferentiated liposarcomas [14]. In June 2013 Taishi Fujii et al., reviewing the literature, had found only 9 publications (13 cases) that described dedifferentiated liposarcomas with osteosarcomatous components [15]. Considering the case presented by the Japanese group and our patient the cases become 15. Focusing our particular attention on this group we can differentiate 8 male and 6 female (in one case patient's sex is not specified), whose mean age is 60.7 year. The most common places of occurrence are retroperitoneum (7 cases) and tight (4 cases); in 7 cases tumor arises primary, 2 cases were recurrences (in the other 6 cases the origin is not reported). Moreover we can say that, reviewing literature, we didn't found any publication that reports an association between malignant pleural mesothelioma and liposarcoma.

We propose a strictly follow up to our patient with a TC abdominal examination after 6 months from surgery.

Riassunto

Si riferisce del caso di un gigantesco liposarcoma retroperitoneale sdifferenziato con componente osteosarcomatosa. Questo tumore è stato asportato da una donna di 63 anni sostanzialmente asintomatica.

L'esame istopatologico ha dimostrato una prima componente neoplastica differenziata ed una seconda componente neoplastica sdifferenziata con prevalenti aspetti di osteosarcoma.

A nostra conoscenza in letteratura sono stati riportati soltanto altri 14 casi del genere.

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