Giant liposarcoma of the mesentery.
Report of a case

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Guido Cerullo, Daniele Marrelli, Bernardino Rampone, Ester Perrotta, Stefano Caruso, Franco Roviello

Department of General Surgery and Surgical Oncology, U.O. Surgical Oncology, University of Siena, Siena, Italy

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Liposarcomas represent the single most common type of soft tissue sarcoma. Its abdominal localization is rare, occurring in only 5% of cases. A 55 year old male was found to have a case of primary giant liposarcoma of the mesenterium with a maximum diameter of 40 cm and weight of 9 kg. Computed tomography revealed the presence of a large mass presenting a dishomogeneous density with an adipose component, probably of mesenteric origin. A xifopubic laparotomy confirmed the presence of a pedunculated growth originating from the mesentery. The mass was removed and the histopathological report noted a well differentiated sclerosing liposarcoma with the peritoneal liquid positive for malignant cells. Surgery currently represents the only possibly curative therapy for this type of tumour but close long-term follow up and accurate evaluation of the clinicopathologic parameters are needed.

KEY WORDS: Liposarcoma, Soft-tissue tumours, Tumours of the mesenterium.

Introduction

Soft-tissue sarcoma is an unusual tumour, representing less than 1% of all malignancies. Liposarcomas represent 20-30% of adult soft-tissue tumours [1-4]. Abdominal localization of liposarcomas (retroperitoneal, pelvic and visceral) represents 10-15% of all liposarcoma [5] and retroperitoneal forms are much more common than those deriving from the mesentery or mesocolon [1,6].
Liposarcomas are most common in the fifth to sixth decade of life and in males [7-8]. Its principal histological subtypes (well differentiated, myxoid and pleomorphic) are entirely separate disease with different morphology, genetics and natural history [9,10]. Although these neoplasias grow to conspicuous proportions [1,11-15], diagnosis often occurs at a late stage and by chance. Radical surgical excision is both of fundamental therapeutic importance and necessary for correct histologic diagnosis. We report a rare case of giant liposarcoma of the mesenterium that was observed and operated on at our Institute.

Case report

A 55 year old male presented at our Institute reporting abdominal distension associated with a gradual weight loss (3 kg) over the previous sixth months. Relevant patient history was a surgical operation to remove a lipoma (19x7 cm) from the right thigh 3 years previously. On physical examination, superficial palpation of the abdomen revealed the presence of a coarse non-painful growth in the mesogastric area. Routine blood tests, electrocardiogram and chest x-rays were normal, except for a slight increase in the total bilirubin (1.2 mg/dl) and direct bilirubin (0.3 mg/dl) and lipases (84 IU/l). Tumour markers (CEA, CA 125, CA 15-3, CA 19-9, CA 72-4 and AFP) were within normal limits. A Computed Tomography (CT) scan of the abdomen showed the presence of a large expanding growth, probably of mesenteric origin, with a transverse diameter of 29.8 cm (Fig. 1). On examination, the mass presented

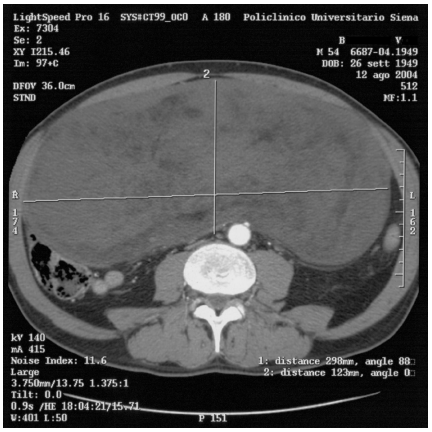

Fig. 1: Abdominal CT: images of a voluminous abdominal mass with adipose component originating from the mesentery, but not infiltrating surrounding structures.

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For correspondence: Dr. Guido Cerullo, Via Casalpiano 79, 53018 Sovicille (SI), Italy (e-mail: drcerullo@tiscali.it).

a dishomogeneous density with an adipose component that was not particularly vascularized but was well defined and showed no signs of infiltration of the surrounding structures. There was a minimal amount of ascites in the Douglas' space. The patient was therefore operated on: following a xifopubic laparotomy, exploration of the abdominal cavity confirmed the presence of a pedunculated growth originating from the mesentery. The growth was capsulated, with distinct borders and a tight elastic consistency. The vascular peduncle was isolated, tied and sectioned, then the growth was carefully excised. The mass that was removed had a maximum diameter of 40 cm and weighed 9 kg (Fig. 2). The histopathological report noted a well differentiated sclerosing liposarcoma (G1); the histological diagnosis was confirmed by electron microscopy. The peritoneal liquid showed the presence of numerous mesothelial cells and occasional cellular elements with cytological characteristics of malignancy. Recovery was uneventful and the patient was discharged six days after the operation. After 1-years' follow-up (clinical and radiological) he is healthy.

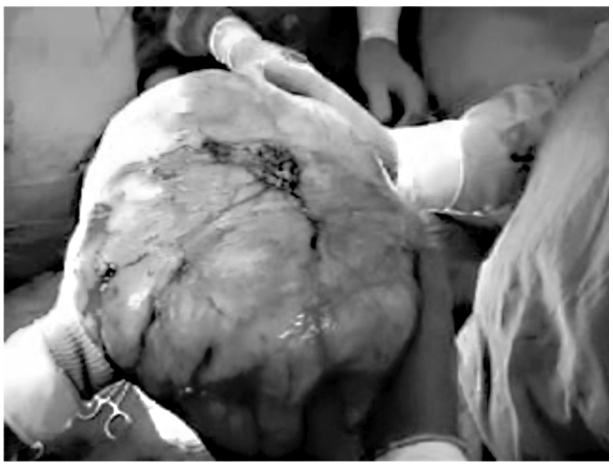

Fig. 2: Excised abdominal mass. Maximum diameter of 40 cm and weight of 9 kg.

Discussion

Liposarcomas of the mesentery are extremely rare neoplasias more than the localization of the extremities or retroperitoneum, as can be seen from international cases studies. They become clinically evident at a late stage, when they have already reached large proportions, and as abdominal masses they pose differential diagnosis with many other pathologies. Among imaging methods, CT is useful for diagnosis and is necessary both for the confirmation of clinical suspicions and for an initial evaluation of the anatomical relationship of the mass with the organs and abdominal structures. In our case CT highlighted the mesenteric origin of the neoplastic mass and its dimension, which was much larger than many cases reported in the literature. CT is therefore fundamental in the first step of the process of differential diagnosis, inasmuch as it can identify the different density of the liposarcoma's adipose component and, where possible, indicates its anatomical origin. However, a definitive diagnosis is only possible in the operating theatre [1] and, considering the great controversy regarding the real benefits of adjuvant therapies, surgery currently represents the only possibly curative therapy for this type of tumour [1, 4-7, 8, 16-20]. In fact, adjuvant treatment for selected cases of extremities liposarcoma is widely accepted as a standard, while it is still investigational for retroperitoneal/abdominal liposarcoma. Some authors maintain that neither radiotherapy nor adjuvant chemotherapy can bring about any real benefit in terms of long term survival [21]. Others have tried to set down guidelines for adjuvant therapy of liposarcomas, including: age under 50, high grade tumour and incomplete resection margins [22]. Anyway, a metaanalysis of adijuvant chemotherapy for Soft-Tissue Sarcomas did not demonstrate an overall survival advantage, although progression-free survival improved [26]. Although radical, surgical excision should not be considered as a definitive cure for these neoplasias. In fact, localised recurrence of the disease occurs extremely frequently, even after a lapse of 10 years, thus rendering careful follow up necessary for at least 5 years [4]. The real risk of formation of lymphnode or multi-organ metastases must be evaluated on the basis of the histopathologic report because it is closely related to the different histological type of liposarcoma [23]. The survival rate for abdominal presentations in international cases is 30-40% [2, 6, 14, 15] but the prognosis is linked to various parameters: histological type, degree of differentiation, tumour site and the radicality of surgical excision [1, 4, 6]. However, the most relevant prognostic factor seems to be the size of the mass [1, 4, 6, 11], with a cut-off point at 10 cm [6, 24] or 20 cm [11] in diameter, according to the cases studied. In our case, although the tumour was well differentiated, the prognosis for the patient could not be positive due to the large size of the neoplastic mass, which was well over the 10-20cm in diameter considered as a useful cut-off point to distinguish between cases with good or bad prognosis [6, 11, 24]. Furthermore, cytologic analysis of the peritoneal liquid was also positive for malignant cells. This parameter is cited in few references [25] of the international literature on liposarcomas but may be a useful prognostic factor and provide additional information for the identification of patients at greater risk of local recurrence. Even if cytology has a limited role for diagnosis of soft tissue sarcoma, further random clinical studies with large numbers of cases may be useful for the evaluation of the risk and time scale of local recurrence in relation to the size of the neoplasia and the cytologic results on peritoneal liquid. In conclusion, abdominal liposarcomas are rare but require radical excision, close long-term follow up of the patient and accurate evaluation of the clinicopathologic parameters.

Riassunto

I Liposarcomi a localizzazione addominale rappresentano un entità patologica rara. Presentiamo il caso di un uomo di 55 anni portatore di una massa endoaddominale, confermata con l'ausilio dell'esame CT scan. L'asportazione chirurgica della neoformazione e l'esame istologico hanno permesso di giungere alla diagnosi di liposarcoma sclerosante ben differenziato. Attualmente, infatti, la chirurgia rappresenta l'unico atto diagnostico e terapeutico per questo tipo di tumori ed uno stretto follow-up si rende necessario per una accurata valutazione della ripresa locale di malatti

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