Re-do Surgery for Recurrent Giant Abdominal Liposarcoma: Description of a Case Report and Literature Review

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Abstract

Introduction: Soft tissue sarcomas are rare and among them giant liposarcoma represents a very uncommon disease.

Case Report: We report a case of a 64 year-old man affected by dedifferentiated retroperitoneal liposarcoma that underwent surgery in 2014. The postoperative course was uneventful and adjuvant chemotherapy was given. Two years later, he was referred to our department because of changing in bowel habits, and nausea. CT scan revealed a huge abdominal mass infiltrating left colon and ileum. Radical re-surgery with colonic and ileal resection was performed.

Conclusion: Liposarcoma and giant liposarcomas are rare. A multidisciplinary approach and surgery are suggested even in recurrences.

Introduction

Liposarcoma is an uncommon disease; its incidence was reported in up to 45% of all soft tissue sarcomas [1]. Tumor size is highly variable and huge dimensions can be reached [2]. Location is inconstant since from neck to gastrointestinal tract, different organs can be involved. The retroperitoneum is the most frequent site, representing 10-15 % of all cases. Diagnosis is often reached in advanced stage because symptoms are unspecific and mostly related to its localization. Surgery represents the treatment of choice in almost all patients. Complete tumor removal with wide margins including, if needed, multivisceral resections is recommended [3]. A case of a 64 year-old patient with recurrent giant liposarcoma is described. Literature review focusing on the topic of re-do surgery in abdominal liposarcoma surgery has been performed too.

Case Presentation

A 64 year-old man underwent surgery for a retroperitoneal dedifferentiated liposarcoma followed by Epirubicin, Ifosfamide and Trebectedine adjuvant chemotherapy in 2014. The treatment was interrupted because of hepatic toxicity after two weeks. In 2016 a new Ifosfamide chemotherapy, interrupted because of clinical worsening, was started. Indeed, he referred to our department because changing in bowel habits, fever and abdominal pain. Abdominal physical examination showed a mass in the medium quadrants of abdomen. All laboratory tests were within normal ranges. CT scan revealed a huge retroperitoneal mass with sigmoid perforation; a conservative approach with fluids and antibiotics was chosen. Two weeks later, a CT scan confirmed multiple tumors localized in both peritoneal and retroperitoneal space. Small bowel was infiltrated as sigmoid colon; the left kidney was dislocated (Figure1-4). The largest mass (30 cm) was located in the pelvis. The patient underwent relaparotomy: multiple masses occupying the abdomino-pelvic cavity and the retroperitoneal space were detected. All the masses were removed with macroscopic free margins (Figure 5 and 6); two segmental small bowel resections with side-to-side mechanic anastomosis were performed. Sigmoid colon was resected too and colo-colic side-to-side manual anastomosis was performed.

Histopathological examination revealed a dedifferentiated liposarcoma (DDLPS). In the immediate aftermath of the operation, the patient was transferred to the intensive care unit for invasive monitoring. On the second postoperative day, because of abdominal pain and fever, he underwent an abdominal CT scan that showed a fluid collection between intestinal loops and subfrenic air; moreover enteric material from abdominal drains was observed. Therefore, the patient...
underwent ileal anastomotic resection and protective ileostomy because of anastomotic dehiscence. The rest of postoperative course was uneventful and patient was discharged in 10th postoperative day.

**Discussion**

Sarcomas are a heterogeneous group of mesenchymal tumors with an uncertain etiology. According to the World Health Organization classification of soft tissue tumors, 5 categories of liposarcomas are reported: (1) well differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell; and (5) pleomorphic [4]. These tumors affect both women and men aged between 40 and 60 years [5]. Tumor size is highly variable, ranging from few kilos to huge dimensions: those over 20 kg are called “giant liposarcoma”, as in our case, and they are extremely rare. Liposarcoma can affect the whole body from head, neck, trunk, mediastinum, upper and lower extremities, the gastrointestinal tract, and retroperitoneum [2]. In presence of retroperitoneal localization, the tumor can grow and reach huge dimensions with late symptoms and worse prognosis [6]. Abdominal pain, changing in bowel habits, dyspepsia, weight loss and anemia are the most common clinical presentations. As reported in our case, the most characteristic sign is a painless abdominal mass that according to literature is detected in up to 78% of cases [7]. Abdominal symptomatology is due to compression of the adjacent organs [1].

Diagnosis is often late and requires different examinations: ultrasonography, computed tomography scan (CT) and MRI. They provide information about localization, size, tumor extension and its relationship with the adjacent organs. Ultrasonography is usually the first choice in patients referring to the hospital for abdominal pain, even if its value is limited in obese and in those with an abdominal girth. CT scan and MRI are the gold standard for diagnosis, and they are highly sensitive and specific in differencing intra abdominal and retroperitoneal mass, necrotic areas, organ invasions. Biopsy is not required, because as reported by Chew, it adds no value to clinical and radiologic assessment of the patient with resectable large
retroperitoneal mass [8]. In our case we didn’t perform any biopsy also because considering the history of the patient, it was highly suspected to be a recurrence. Surgery represents the gold standard in treatment of non-metastatic retroperitoneal liposarcoma [9]. If required, multiorgan resections can be performed with the aim of a wide free margin around the tumor to prevent recurrence. Although some patients can have metastatic disease (usually lungs), mostly have recurrence in the primary site [10]. Indeed, locoregional recurrence remains the main problem, whose rates ranging from 50% to 80% [11]. Surgery remains the only curative treatment in recurrent liposarcoma too [1,7,12]. Even with complete removal of the liposarcoma, prognosis remains poor. The 5-years survival rate of well-differentiated retroperitoneal liposarcoma is 83%, while it is 20% for the dedifferentiated tumor subtype [13]. Although this poor prognostic rate, Bautista showed that meaningful long-term survival could be achieved with reoperations for recurrent disease [14], and moreover when repeated surgery can be successfully performed [15]. Recurrent sarcoma shows the same patterns of primary ones: they are asymptomatic until they reach huge dimensions, as reported in our case; radiological close follow up represents a useful tool to early detect these masses. As for primary tumors, local recurrence survival after re-surgery depends on the grade and the size of the tumor [16]. Lahat [17] demonstrated that patients with tumors sized >15 cm were at increased risk of developing distant recurrence and exhibited higher disease-specific mortality compared with those with smaller tumors. Surgery in combination with radiotherapy can prevent recurrence in about 85-90% of liposarcomas, when compared with surgery alone [18,19]. The role of chemotherapy is not clearly defined, but it may be recommended in patients at high risk of recurrence or in presence of metastatic disease [20].

References