Journal of Gastrointestinal Surgery (2019) 23:1521–1523
https://doi.org/10.1007/s11605-018-3851-5

GI IMAGE

Giant Primary Retroperitoneal Dedifferentiated Liposarcoma

Fabio Carboni[1] · Mario Valle[1] · Orietta Federici[1] · Renato Covello[2] · Alfredo Garofalo[1]

Received: 5 June 2018 / Accepted: 18 June 2018 / Published online: 29 June 2018

Keywords Dedifferentiated liposarcoma · Giant liposarcoma · Retroperitoneum · Diagnosis · Surgery

Clinical Data

A 65-year-old woman presented with vomiting, pain, and significant abdominal distension. Physical examination demonstrated a painful large palpable mass involving the whole abdomen (Fig. 1a). Laboratory investigations showed low hemoglobin (7.8 g/dl) and protein (5.2 g/dl) level. Computed tomography (CT) scan showed a 34 × 32 × 35-cm heterogeneous fatty mass with septations and solid nodules within the lesion showing high contrast enhancement, filling the entire abdominal cavity, with right kidney encasement (Fig. 2a). Abdominal visceral displacement was observed (Fig. 2b). Preoperative biopsy showed lipomatous tumor. At laparotomy, the well-circumscribed, lobulated mass, weighing about 30 kg underwent complete excision, including a right nephrectomy (Fig. 1b), and the greatest difficulty was to obtain vascular control of major vessels. Microscopically, the tumor was characterized by the presence of pleomorphic lipoblasts with large, hyperchromatic nuclei scalloped by cytoplasmatic vacuoles (Fig. 3a). Immunohistochemical analysis was positive for CDK4 and MDM2 (Fig. 3b). Diagnosis was dedifferentiated liposarcoma grade III. Oozing hemorrhage without overt source requiring relaparotomy occurred in postoperative day 5. The patient was discharged 10 days later. She was lost at follow-up after 12 months without evidence of recurrence.

✉ Fabio Carboni
fabiocarb@tiscali.it

[1] Department of Digestive Surgery, IRCCS, Regina Elena National Cancer Institute, Via Elio Chianesi 53, 00144 Rome, Italy

[2] Department of Pathology, IRCCS, Regina Elena National Cancer Institute, Rome, Italy

Discussion

Soft tissue sarcomas represent the rarest form of solid tumor malignancy, accounting for only 1% of all tumors in the adult population and including more than 50 subtypes. Approximately 12–15% develop in the retroperitoneum (RP) with liposarcomas (LS) representing almost 70% of cases.[1] These are a heterogeneous group with a varying biological and clinical behavior. According to the last World Health Organization classification, four major subtypes can be recognized: well-differentiated (WDL), dedifferentiated (DDL), myxoid, and pleomorphic.[1] Dedifferentiated subtype represents about one third, arising de novo in most cases, while few occur following recurrences of WDL. It consists of well-differentiated lipogenic mass juxtaposed to non-lipogenic sarcoma, of variable histologic grades. DDL shows more complex genetic abnormalities as compared to WDL mainly involving chromosome 12q13-15, with high amplification levels of MDM2 (100%) and CDK4 (90%) oncogenes, correlating also with poor outcome.[1]

Owing to the absence of symptoms in the RP space, LS may reach a large size by the time of diagnosis, often causing symptoms of compression on adjacent organs. Those weighing over 20 kg are extremely rare and considered to be "giant" LS. A recent review identified only 13 cases reported in the literature, with DDLs accounting for less than one third.[2] Contrast-enhanced CT scan is the imaging technique of choice to evaluate these tumors. Differential diagnosis with WDL is based on the presence of the non-lipomatous component with DDL usually appearing as heterogeneous fatty mass with septations and solid nodules within the lesion, showing high contrast enhancement.[2] However, with the exception of WDL and angiomyolipoma, identification of subtypes may be challenging. Percutaneous core needle biopsies can be safely performed to establish a correct diagnosis and the most appropriate treatment when radiologic appearance is unclear or

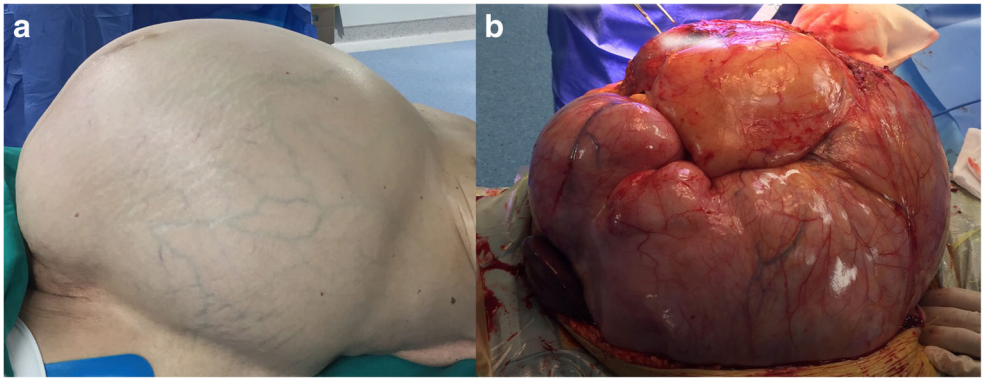

Fig. 1 **a** Palpable mass involving the whole abdominal cavity. **b** Macroscopically well-circumscribed, lobulated mass

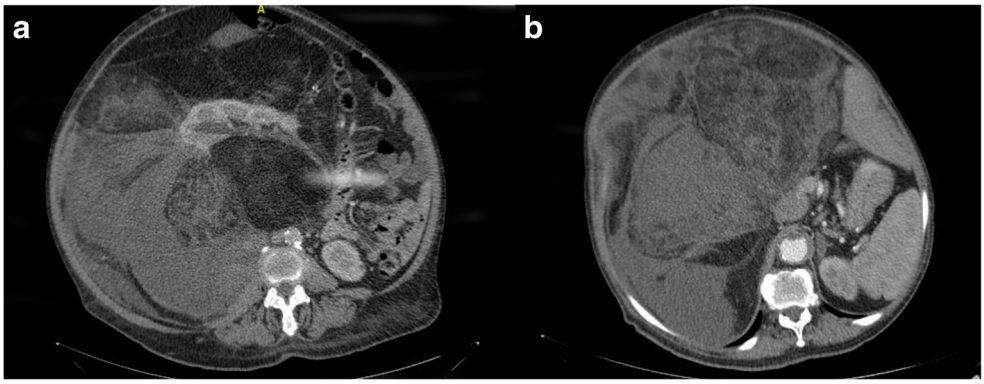

Fig. 2 **a** CT scan showing heterogeneous fatty mass with septations and solid nodules within the lesion with high contrast enhancement, encasing the right kidney. **b** Abdominal organs appear displaced

neoadjuvant treatment is planned, but overall accuracy is low for DDL.[2]

Complete surgical resection is the mainstay of treatment, but the large size and deep location make resectability difficult. The most important prognostic factors are grading and degree of resection. A radical resection should be obtained, often requiring en bloc removal of involved adjacent structures. Tumor integrity, extent of resection, gradinge, multifocality, and organ invasion impact patient's outcome.[3] Compartmental resection of uninvolved structures is associated with no survival benefit and increased postoperative morbidity and mortality. Despite aggressive surgery, mortality is predominantly due to local failure, and histologic subtype of DDL is a significant predictor of local recurrences in up to 40–80% of patients.[3] Distant metastases have been observed in half of cases in grade III disease, mostly occurring in the lung. Incomplete, piecemeal resection is associated with a dismal prognosis and should be avoided unless for symptomatic reasons.

Postoperative radiotherapy may constitute a valuable option in the presence of involved margins or high-grade tumors, but no studies have proven the efficacy following gross

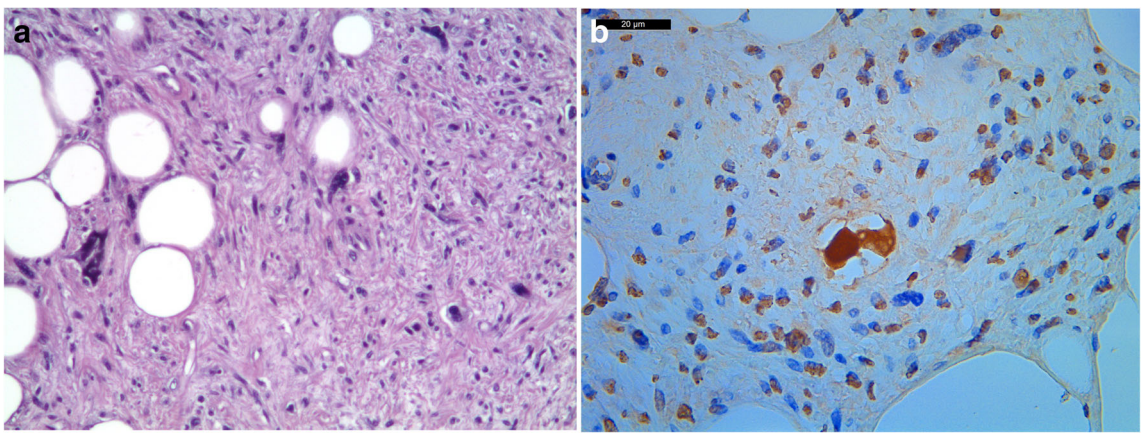

Fig. 3 **a** Areas of undifferentiated pleomorphic sarcoma and areas containing lipoblasts (H&E, × 10). **b** Immunohistochemical analysis showing intense nuclear staining for MDM2 (× 200)

resection. Preoperative radiotherapy produced higher rates of negative surgical margins and improved local control, but no survival benefit was obtained compared to surgery alone.[2] Systemic chemotherapy showed poor response rate in DDL, and it is reserved for palliative treatment of advanced or metastatic disease.

Conclusions

Novel molecular targets are necessary to provide new treatment options in the future. At the moment, the main factor improving patient's prognosis is adequate initial treatment in specialized multidisciplinary centers, including aggressive surgery.

Author Contribution FC, MV, OF, RC, and AG participated in the:

- Substantial contributions to the conception or design of the work, or the acquisition, analysis, or interpretation of data for the work;
- Drafting the work or revising it critically for important intellectual content;
- Final approval of the version to be published;
- Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

References

1. Thway K, Jones RL, Noujaim J, Zaidi S, Miah AB, Fisher C. Dedifferentiated liposarcoma: updates on morphology, genetics and therapeutic strategies. Adv Anat Pathol 2015; 23: 30–40
2. Zeng K, Liu W, Wu X et al. Clinicopathological characteristics and experience in the treatment of giant retroperitoneal liposarcoma: a case report and review of the literature. Cancer Biol Ther 2017; 18: 660–5
3. Keung EZ, Hornick JL, Bertagnolli MM, Baldini EH, Raut CP. Predictors of outcome in patients with primary retroperitoneal dedifferentiated liposarcoma undergoing surgery. J Am Coll Surg 2014; 218: 206–17