Dedifferentiated liposarcoma of the retroperitoneum

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Abstract

Retroperitoneal liposarcoma is a rare tumor in clinical practice, that is difficult to diagnose and treat. We present the case of a 64 years old female patient diagnosed by computed tomography imaging with a massive left renal tumor, after the emergence of an autopalpable mass within the abdominal left flank. The CT showed a large left renal mass without a clear limit from the adiacent structures (pancreas, spleen, colon). After a correct and complete surgical resection, a retroperitoneal dedifferentiated liposarcoma was histopathologicaly diagnosed, witch encapsulated the left kidney and adrenal gland without microscopic invasion of these structures.

Keywords: retroperitoneal liposarcoma, CT imaging, surgical resection

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Introduction

Liposarcoma is the most common malignant tumor with origin in the connective tissue of the retroperitoneum. Based on morphological and cytogenetic aberrations, liposarcoma are classified into four histologic subtypes: 1- well differentiated, 2 - dedifferentiated, 3 – myxoid / round cell and 4 – pleomorphic (1).

Dedifferentiated subtype, in witch in the same tumor there are findings of well differentiated liposarcoma, undifferentiated liposarcoma and non-lipomatous sarcoma items (2), has several features: histological diagnosis is difficult and it has a more reserved prognosis compared with other subtypes (5-year survival of approximately 20%, local recurrence in 40-83% of cases and distant metastases in 15-30% of cases) (3, 4, 5) (fig.1).

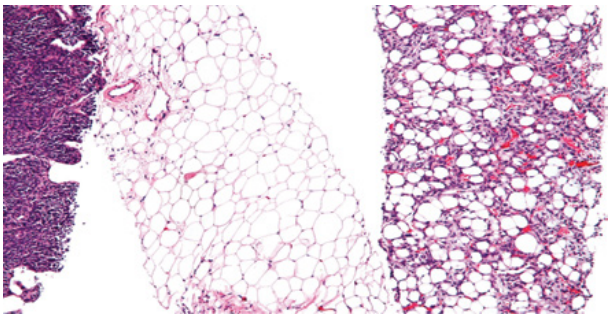


Fig.1 Histopathological aspect

Radical excision with a safety margin is the standard surgical procedure that ensures the greatest long-term survival. Adjuvant techniques as chemotherapy and radiotherapy, still do not have a well established role in the treatment of this tumor, so they are being reserved for cases in which complete surgical excision was not possible (6, 7).

Case presentation

The patient aged 64 years with no significant medical history, recently diagnosed with a left renal tumor (based on the diagnosis of abdominal and pelvic CT-fig.2) addresses our clinic for specialized treatment. Clinically, the patient had left abdominal enlargement with pain and nausea. Physical examination revealed normal vital signs and solid, large, palpable abdominal mass in the left flank. In terms of biological status: persistent elevated creatinine levels specific for the pre-uremic stage of chronic renal failure (creatinine = 1.44mg/dL), slight hypoproteinemia with hypoalbuminemia (total protein = 5.9g/dL, albumin = 3g/dL), increased GGT and LDH (GGT = 163U/L, LDH = 443U/L).

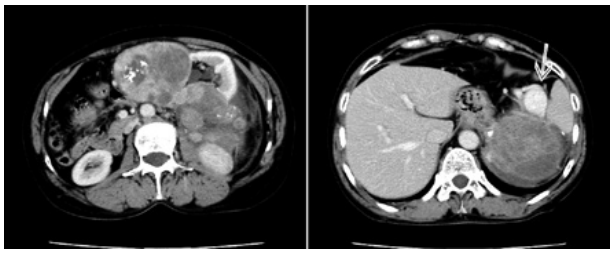


Fig.2 (right) Accessory spleen – preoperatory aspect; (left) Tumor - preoperatory aspect

The patient underwent left radical nephrectomy by anterior, transperitoneal, approach, with preservation of the adjacent organs (accessory spleen was identified during the surgery, and preserved). Bulky tumor mass of larger diameter of 26 cm was excised (fig.3).



Fig.3 (right) Macroscopic aspect – excised tumor; (left) Dissected aspect

Macroscopically the tumor was solid pseudo capsulated with poly cyclic shape, multilobulat polimorfe appearance (some whitish, some pink-yellow and/or yellowish-translucent tumor lobules, with variable consistency, mostly solid) and it encapsulated almost entirely the left kidney and left adrenal gland. Microscopic examination revealed elongated malignant mesenchymal tumor cell proliferation, dedifferentiated liposarcoma type in multi nodular pattern, sometimes intimately adherent to the renal capsule, without invasion of the parenchyma, and incorporating the adrenal gland.

Postoperative evolution was favorable, except for the development of a left lumbar lymphocele that required drainage. Revalued to 2 months, the patient was asymptomatic and computer tomography examination showed complete excision of retroperitoneal tumor, with no evidence of local recurrence (fig.4).

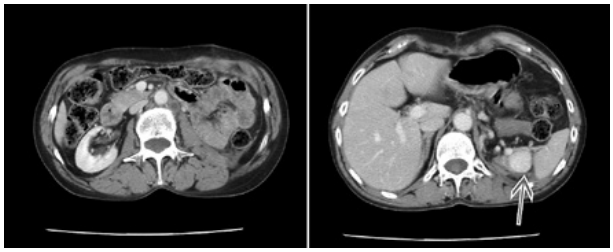


Fig. 4 (right) Accessory spleen – postoperatory aspect; (left) Left flank without tumor recurrence, occupied by the intestines.

Discussions

Dedifferentiated liposarcoma of the retroperitoneum is a rare form of cancer, but has a very high mortality rate in the absence of surgical treatment. It can occur at any age, but the highest incidence is recorded in the fifth and sixth decade, especially in men. In most cases (80-90%), the tumor appears de novo, with no prior injury.

The presence of palpable tumor is the main clinical sign of soft tissue sarcomas, constituting, as a rule, the reason for addressing the doctor. In large tumors, clinical signs are secondary to compressive effect exerted on the surrounding structures.

Imaging studies (computed-tomography, MRI) do not provide information that can indicate the presumed histogenesis tumor process, but are essential for assessing the exact location, size, extent of tumor and involvement of adjacent structures (lymph nodes, vessels, nerves, bones). Tumor biopsy with appropriate imaging studies is not recommended for two main reasons: "blind" biopsy may be uninformative, and may jeopardize the possibility of a future radical surgical excision.

The only curative treatment that can provide long-term survival is complete surgical excision of the tumor. This goal is not always possible for the following reasons: most of the tumors were large at the time of diagnosis (often > 10 cm); retroperitoneal space doesn't have a very clear anatomical partition, so that tumors extend easily and include various structures and organs; intraoperative difficulty of establishing a clear boundary for separating the high differentiated adipocytes of the liposarcoma structure and retroperitoneal adipose tissue.

Soft tissue sarcomas tend to develop along the fascial tissue in areas with the lowest resistance. As a result, the surrounding soft tissues are compressed and form a pseudocapsule, which gives the appearance of a tumor sarcoma well encapsulated. This aspect is often misleading, because "the enucleation" or marginal excision of the tumor lead to a probability between 50 and 90% of the local recurrence.

Prognostic factors that are associated with high rates of morbidity and mortality in case of a liposarcoma are: anatomic setting (retroperitoneal liposarcoma has lower survival rates compared to other sites), histology (dedifferentiated liposarcoma shows high risk of local recurrence and distant metastasis) and incomplete surgical excision (8, 9, 10).

Conclusions

Retroperitoneal liposarcoma is a challenge in practice, because it is a rare tumor, but with an aggressive development and high mortality, so early diagnosis, histopathological staging and correct and complete surgical excision of the tumor is the key to getting a good, long term survival.

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