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clinical practice

Surgical treatment for giant retroperitoneal well-differentiated liposarcoma (WDLPS): case report and literature review

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SUMMARY: **Surgical treatment for giant retroperitoneal well-differentiated liposarcoma (WDLPS): case report and literature review .**

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WDLPS are very rare retroperioneal tumors that can reach huge size before becoming symptomatic. The aim of this article is show the diagnostic management and the surgical approach to giant WDLPS.

A standard treatment has yet to be established because the preoperative diagnosis is very difficult.

We present a case of a 69-year-old male patient with progressive increase of abdominal girth, weight loss and light abdominal pain and with an abdominal mass of over 15 kg that displaced the right kidney.

A complete tumor resection and right nephrectomy were performed. Histology revealed a well-differentieated liposarcoma.

CT scan is the most common imaging technique and laparotomic open radical resection represent the most common surgical approach.

KEY WORDS: Surgery - Retroperitoneal - Tumor - Liposarcoma - WDLPS.

Introduction

Soft tissue sarcomas (STS) are rare tumors, they account for <1% of all malignant tumors (1, 2). Liposarcoma (also known as atypical lipomatous tumor) (3) is the most common (4) and represent the 20% of all STS (1, 2). They are tumors of embryogenic mesodermal origin presenting in the following histologic variants: well-differentiated (WDLPS), myxoid, round cell, pleomorphic, and dedifferentiated (DDLPS) (5). Rarely located in the gastrointestinal tract, liposarcomas can develop in the extremities or trunk, but the retroperitoneum is the most common location (6, 7), in fact it is the primary site in about 15% of STS (8, 9).

The incidence of retroperitoneal liposarcomas is 0,3% to 0,4% per 100,000 individuals (7). The two most common retroperitoneal types are well differentiated (WDLPS) and dedifferentiated (DDLPS). These two variants share the molecular hallmark of MDM2 gene amplification which differentiates these from other retroperitoneal tumors (10). Retroperitoneal liposarcomas have a higher incidence in the 6th and 7th decade, they present with non-specific symptoms like bleeding, weight loss, and abdominal pain (11). Usually, retroperitoneal liposarcomas reach huge sizes because they can grow without constraints before becoming symptomatic (10, 12).

Tumor behaviour depends on the liposarcoma subtype. WDLPS is locally aggressive but does not metastasize, whereas DDLPS has the potential to metastasize (20-30% distant recurrence rate). DDLPS also has a higher local recurrence rate than WDLPS and six times the risk of death (13, 14).

The rarity of retroperitoneal liposarcomas and the variety of histologic subtypes make difficult the

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management of these neoplasms. Because of the large potential spaces of the retroperitoneum and local invasiveness of liposarcomas, these lesions are very large when are diagnosed and often involve adjacent organs and structures (11). Surgery is the first step in resectable disease, however local recurrence rate can be >80% in DDLPS (14). Studies that have evaluated preoperative chemotherapy followed by surgery have reported inconsistent findings (15).

We describe a case of a giant retroperitoneal well differentiated liposarcoma and discuss about two still open questions: the difficulty of a precise preoperative diagnosis and the surgical approach. This work is reported in line with the Surgical Case Report Guidelines (SCARE) criteria.

Case report

A 69-year-old male patient, presented a gradual increase of abdominal girth in the last months, weight loss and light diffuse abdominal pain, was admitted at our department for further diagnosis and treatment. No other symptoms were reported. Physical examination on admission revealed a big fixed abdominal mass with ill-defined margins occupying the entire abdomen. Pre-operative laboratory tests were normal. Tumor markers were negative. Abdominal US showed a very big abdominal mass of over 30 cm of diameter. Contrast enhanced CT showed a huge retroperinonel mass of 30 × 21 × 33 cm presenting a lipoma-like aspect with mixed density and pathological contrast enhancement. The mass displaced right kidney in epigastrium and pressing on it (Figures 1, 2). CT findings suggested a giant fatty tumor, including suspected dedifferentiated areas with diffuse calcification. Surgery was decided and a midline xyphoid to pubis incision was performed. Surgical exploration revealed a voluminous mass involving right kidney without a clear cleavage plane between the mass and the right kidney and "en-bloc" resection of tumor mass and right kidney was performed. The mass weight was of over 15 kg (Figure 3).

The recovery was uneventful: oral feeding restarted in 2nd p.o. day and normal bowel functions restored in 3rd p.o. day. Drains were removed in 8th p.o. day and the patient was discharged in 12th. Histological examination showed a well differentiated liposarcoma with 'floret celles' aspect with R0 margin of resection.

The immune-histochemical results confirmed the diagnosis (CD34; CD99; CD117; S-100; Desmin; ki-67; Vimentin; p53; p16).

There was no report of relapse on the follow-up survey after 6 months and after 1 year.

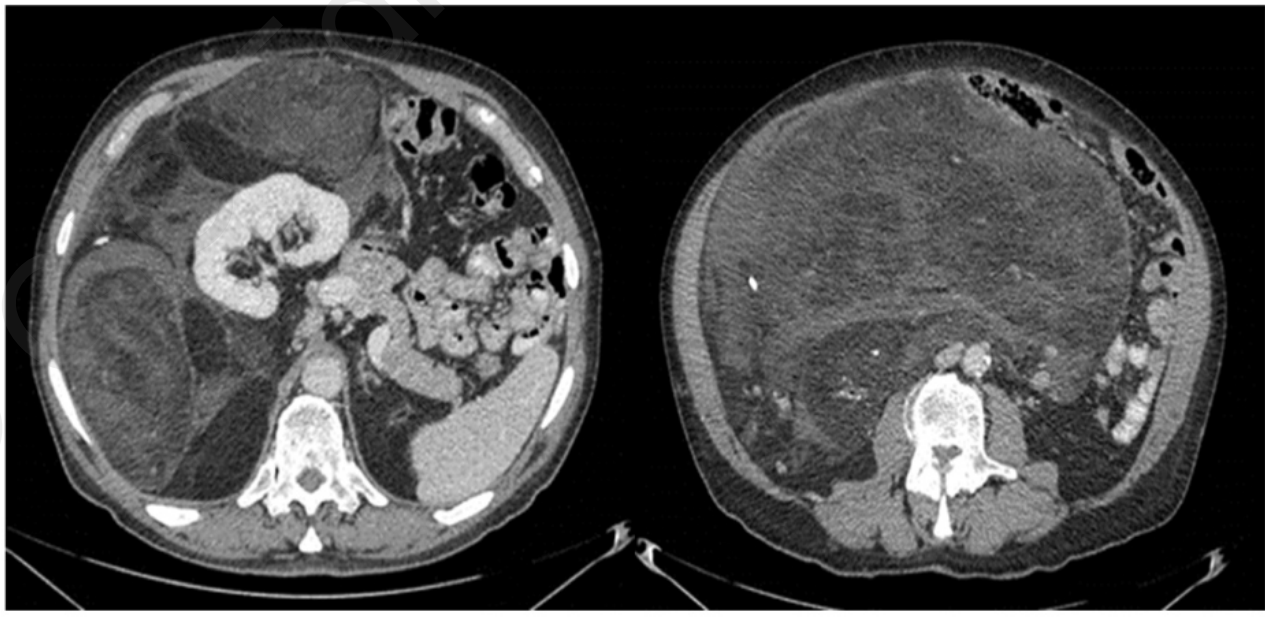

Figure 1 - CT scan: right kidney displaced in epigastrium.

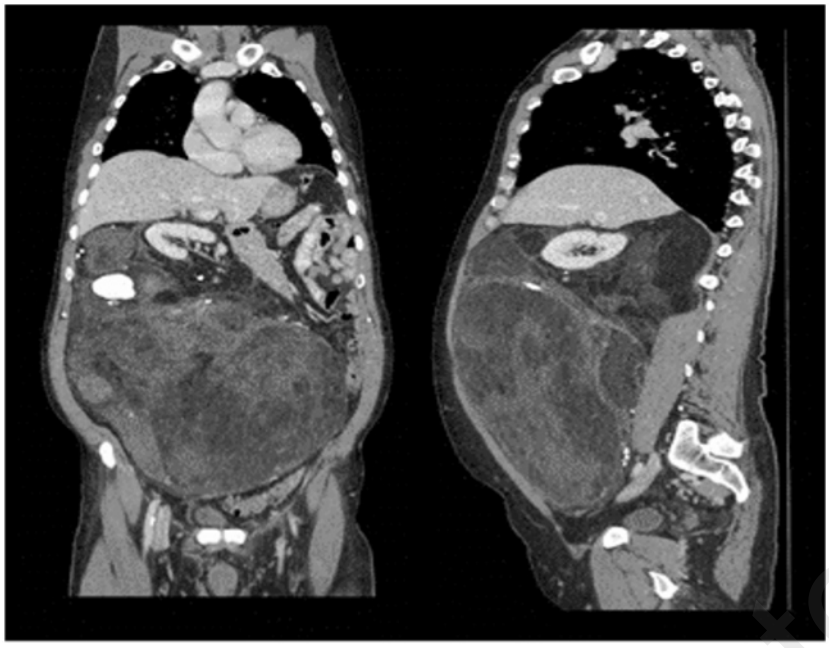

Figure 2 - The mass occupying the entire abdomen.

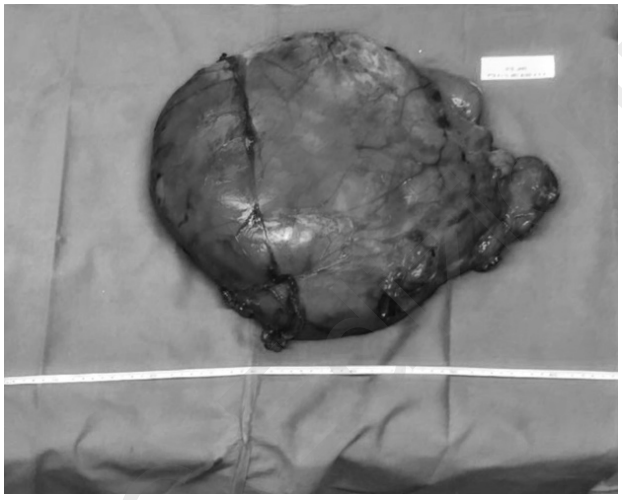

Figure 3 - Operative sample.

Discussion

Liposarcomas are the commonest type of retroperitoneal sarcomas (11, 16, 17). Generally, patients with sarcomas present late symptoms, since the retroperitoneal area provides a potential space for the growth of tumors (18). Most of these patients may remain asymptomatic or present nonspecific symptoms because the tumors grow without pressing on visceral organs (19). However, some patients with retroperitoneal sarcomas can present gastrointestinal symptoms when the tumor presses the gastrointestinal tract, while others can have urinary disorder, when the tumor involves the kidneys. Similarly, patients can present an increased frequency of micturition or urgency when the urinary bladder is affected, and pressure on the nerves can present as neural syndromes.

Performing Ultrasound is very difficult to differentiate liposarcomas from other types of retroperitoneal tumors (11).

CT is the most common imaging technique for the identification, localization, and staging of retroperitoneal sarcomas (18). CT can help in clarifying the anatomical location, size, and possible origin of the tumor, the relation with the adjacent organs, nerves and vessels, and the presence or absence of metastasis.

CT, also, can suggest images of malignancy including large lesion size, presence of thick septa, presence of nodular and/or globular or non adiposes mass-like areas, and decreased percentage of fat composition (20).

MRI is the best techniques for evaluating retroperitoneal tumors (21). MRI is required to further investigate the level of invasion of the tumor,

determine the source of the tumor, and identify the neurovascular or muscle invasion. MRI combined with enhanced-CT can help in the differential diagnosis of various histological subtypes of liposarcomas (22). Along with radiological investigations, pathological, and immunohistochemical investigations form the gold standard for the diagnosis of retroperitoeal liposarcomas.

Percutaneous needle biopsy has low accuracy for the diagnosis of retroperitoneal liposarcomas. A retrospective study on 256 patients underwent to surgical resection with pathologic results of WDLPS or DDLPS noted an unexpectedly low specific diagnostic accuracy of pre-operative percutaneous biopsy: 64% of DDLPS patients and 15% of WDLPS patients were incorrectly diagnosed with other types of tumours, and 53% of false-negative DDLPS cases were diagnosed as WDLPS. In this perspective, the poor accuracy of preoperative biopsy could lead to avoid percutaneous sampling in many cases, especially in symptomatic patients in which surgery is necessary (23).

Surgical resection with appropriate negative margins is the standard primary treatment for most patients with STS (15). In addition to the difficulties to identify and consequently preserve retroperitoneal organs and structures, the high degrees of adipocyte differentiation in the tumour can pose difficulty at distinguishing it from retroperitoneal fat. Thus, the determination of a safe margin for resection becomes difficult.

According to guidelines of NCCN (National Comprehensive Cancer Network) complete anatomic compartment resection is not routinely necessary (15). Prophylactic compartmental resection has not been consistently shown to improve outcomes and is not universally accepted as standard of care (24).

In a study involving 228 patients with retroperitoneal liposarcoma undergoing surgery, patients with combined organ surgeries has a lower 10-year survival rate of 26% as opposed to 35% rate for patients not requiring multiorgan resection (25).

If resections with microscopically positive or grossly positive margins are anticipated, surgical clips should be left in place to identify high risk areas for recurrence, particularly for retroperitoneal or intra-abdominal sarcomas to help guide potential future radiotherapy. Retroperitoneal liposarcoma tends to infiltrate adjacent tissues and structures with skip areas: for this reason it can be explained why it frequently recurs (from 6 to 24 months after initial surgery) despite complete surgical resection with negative margins (26). Beyond these recent evidences there are some cases which need a "tailored" surgical strategy. In our case we report an anatomical condition in which an extent surgery is recommended of necessity, in fact the liposarcoma enveloped and displaced the right kidney and nephrectomy was necessary. Palliative resection is beneficial in case of tumor invasion of the surrounding organs.

Chemotherapy and radiotherapy does not have known definitive effects on retroperitonel liposarcomas (27). Studies have shown that the complete resection of tumors visible to the naked eye improves the prognosis (28, 29). Furthermore, a retrospective analysis revealed that complete surgical resection led to a 3- and 5-year survival rate of 87% and 49%, respectively (30).

The prognosis of retroperitoneal liposarcomas depends on the extent of differentiation, histologic subtype, margin of resection, complete resection of the tumour, and need for contiguous organ resection (14). Prognosis of WDLPS is generally excellent where the 5-year survival rate is 90% with 5-year local recurrence rate at approximately 50%.

Following R0 resection, a well-differentiated subtypes has a 5-year survival rate of 90% while for pleomorphic subtypes the rate is only 30-50% (31, 32).

In contrast, other histologic types have lower 5 years survival rate and necessitates a more aggressive approach (14).

Conclusion

WDLPS are rare tumors. Due to the absence of distinguishing features on presentation imaging the can be misdiagnosed. The gold standard treatment is represent by surgery even if it is difficult a precise pre-operative diagnosis.

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