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Case Report

Case report of giant retroperitoneal liposarcoma in a young woman

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ABSTRACT

Retroperitoneal liposarcoma is a rare malignant mesenchymal tumor with an annual incidence of approximately 2.5 cases per 100,000 inhabitants. It does not present specific symptoms, so its early diagnosis is difficult and in most cases it is performed when it has a large size. A 45-year-old patient with a history of increased abdominal perimeter and local discomfort was admitted in our center. After performing an abdominopelvic CT, she was diagnosed of a large, bulky tumor of lipomatous origin that significantly displaced the intestinal loops and the rest of neighboring organs. In view of the clinical suspicion of a liposarcoma, the patient underwent a resection of the lesion located in the retroperitoneum. The histopathological study determined a well differentiated retroperitoneal liposarcoma. The well-differentiated liposarcoma located in the lower limbs and the retroperitoneal space. Its age of presentation is between the fourth and sixth decade of life without finding differences between men and women. Its treatment is surgical and involves the resection of the tumor mass with non-affected surgical margins. Retroperitoneal liposarcoma is a malignant tumor whose treatment is fundamentally surgical. These tumors tend to be resistant to radiotherapy and / or chemotherapy. Among the most important prognostic factors related to survival is surgery with non-affected margins.

Keywords: Liposarcoma, Retroperitoneal, Sarcoma, Surgery, Well-differentiated

INTRODUCTION

Retroperitoneal sarcomas are rare mesenchymal tumors that represent 10-15% of all soft tissue sarcomas.[1,2] They represent less than 1% of the total of malignant tumors in the adult and between 10-15% are located in the retroperitoneum.[3] Among them, retroperitoneal liposarcoma (RLPS) is the most frequent (20-45%), which is followed by other types of tumors such as leiomyosarcoma, malignant fibrous histiocytoma or undifferentiated pleomorphic sarcomas.[1,3,4] Most RLPS originate from perirenal fatty tissue.[5,6] Next, we report the case of a 45-year-old woman who underwent surgery in our center for a large retroperitoneal tumor that caused an increase in the abdominal perimeter with local discomfort whose histopathological analysis revealed a well differentiated retroperitoneal liposarcoma.

CASE REPORT

A 45-year-old female with no medical history of interest who was admitted for study in our center due to an increase in the abdominal perimeter of months of evolution without other associated symptoms. He did not report abdominal pain or weight loss. The physical examination showed an asymmetry of the abdominal wall

with mass effect in the right of the abdomen. The blood tests showed no changes except for a slight elevation of liver enzymes (ALT and AST), so an abdominal ultrasound was performed to complete the study. The ultrasound revealed an increase in retroperitoneal space content with heterogeneity and displacing the visceral bundle.

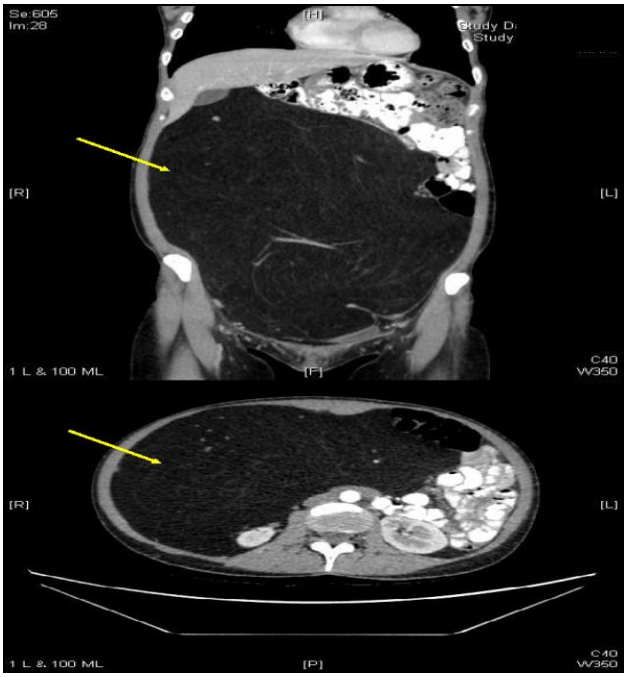

Figure 1: Mass of fat density in CT scan (yellow arrows).

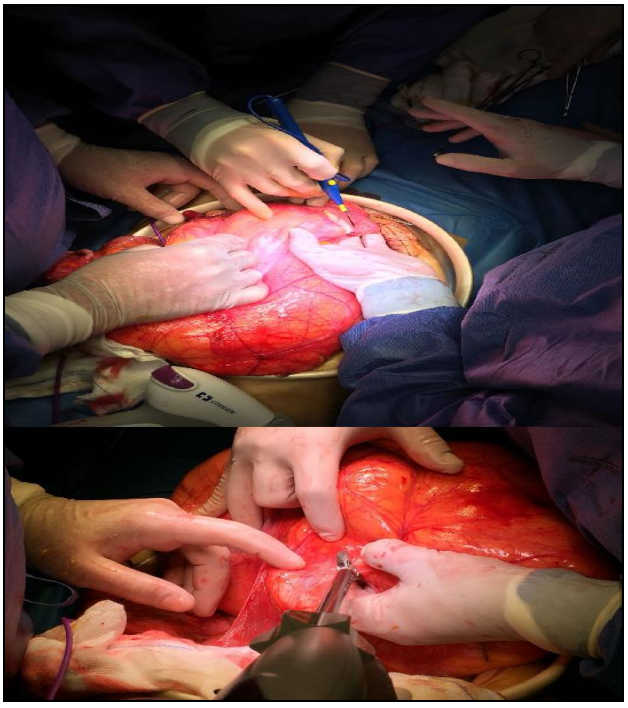

Figure 2: Dissection of giant retroperitoneal liposarcoma.

A thoraco-abdominopelvic CT scan with oral and intravenous contrast was performed, showing an occupation of the abdominal cavity by a mass of fat density, without solid poles or calcifications, of dimensions 330x230x170 mm, originated in the retroperitoneal space, displacing the intestinal loops towards the left hypochondrium (Figure 1). No vascular or visceral invasion was observed. There were no secondary lesions in liver or lung.

The patient underwent surgery, performing under general anesthesia, a suprainfraumbilical laparotomy that showed a large lipomatous tumor that occupied practically the total of the abdominal cavity, displacing the intestinal loops (Figure 2). (the ascending colon and the duodenum were in the left hypochondrium). The tumor was dissected with respect to the adjacent structures (inferior vena cava, aorta, ascending colon and duodenum) without extensive resection, except for the left gonadal vessels that were intimately attached to the tumor. Both ureters could be preserved.

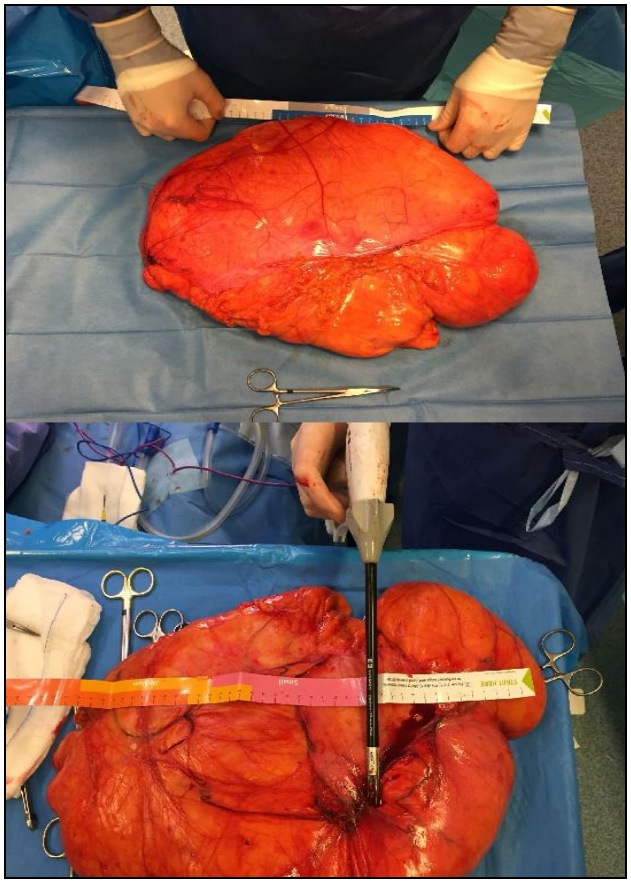

Figure 3: Retroperitoneal liposarcoma removed.

The histopathological result revealed a low-grade, well-differentiated retroperitoneal liposarcoma, measuring 380x240x170 mm, with 5.432 grams of weight (Figure 3). The surgical margins were free of tumor infiltration. The patient was discharged on the tenth day of hospital admission without incidences, currently requiring follow-up without receiving adjuvant treatment.

DISCUSSION

The first description of a retroperitoneal lipomatoustumor excision was made in 1761 by Giovanni Battista Morgagni during the autopsy of a 60-year-old woman.[3]

RLPS are uncommon malignant tumors, representing between 0.07 and 0.2% of all neoplasms.[5] They have an incidence of approximately 2.5 inhabitants per 100,000, with an average age of presentation between 40 and 60 years, with a distribution in both sexes equally.[1,7]

Most of them are diagnosed incidentally, when performing an imaging test for another reason, since most are asymptomatic. They can produce during their growth (usually when they exceed 20 centimeters) nonspecific abdominal pain, early satiety, neurological or obstructive symptoms (urinary or digestive) by compression.[2,5,8]

For its diagnosis, the test of choice is computed tomography (CT) with intravenous contrast, since it allows in most cases an adequate staging and preoperative evaluation. The RLPS appears as a large homogeneous encapsulated mass of fatty tissue with fine septa displacing the renal parenchyma or even the intestinal bundle as in our case.[1,5]

Abdominal ultrasound may confirm the presence of a hyperechoic mass and may be useful at the beginning of the study.[5] Pelvic magnetic resonance is reserved to rule out muscular or vascular invasion as well as to discover satellite lesions or recurrences.[5,7]

The differential diagnosis should be made mainly with renal or adrenal tumors, pancreas, lymphomas, lymphangiomas, retroperitoneal fibrosis, advanced gastrointestinal carcinomas or even with testicular tumors.[5]

The World Health Organization (WHO) has classified the liposarcomas into two groups according to the degree of differentiation in low grade (where the well-differentiated liposarcoma and the myxoid are found) and high degree (dedifferentiated, pleomorphic liposarcoma and of mixed cells).[6] The well-differentiated and the dedifferentiated liposarcoma are the most frequent types. Well-differentiated liposarcoma have a slower growth rate and have a less aggressive behavior with a lower rate of distant metastasis with respect to dedifferentiated liposarcomas.

Its management is fundamentally surgical. The use of neoadjuvant or adjuvant chemotherapy and / or radiotherapy is controversial given the low sensitivity of these tumors.[9] Lines of treatment with doxorubucin present a rate of 18-29% of responses.[1] Authors such as Balle have not found benefits in the use of radiotherapy, describing various side effects such as neuropathy, hydronephrosis, ureteral fistula and / or intestinal obstruction.[10] However, the use of adjuvant radiotherapy is used for tumors larger than 5 cm with positive surgical margins to reduce recurrence without increasing survival.[1]

The most important prognostic factor in these tumorsis complete resection with free margins. Complete resection (R0) increases survival from 16.7% to 58% according to series, with median survival of 103 months versus 18 months in R1 and R2 resections.[5,7] Some authors recommend resection of the organs in contact with the tumor (extensive resection) while others postulate dissection with unaffected macroscopic margins (colon, pancreas, ureters, iliac vessels, aorta, inferior vena cava). For authors like Gronchi as well as for the French Group of Sarcomas, they describe a relapse rate between two and three times lower in those patients who undergo resections that involve the removal of the tumor mass together with other related organs.[11,12] In contrast, for other authors, extensive resection of these tumors does not decrease the recurrence rate, increasing only morbidity and mortality.[9] Lewis describes a perioperative mortality of 4% (secondary to hemorrhage, sepsis, coronary event or multi-organ failure).[13] However, morbidity increases when more than three organs are resected.[9]

In conclusion, liposarcomais a rare tumor that due to its retroperitoneal location does not present specific symptoms, being diagnosed when they present a large size and produce compressive symptoms. Its management is surgical fundamentally, being the use of the chemotherapy and /or radiotherapy controversial due to the low sensitive of these types of tumors.

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