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Retroperitoneal liposarcoma a case report of a rare mesenchymal tumor

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ABSTRACT

Retroperitoneal liposarcoma is a rare clinical entity characterized by its initial asymptomatic development and its progressive increase in size leading to compression of neighboring organs, thus constituting the most frequent mode of revelation. Surgery is the only potentially curative treatment. The use of neoadjuvant or adjuvant chemotherapy and/or radiotherapy is controversial due to the low sensitivity of these tumors. Despite adequate management of the tumor, the risk of recurrence is high, often necessitating one or more additional resections.

Introduction

Retroperitoneal liposarcoma is a rare tumor belonging to the sarcomas, developing silently for years before symptoms due to compression and invasion of neighboring organs. We present here the case of a patient operated for a liposarcoma in the department of digestive cancer surgery and liver transplantation at University Hospital Center Ibn Rochd of Casablanca. The aim is to analyze the epidemiological aspects and diagnostic difficulties of this condition, and to specify the therapeutic means and prognostic elements involved.

Case :

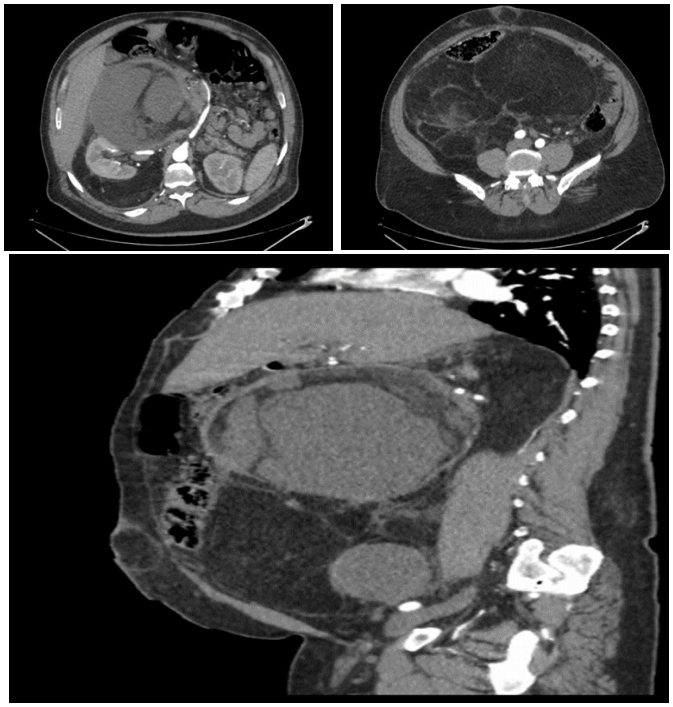
We report the case of a 54-year-old patient who presented an abdominal distension evolving for 3 months and a weight loss of 15 kg.

Clinical examination revealed an abdominal mass extending from the right hypochondrium to the right iliac fossa, extending beyond the midline towards the contralateral side, with poorly defined contours measuring approximately 30 cm in long axis, An abdomino-pelvic angioscan was performed, which revealed:

- A first subhepatic mass luxating the pancreaticoduodenal junction towards the midline and to the left. It measured 182 x 155 x 120 mm. It displaces the superior mesenteric vessels to the left and laminates the IVC posteriorly, It exerts a significant mass effect on the right kidney, which is pressed back against the posterior abdominal wall.
- The second mass is located in the right iliac fossa. It is hypodense, poorly enhanced and

well limited, measuring 80 x 81 x 57 mm.

- The third mass is retro-umbilical, displacing the colon and an intestinal ansae peripherally. It is fatty in nature, with inhomogeneous components. It measures approximately 285 x 200 x 140mm.



Figures (1) and (2) CT scan pictures showing the three masses as described

The case was discussed at a multidisciplinary consultation meeting on digestive and vascular oncological surgery, where the decision was taken to perform a carcinological resection of the tumor. Surgical exploration revealed a 60 cm long retroperitoneal mass with a double tissue and fat component, pushing the right colon forward and adherent to the duodenum and the pancreas, the right colonic angle and the lower surface of the liver. This mass encompasses the right kidney and compresses the inferior vena cava, with the development of collateral venous circulation.Without visualization of effusion, metastases or peritoneal nodules.

A carcinological resection of the mass was performed, removing the right kidney and the right adrenal gland, with recutting of the liposarcoma capsule at the level of the right renal pedicle and the serosa of the right colon. The tumor weighed 10 kg.

The post-operative course was straightforward, and the patient was discharged on the 5th post-operative day.

Anatomopathological examination of the surgical specimen showed a myxoid liposarcoma with pathological resection margins, and the recuts on the renal pedicle and serosa of the right colon were healthy.

Clinical follow-up and follow-up imaging after one year did not reveal any tumor recurrence.

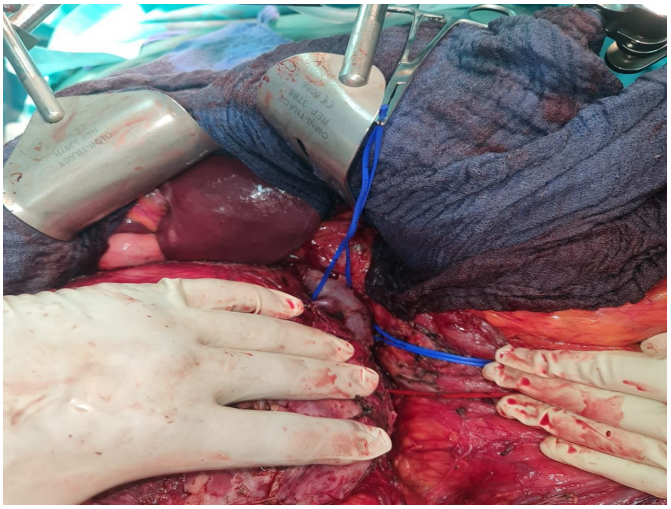


Figure 3 : Image showing the mass in contact with the inferior vena cava and the right kidney with its pedicle

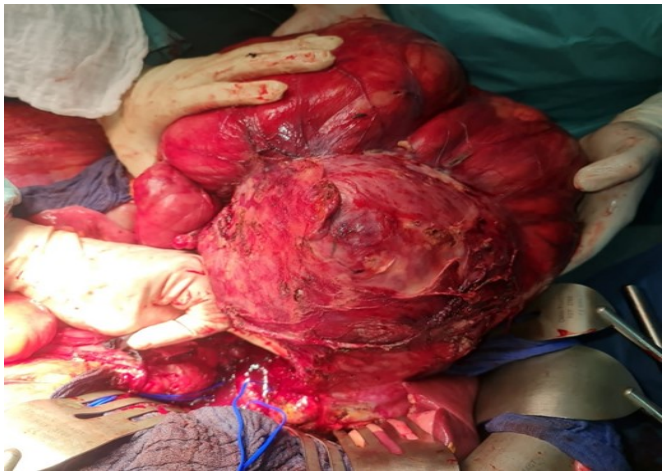
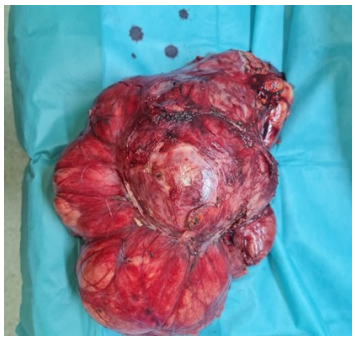
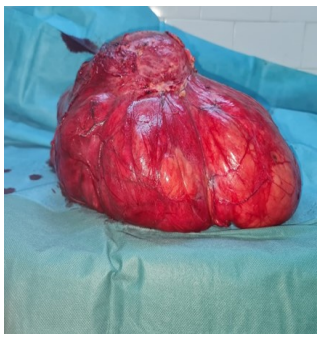


Figure 4: Intraoperative image of the mass during its dissection





Figures 5&6 : “Image of the mass after total resection”

Discussion

Soft tissue sarcomas are rare mesenchymal neoplasms with over 150 different histological subtypes, accounting for 1% of adult malignancies. Liposarcomas are a common subfamily, collectively accounting for 10-15%. (1) (2). They are a rare tumor of adipocytic origin, with an estimated incidence of 1 case per 100,000 people per year, of variable location, most frequently in the retroperitoneum which account of 0.07 to 0.2% of all cancers (3), affecting subjects aged between 40 and 60, with a slight male predominance (4).

The liposarcoma subtypes defined by the World Health Organization (WHO) include atypical lipomatous tumors (ALT), well-differentiated liposarcoma (WDLPS), dedifferentiated liposarcoma (DDLPS), myxoid liposarcoma (MLPS), pleomorphic liposarcoma (PLPS) and myxoid pleomorphic liposarcoma (MPLPS). The quantity of lipids inside the cells, mucoid lipids and the degree of cellular differentiation are the essential elements of classification. (6)

Well-differentiated liposarcoma WDLPS is the most common type in the retroperitoneum (7), and is a slow-growing tumor that often recurs locally, but has minimal metastatic potential. Undifferentiated liposarcoma may be primary or develop from a pre-existing WDLPS, and is generally associated with a higher rate of growth and occurrence of metastasis. MLPS have an aggressive clinical course and a marked propensity to metastasize, often to unusual sites such as bone and skin. PLPS typically occur in the elderly, with a marked tendency to metastasize (3). Myxoid, round-cell and pleomorphic liposarcomas are rare in the retroperitoneum. (8).

Liposarcomas are often asymptomatic in 78% of cases (2), until they reach a significant size, with an average diameter of 20 to 25 cm and a weight of 15 to 20 kg, before producing symptoms due to compression of other adjacent organs (1). 60 to 80% of masses are palpable, and almost 50% of patients present with abdominal pain (9).

Abdominopelvic CT is the key examination, demonstrating a heterogeneous, hypodense tumor, clarifying relationships with neighboring organs, guiding puncture and biopsy, and enabling postoperative monitoring. MRI can detect vascular invasion, especially of the inferior vena cava and the abdominal aorta (10).In our case, abdominal angioscan was performed to better characterize vascular relationships.

Percutaneous biopsy is the examination of choice for confirming the diagnosis of liposarcoma, the indication for which must be discussed in multidisciplinary consultation meeting on a case-by-case basis(11) (2).

Complete resection is the treatment of choice for retroperitoneal liposarcoma. The use of neoadjuvant or adjuvant chemotherapy and/or radiotherapy is controversial due to the low sensitivity of these

tumors. The principle of surgery is monobloc R0 resection of the tumor and all invaded organs. Complete resection of retroperitoneal liposarcoma can increase the 5-year survival rate from 16.7% to 58%.(12)(17)

The kidney is the most frequently resected organ, followed by the colon. In our case, resection of the right kidney was necessary. It should also be noted that, given the difficulty of differentiating between tumor tissue and healthy tissue in certain cases, tumor resection must include a significant amount of adjacent peritoneal fat. Local recurrence remains the leading cause of mortality in retroperitoneal liposarcoma.(1)(13)

Survival rates were better in patients who had undergone complete resection of their recurrent tumor than in those who had not. Therefore, the gold standard of treatment remains R0 complete resection of the tumor and possible recurrences. (14)

The prognosis of liposarcoma remains poor due to the volume of the tumor at diagnosis and its proximity to large blood vessels, which often leads to incomplete resection (15).

Retroperitoneal liposarcoma often recurs within 6 months to 2 years after initial surgical resection, despite complete surgical resection. It presents rapid growth, with a doubling of tumor volume over 100 days .The 5-year survival rate for well-differentiated retroperitoneal liposarcoma is 83%, compared with 20% for the dedifferentiated subtype.(3)(16)

Conclusion

Retroperitoneal liposarcoma is a rare entity characterized by its rapid and silent evolution. Surgical treatment is the only curative treatment for the pathology, and a multidisciplinary approach is required for its management, which must be discussed first at the multidisciplinary consultation meeting in order to optimize surgical outcome and improve patient survival. Long-term follow-up of patients is essential, given the high risk of recurrence.

Conflicts of interest:

The authors declare having no conflicts of interest for this article

Ethical approval:

I declare on my honor that the ethical approval has been exempted by my establishment

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