

RARE: ENORMOUS RETROPERITONEAL LIPOSARCOMA CASE REPORT***R. Ouaddane Alami, O. Chama, M. Ahsaini, S. Mellas, J. El Ammari, MF. Tazi, MJ. El Fassi and MH. Farih**

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ABSTRACT

Retroperitoneal liposarcomas are malignant mesenchymal tumors that develop from fat cells in the retroperitoneal space. They are primitive and independent of the organs in this space, sometimes invading the kidney. They are rare and have benefited from imaging techniques, in particular ultrasound, CT and MRI. However, these tumors still pose diagnostic and therapeutic problems, they are often diagnosed as a palpable mass. The reference treatment is complete tumor resection with healthy margins and without invasion. For this purpose, some authors recommend an extension to the proximal organs. However, the anatomical constraints of the retroperitoneal space and the often large tumor volume limit the possibilities of achieving satisfactory clearance.

KEYWORDS: Retroperitoneal liposarcoma, enormous, giant tumor, multidisciplinary.**INTRODUCTION**

Sarcomas are rare malignant tumors. They occur in the soft tissue in more than 50% of cases.^[1] Soft tissue sarcomas (STS) account for 1% of all soft tissue tumors, less than 1% of all adult cancers, and about 15% of all childhood cancers.^[2, 3] Retroperitoneal sarcomas (RPS) are rare and heterogeneous tumors. They represent 15% of soft tissue sarcomas, with an annual incidence of 1000 in the United States and 300 in France.^[4,5] The basic treatment is complete surgery with healthy margins and without invasion, but this is often difficult. The quality of the excision is the determining prognostic factor found in the literature.^[6-7] The prognosis of these tumors remains unfavorable due to a high rate of locoregional recurrence. The five-year overall survival rate of all PRS does not exceed 15 to 30%.^[8] In order to optimize the therapeutic outcome of these rare tumors, a multidisciplinary management is mandatory.

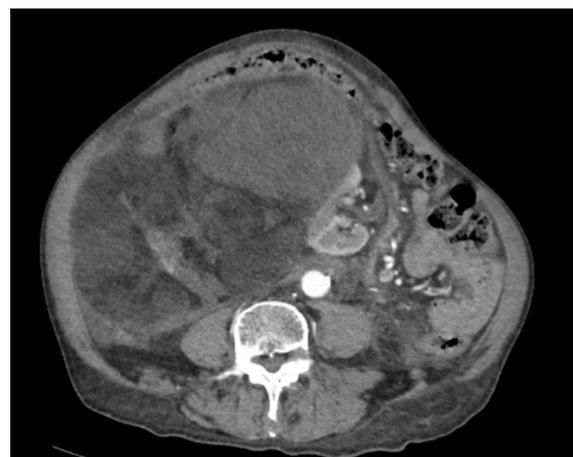
We report the case of a patient diagnosed with a giant Liposarcoma of the renal lodge collected at our department in order to underline the anatomical and surgical particularities of this localization within the retroperitoneum, and its possible prognostic repercussions.

CASE REPORT

A 78-year-old man with no previous pathological history, was admitted to hospital for the assessment of a right flank mass, with a 2 year history of intermittent abdominal pain and a palpable abdominal mass, all

evolving in a context of deterioration of the general state and anorexia. The examination showed a mass on the right flank, extending upwards towards the right hypochondrium, hard and non-mobilizable, painful to palpation, with a lumbar contact. The rest of the examination was normal.

Abdominal and pelvic CT scan showed a huge limited mass occupying the right hemi-abdomen, measuring 14x19x20cm (Fig 1). It crushes and pushes the kidney medially. It exerts a mass effect on the surrounding structures without signs of invasion. It is of hypodense tissue density, heterogeneously enhancing after injection of contrast medium and without signs of necrosis. This aspect suggests the malignant adipose nature of the process developed in the renal pelvis.



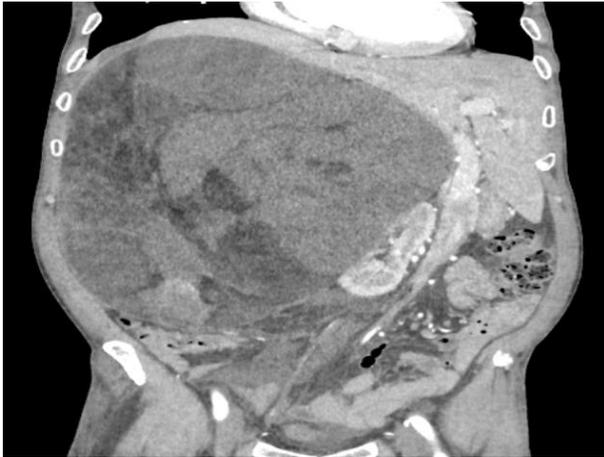


Figure 1: Abdominal and pelvic CT scan showed a huge limited mass occupying the right hemi-abdomen, measuring 14x19x20cm.

On these data, the points discussed on the technical level were: how to have a good operative exposure and how to approach the renal artery because of the large tumor volume and the way the mass develops in contact with the renal pedicle. The patient was operated by a median xyphopubic laparotomy, enlarged by a right transverse split. On exploration, the tumor occupied the right supra- and sub-mesocolic level, and plunged into the pelvis crossing the midline to the left. The key moment of the operation was, after detachment of the right colon, the liberation of the lower pole of the tumor allowing the location and control of the renal artery before any dissection of the left renal vein molded on the tumor.

The operation lasted 3h30min and the bleeding was evaluated at 300ml. The anatomopathological examination was performed on an enlarged tumor removal specimen taking the whole renal compartment and its contents, weighing 4200g and measuring 30x20x17cm (Fig 3).

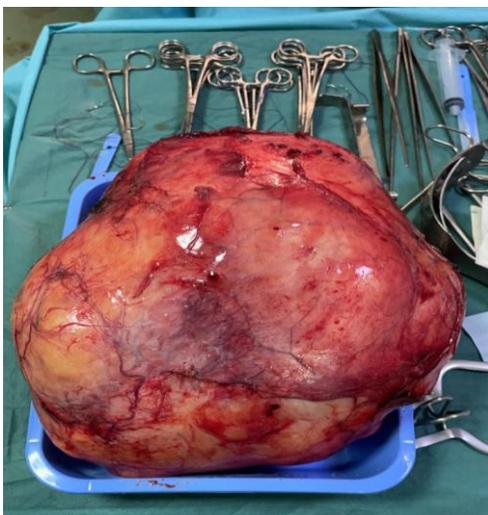


Figure 3: Postoperative image showing a gigantic retroperitoneal liposarcoma.

Histopathology revealed a well-defined retroperitoneal liposarcoma. The patient was doing well without recurrence at 18 months, no adjuvant treatment has been performed.

DISCUSSION

Liposarcomas are rare tumors representing less than 1% of cancers.^[9] They occur at any age (3-76 years), but are more frequent in young adults (mean age 44 years)^[10], with a slight female predominance.

These tumors can be large (20 cm or more) and of considerable weight (3.5-9 kg).^[11] Histologically, well differentiated, moderately differentiated (myxoid) and undifferentiated forms are described.

The clinical symptomatology is variable. Abdominal pain is present in 50 to 88% of cases and sometimes intense, often dorsolumbar or pelvic.^[11] Digestive signs are observed in 60% of cases^[4], such as vomiting, digestive hemorrhage and transit disorders. Other signs may be found: neurological (10-30% of cases^[8]), urinary (6-11% of cases^[3]), venous compression (10-11% of cases.^[2,11]

Abdominopelvic ultrasound allows the diagnosis in 30-50% of cases. Abdominal and pelvic CT shows a hypodense, heterogeneous tumor and specifies the relationship with the neighboring organs; it represents the best means of postoperative surveillance; it allows fine needle biopsies to be performed, but this puncture is not unanimously accepted by the authors, because of the possible existence of different cell types within the same tumor.^[9] Normal intravenous urography does not eliminate a retroperitoneal tumor. MRI clarifies the characteristics of the mass, and can detect vascular invasion by specifying the extent of the thrombus. Paraneoplastic hypercalcemia has been described.^[7, 9]

Surgery is the treatment of choice for these tumors. The transperitoneal approach is the most commonly used^[2] and depending on the location of the tumor, it can be a thoraco-abdominal approach, a median laparotomy or a transverse approach. Exploration is a crucial time to judge the resectability of the tumor and among the factors that oppose resection, invasion of the large vascular trunks, hepatic metastases and invasion of the root of the mesentery should be noted.^[11,12] Sometimes only an incomplete excision could be performed.^[9] Surgery is responsible for 10% of mortality.^[2, 6, 12]

Radiation therapy is used as a complement to surgery and the doses are high (more than 60 Gy). The results depend on the volume of the tumor. It can be preoperative for unresectable tumors, or postoperative between two recurrences.^[7] The response rate to chemotherapy is between 20 and 35%.^[5,10] Chemotherapy can be used in the treatment of metastatic tumors, in the adjuvant or neoadjuvant setting.^[13]

CONCLUSION

Retroperitoneal sarcomas are rare tumors with a poor prognosis. Complete surgery is formally recommended to reduce the risk of local recurrence and metastasis. The role of radiotherapy and neoadjuvant chemotherapy is currently being evaluated. Multidisciplinary management by a specialized team is recommended. Given the absence of anatomical boundaries or barriers outside the renal pelvis in the retroperitoneum and the large size of these sarcomas, successful resection with uninvaded microscopic margins is difficult to achieve even with macroscopically complete resections.

Conflict of interest

None.

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