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Case Report

Surgical Resection of A Giant Liposarcoma in the Renal Compartment: A Case Report and Literature Review

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Abstract: Retroperitoneal sarcomas are rare tumors that develop in the mesenchymal tissue, where liposarcoma is the most frequently encountered subtype. The symptoms they induce are often vague and nonspecific, thus requiring thorough evaluation. Their diagnosis primarily relies on imaging techniques such as CT scans and MRI, and is confirmed by biopsy. Despite excision surgery, which remains the main treatment aiming for total resection, local recurrences remain a frequent challenge. Although radiotherapy and chemotherapy in the perioperative period are sometimes considered, their effectiveness remains subject to debate. The histological type of the tumor is of paramount importance in prognosis, with well-differentiated liposarcomas being associated with better outcomes than myxoid or dedifferentiated forms. Optimal management of these tumors requires a multidisciplinary approach, involving close collaboration between surgeons, oncologists, radiologists, and pathologists to ensure adequate and personalized treatment. Hence, the interest of a case of giant retroperitoneal liposarcoma in a 65-year-old man that we managed and report here.

Keywords: Surgical Resection, Giant Liposarcoma, Renal Compartment.

INTRODUCTION:

Sarcomas are types of primary tumors of mesenchymal tissue, rare, accounting for about 1% of all malignant tumors [1]. The majority of tumors localized in the retroperitoneal region are malignant, with approximately 30% of them identified as sarcomas, among which liposarcoma constitutes approximately 40 to 50% of cases [2]. Although most sarcomas develop de novo, two risk factors have been identified: radiotherapy, which appears to be associated with an increased risk of radiation-induced sarcomas, characterized by their aggressiveness and poor prognosis, as well as several hereditary conditions, such as neurofibromatosis type I, which may increase the risk of developing soft tissue sarcomas [3]. Various histological varieties of sarcomas with increasing degrees of malignancy have been identified, including well-differentiated liposarcoma, myxoid, pleomorphic, mixed, and dedifferentiated [4].

OBSERVATION:

A 65-year-old patient presented with diffuse abdominal pain, more accentuated in the right flank, with a sensation of abdominal heaviness and altered bowel movements resembling constipation. Clinical examination was entirely normal. An initial ultrasound revealed a huge retroperitoneal mass, with homogeneous echogenicity and regular contours, extending from the right pelvic region to subhepatic without local infiltration or deep lymphadenopathy (Figure 1 and 2). A thoracoabdominopelvic computed tomography scan showed a large fatty mass in the right lumbar fossa, encasing the right kidney, extending down to the ischial fossa, measuring 122 x 100 x 230 mm in height, without signs of local or distant aggressiveness (Figure 3 and 4). An MRI revealed a large solid abdominopelvic mass lateralized to the right, appearing hyperintense on T1 and T2 signals, containing septations, enhancing after contrast injection, measuring 134 mm x 269 mm in height,

suggestive of an expansive retroperitoneal liposarcoma (Figure 5). A biopsy was performed using a protected needle, and histopathological examination suggested a well-differentiated liposarcoma with focal positivity of cells to anti-MDM2 antibody on immunohistochemistry.

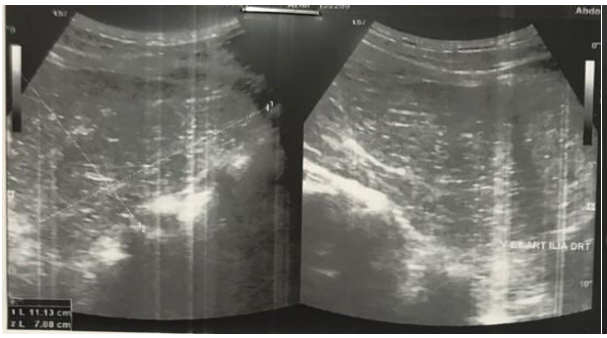

Figure 1: Ultrasonographic appearance of the mass

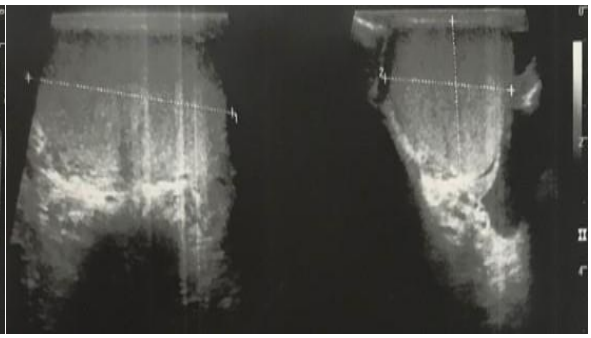

Figure 2: Ultrasonographic appearance of the mass

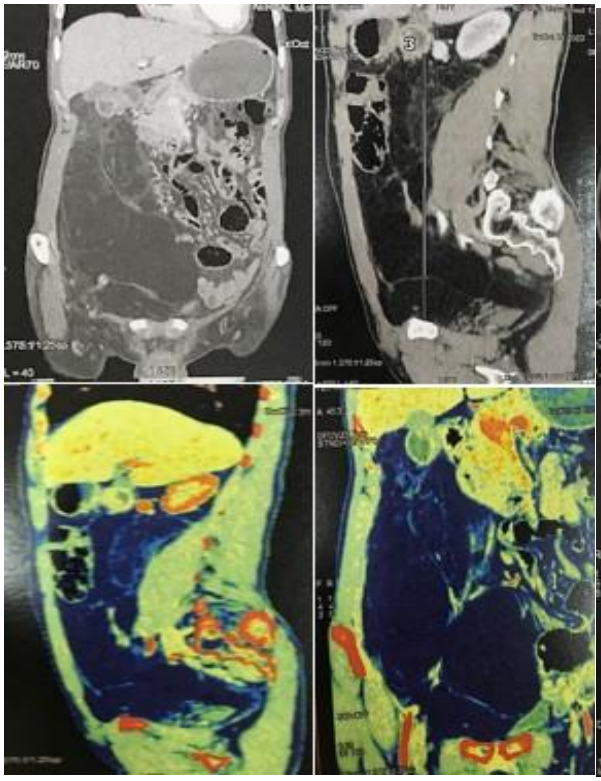

Figure 3: Appearance of the liposarcoma on CT scan

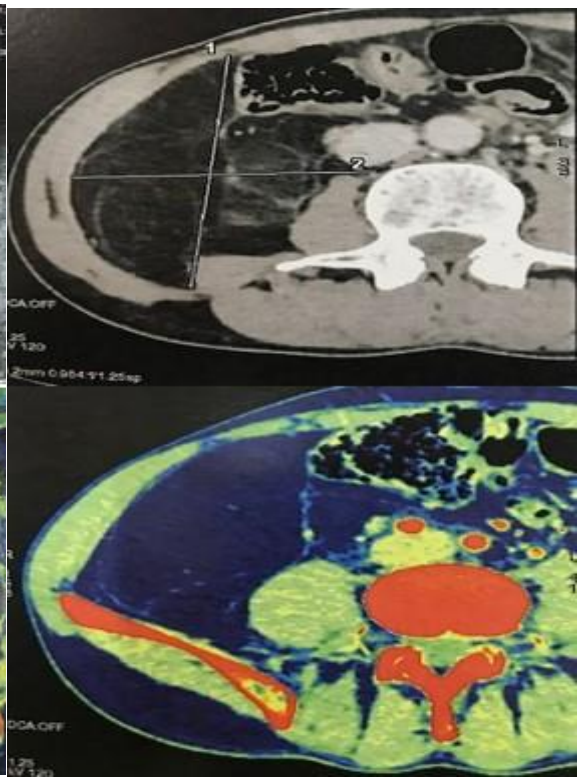

Figure 4: Cross-sectional view on CT scan

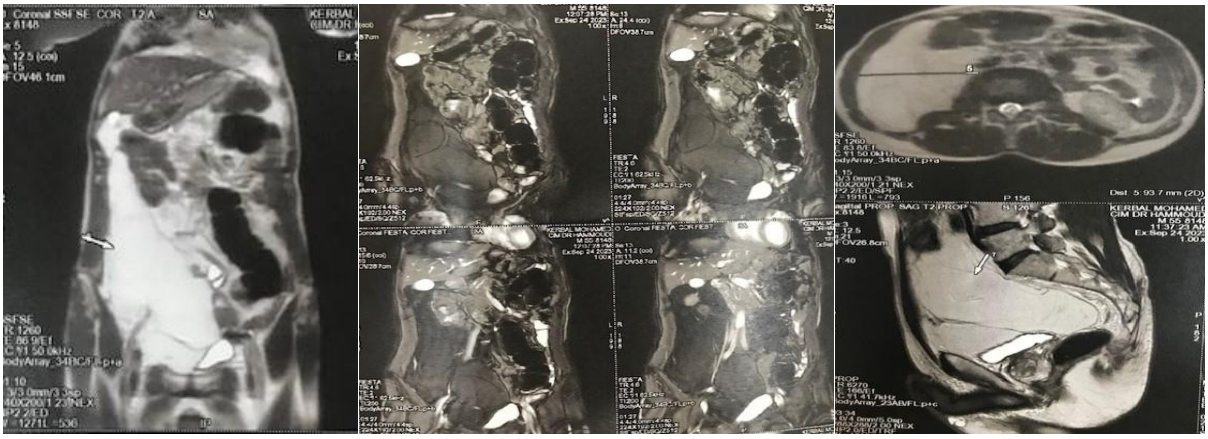

Figure 5: Appearance of the liposarcoma on MRI

The patient underwent surgery, approached through a midline incision, after right coloparietal detachment. A lobulated fatty mass was found, extending from the suprarenal region to the pelvis without infiltration of adjacent organs and without presence of lymphadenopathy (Figure 6-9). A monobloc resection was performed without tumor breach, preserving the right kidney. Postoperative recovery was uneventful, and the patient was discharged on the 5th day after the procedure. Histopathological and immunohistochemical examination of the tumor revealed features consistent with a well-differentiated liposarcoma (WHO 2020) of grade 1 according to the FNCLCC classification, located in the abdominopelvic region. Subsequently, the patient underwent radiotherapy.

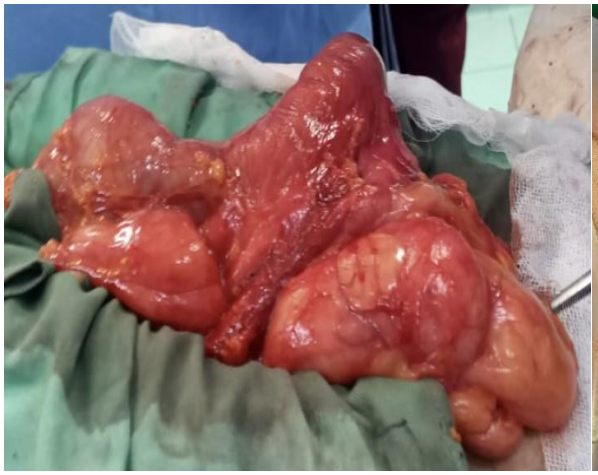

Figure 6: Appearance of the mass after dissection

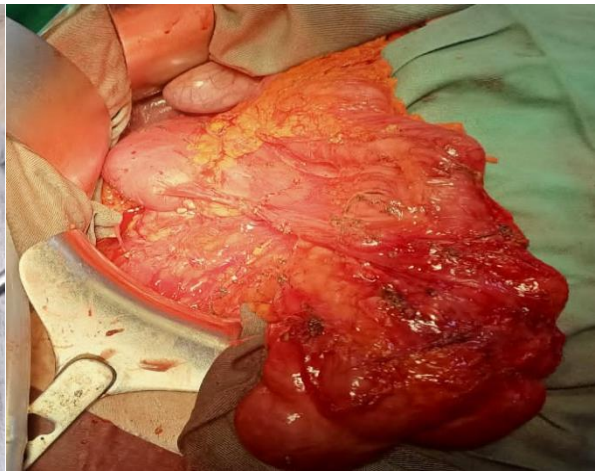

Figure 7: Appearance of the right kidney

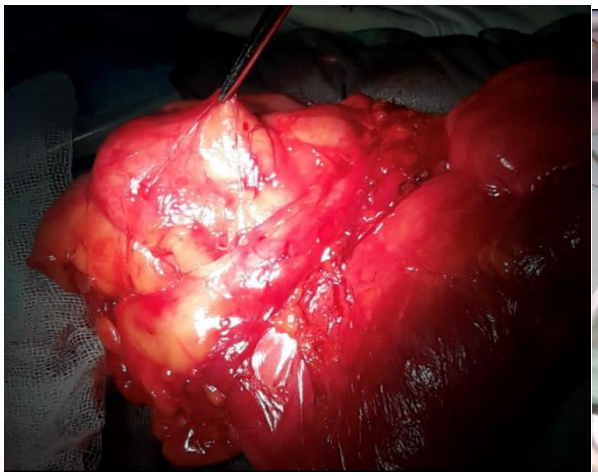

Figure 8: Perioperative appearance of the capsule

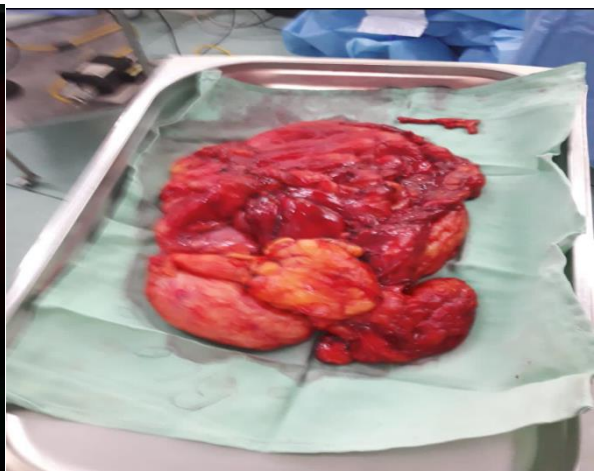

Figure 9: Appearance of the mass after resection

DISCUSSION:

Retroperitoneal liposarcomas are malignant tumors that can affect both sexes indiscriminately. Although the average age at diagnosis typically falls around the fifth decade of life, it is important to note that this disease can occur at any age, thus affecting all age groups of the population [5,6]. Symptoms associated with retroperitoneal liposarcomas are often nonspecific, usually resulting from compression of surrounding organs by the tumor. The most common manifestations include abdominal pain or a feeling of heaviness observed in the majority of patients, as well as the presence of an abdominal mass. Less frequently, these symptoms may be accompanied by urinary and digestive disorders, deterioration of general health, and sometimes even hyperthermia related to tumor necrosis. Overall, while these signs may vary from patient to patient, they are generally related to compression of adjacent structures by the tumor [7-9].

Performing a contrast-enhanced thoracoabdominal-pelvic CT scan is essential in the diagnostic process and in formulating the therapeutic plan for retroperitoneal liposarcomas. It allows for a positive diagnosis in the presence of a lesion localized in the retroperitoneum and not associated with a specific organ. Retroperitoneal localization can sometimes be difficult to assess, especially with large masses, but the displacement of retroperitoneal organs provides useful clues. Liposarcomas can spread through various anatomical passages such as the inguinal canal in our patient. Regarding histological nature, liposarcomas generally present with fatty density

components and vascularized septa. Similarly, CT scan enables planning of tumor or metastasis biopsy, as well as evaluation of nodal and systemic extension as part of the staging workup. It can also provide valuable insights to surgeons, enabling them to study resectability criteria to determine the appropriate therapeutic strategy [10].

MRI is often recommended as an adjunct to CT scanning in cases where assessment by CT is challenging. It offers better accuracy in assessing extension, especially in the pelvic region, where it can be particularly useful in delineating the lesion. The MRI protocol typically includes classic T1 and T2 sequences, fat saturation sequences, diffusion sequences, and dynamic sequences. The use of a body coil allows for studying tumor boundaries in all planes, providing a comprehensive view of disease extension. When radiotherapy is considered, MRI aids in delineating the treatment volume, which must be optimal and also include peritumoral edema to ensure effective management [10].

18FDG positron emission tomography-computed tomography (PET-CT) can play an important role in the follow-up of tumors in case of recurrence. Its sensitivity and specificity superior to those of CT in this situation make it a particularly valuable imaging modality. This technique detects areas of increased metabolic activity, characteristic of malignant tumors, using a radioactive tracer, 18FDG, which is selectively absorbed by cancer cells, thereby identifying sites of tumor recurrence even at early stages, thus offering earlier detection than conventional CT. Due to its high sensitivity, 18FDG PET-CT is also useful for assessing treatment response and for detecting potential occult metastases. Its use in regular patient follow-up can therefore contribute to earlier and more precise management of recurrences [11].

The positive diagnosis of retroperitoneal sarcomas generally relies on histological confirmation obtained through biopsy. This approach is essential for establishing an accurate diagnosis, especially since retroperitoneal sarcomas represent only one-third of the tumors present in this region. Image-guided percutaneous biopsy is the standard method for obtaining tissue samples. It is usually performed via an extra-peritoneal route using a protected needle, minimizing the risk of pathway contamination. It is recommended to sample from different areas whenever technically and reasonably feasible, to obtain adequate representation of the tumor and improve diagnostic accuracy [12]. Although there is a risk of biopsy tract contamination, it is generally low and is not associated with an increased risk of local recurrence. Thus, despite this risk, biopsy remains a safe and effective procedure for confirming the diagnosis of retroperitoneal sarcoma [13].

The classification established by the World Health Organization (WHO) in 2020 serves as a major reference in categorizing connective tissue tumors. This classification is based on the distinction of tumors according to their differentiation lineage. For each type of tumor, this classification proposes subcategories including benign varieties, intermediate malignancy tumors, and malignant forms (Table 1). The criteria used to classify connective tissue tumors rely on a combination of morphological, immunohistochemical, and molecular characteristics specific to each tumor type. This approach allows for a more precise and tailored classification to the diversity of tumors encountered in clinical practice. In the abdominopelvic context in adults, well-differentiated/dedifferentiated liposarcomas and leiomyosarcomas are among the most frequently observed subtypes [14,15].

Table 1: WHO Classification of Abdomino-Pelvic Soft Tissue Sarcomas

Adipose Tumors
• Well-differentiated/Dedifferentiated Liposarcoma • Myxoid Liposarcoma • Pleomorphic Liposarcoma

The prognosis of sarcomas has largely been based on morphological criteria, primarily determined by the grade of malignancy established by the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC). This grade takes into consideration several aspects, including sarcoma differentiation, mitotic index, as well as the presence and percentage of necrosis within the tumor. Each parameter receives an independent score, typically ranging from 1 to 3, and the overall grade is then determined by the sum of these scores. It is important to note that grading evaluation is not reliable after preoperative chemotherapy and/or radiotherapy, as these treatments can alter the histological characteristics of the tumor tissue. Consequently, grading assignment is generally performed before the initiation of any treatment. There are also specific prognostic scores for certain histological entities, such as the Demicco score used to evaluate solitary fibrous tumors. These scores provide additional information on prognosis and help refine treatment and follow-up strategies for patients with sarcoma [16,17].

Table: Histoprognostic Grade of Sarcomas by FNCLCC

Tumor Differentiation
• Score 1: Sarcomas resembling normal adult tissue (e.g., well-differentiated liposarcoma) • Score 2: Sarcomas with certain histological diagnosis (e.g., myxoid liposarcoma) • Score 3: Embryonal sarcomas, synovial sarcoma, epithelioid sarcoma, alveolar soft part sarcoma, undifferentiated sarcomas, and those with uncertain histological type.
Mitotic Index
• Score 1: 0 to 9 mitoses • Score 2: 10 to 19 mitoses • Score 3: More than 19 mitoses
Tumor Necrosis
• Score 0: Absence of tumor necrosis • Score 1: < 50% tumor necrosis • Score 2: ≥ 50% tumor necrosis
Grade 1 (sum of scores = 2 to 3): Low metastatic risk Grade 2 (sum of scores = 4 to 5): Tumor with difficult-to-predict evolution, real metastatic risk Grade 3 (sum of scores = 6 to 8): Poor prognosis tumor, high metastatic risk.

Regarding the TNM classification of retroperitoneal sarcomas, it is primarily determined by tumor size (Table 3), which is not really the best criterion as liposarcomas are diagnosed in the majority of cases with already very large sizes and therefore considered inherently advanced [18].

Table 3: TNM Classification of Retroperitoneal Sarcomas

• Tx: Unable to evaluate • T0: No identified primary tumor • T1: Major axis tumor ≤ 5 cm • T2: Major axis tumor > 5 cm and ≤ 10 cm • T3: Major axis tumor > 10 cm and ≤ 15 cm • T4: Major axis tumor > 15 cm
• N0: No pathological lymph nodes or unknown status • N1: Presence of pathological lymph nodes
• cM0: Absence of metastasis • cM1: Presence of distant metastasis • pM1: Presence of confirmed distant metastasis microscopically.

Chromosomal analysis, as well as cytogenetic and molecular studies, currently play a crucial role in the differential diagnosis of adipose tumors. Well-differentiated liposarcomas are characterized by the presence of supernumerary chromosomes, mainly in the form of a circular ring and unusually large in size. It is now widely established that these supernumerary chromosomes result from amplification of the 12q14-15 sequence on the long arm of chromosome 12. Amplification of the MDM2 gene is almost invariably observed, while amplification of the SAS, CDK4, and HMGIC genes is less frequent. Myxoid and round cell liposarcomas are associated with the inverse translocation t12;16 and q13;p11.

Surgical resection of retroperitoneal sarcomas remains the primary therapeutic strategy, aiming to achieve oncologically sound objectives. Macroscopically complete resection is essential to improve patient survival; however, the rate of local recurrence remains generally high despite this surgical approach. The primary oncologic objective is to perform en bloc resection of the tumor with microscopically clear margins (R0). This approach is particularly crucial given the absence of an effective salvage systemic treatment in case of macroscopically incomplete surgery. Additionally, any breach of the tumor capsule during surgery may promote peritoneal dissemination, leading to sarcomatosis. However, aiming for macroscopically clear margins may result in higher morbidity, especially in cases involving major vessels. In this context, a multidisciplinary approach involving different specialists seems indispensable to develop an appropriate therapeutic strategy. Local recurrence occurs in 20 to 80% of cases, varying depending on histological subtype, grade, and quality of initial resection. Although 85% of retroperitoneal sarcomas operated on with curative intent may result in macroscopically complete resection (R0/R1) during the initial surgery, this rate drops below 50% during second-line surgery. Visceral metastases are detected in 10 to 20% of cases at diagnosis but rarely as the site of first disease recurrence. For selected oligometastatic patients, metastasectomy may offer a survival benefit. Surgical resection of lung metastases, regardless of their number, has demonstrated a survival benefit, unlike hepatic metastases for which surgery is not the standard treatment.

The role of radiotherapy is based on low-level evidence. In the neoadjuvant setting, its use relies on lower doses than those used in the adjuvant setting. Retrospective studies have shown favorable outcomes in terms of overall survival for this approach. The recommended dose is generally 50 to 50.4 Gy administered in fractions of 1.8 to 2 Gy. Surgery is usually scheduled 4 to 8 weeks after the end of treatment. Similarly, regarding outcomes on overall survival in the adjuvant setting, they remain contradictory.

The use of perioperative chemotherapy in the treatment of retroperitoneal sarcomas is justified by several factors, including preoperative tumor reduction, sterilization of micrometastases, and assessment of chemotherapy sensitivity from resection specimens. In the neoadjuvant setting, chemotherapy can induce objective responses, typically between 21 and 33%, except for well-differentiated liposarcoma, which is not sensitive to chemotherapy. These responses can notably reduce tumor volume and make surgery more conservative while lowering the resectability threshold. Systemically, it can destroy micrometastases. Its use is often debated for large high-grade tumors, especially when other surgical interventions are considered. In the adjuvant setting, chemotherapy seems to reduce the risk of local recurrence, but its impact on overall survival specifically for retroperitoneal sarcomas has not been clearly established by available studies.

The histological type of the tumor represents the most important prognostic factor in retroperitoneal sarcoma. It is closely associated with rates of local recurrence, metastases, and overall survival. Well-differentiated liposarcomas generally have a better prognosis, with lower metastatic potential compared to other subtypes. However, even after resection, they may present with local recurrences. Conversely, the myxoid subtype, which is the most frequent histologically, is clinically more aggressive, with a tendency to recur rapidly and a poorer prognosis. Pleomorphic, mixed, and dedifferentiated liposarcomas are also associated with an unfavorable prognosis. Other important prognostic factors include resection status (complete or not), presence of synchronous metastases, as well as neurovascular and osseous involvement. These elements must be taken into account in prognosis evaluation and treatment planning for patients with retroperitoneal sarcomas.

CONCLUSION:

Retroperitoneal sarcomas are rare tumors of mesenchymal tissue, with liposarcoma being the most frequent subtype. Symptoms, often nonspecific, and diagnosis rely on imaging, notably CT scan and MRI, confirmed by biopsy. Surgical resection remains the cornerstone of therapy, aiming for complete resection, although local recurrence remains common. Perioperative radiotherapy and chemotherapy seem to have a role; however, their benefit remains debated. The histological type of the tumor is crucial for prognosis, with well-differentiated liposarcomas having a better prognosis than myxoid or dedifferentiated forms. A multidisciplinary approach is essential for optimal management of these tumors.

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