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Uncommon Presentation Of A Giant Retroperitoneal Liposarcoma: A Peculiar Case Study

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Abstract:
Introduction: *Soft tissue sarcomas constitute less than 1% of all neoplasms, with approximately one-third of malignant tumors in the retroperitoneum being sarcomas. Among these, liposarcoma emerges as the most prevalent type. Retroperitoneal liposarcomas with intraperitoneal extension and extension into the inguinal canal are relatively uncommon, presenting a challenge for treatment due to their local aggressiveness and clinical nonspecificity. The clinical difficulty in managing these cases is exacerbated by patients often seeking medical attention late in the disease progression. The slow development and the scarcity of symptoms in the early stages contribute to delayed reporting. Consequently, these tumors are recognized for their capacity to reach enormous sizes before being diagnosed and addressed.*
Case Report: *We present a case involving a 52-year-old man who underwent surgical resection for a massive, well-differentiated retroperitoneal liposarcoma. This tumor exhibited intraperitoneal extension and weighed a substantial 6.8 kilograms.*
Discussion: *Retroperitoneal liposarcomas (RPS) frequently manifest as large, locally advanced lesions. The occurrence of RPS with intraperitoneal extension and extension into the inguinal canal is relatively uncommon. The clinical presentation is often delayed, attributed to the expansive nature of the retroperitoneal space. Evaluating these tumors necessitates a comprehensive, multimodal approach. Given that most RPS are characterized as low-grade, distant metastasis is infrequent, and the primary challenge lies in achieving effective local control and preventing recurrence. The preferred treatment involves radical resection of these tumors, incorporating en bloc resection of involved structures. In certain cases, the strategic use of neoadjuvant and adjuvant therapies, tailored to specific tumor histologies, may enhance local control and overall survival.*
Conclusion: *Effectively managing retroperitoneal liposarcomas necessitates a multidisciplinary approach, ideally conducted at high-volume centers that specialize in the treatment of patients facing these intricate malignancies. Existing data indicate that radical resection stands as the primary opportunity for a cure. In select cases, chemotherapy and radiation therapy may offer a survival benefit, underlining the importance of individualized treatment strategies based on the specific characteristics of the tumor and the patient.*
Keywords: *Giant Retroperitoneal Liposarcoma (RPS), intraperitoneal extension, local recurrence*

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I. Introduction

Soft tissue sarcomas have their origins in mesenchymal cells and constitute less than 1% of all neoplasms. In the retroperitoneum, where one-third of malignant tumors emerge as sarcomas, liposarcoma stands out as the most common subtype.1 The incidence of retroperitoneal liposarcomas (RPS) is relatively rare, ranging from 0.3% to 0.4% per 100,000 population, with the peak occurrence observed in the fifth decade of life. Retroperitoneal liposarcomas with intraperitoneal extension pose a unique challenge due to their local aggressiveness and clinical nonspecificity. Patients often delay seeking medical attention, contributing to late-stage diagnoses.2 The slow progression and limited symptoms in the early stages allow these tumors to attain enormous sizes before detection. In this context, we present a case involving a giant retroperitoneal adipocytic liposarcoma with intraperitoneal extension, weighing an impressive 6.8 kilograms. This case underscores the complexities and challenges associated with the management of such tumors.

II. Case Report

A 52-year-old male presented with a 6-month history of painless, progressively enlarging abdomino-scrotal swelling. The swelling was associated with abdominal distension and extended into the left scrotum. There was no reported history of fever, vomiting, trauma, dyspnea, melena, or alteration of bowel habits. Upon examination, a sizable abdominal lump, measuring approximately 15 x 9 inches, was evident, with extension into the left scrotum. This clinical presentation suggests a significant abdominal mass with scrotal involvement, necessitating further evaluation and diagnostic investigation to determine the underlying cause and guide appropriate management.

Contrast-enhanced computed tomography of the entire abdomen revealed a substantial lobulated soft tissue density within the peritoneal cavity, displaying internal hyperdense septation. This mass extended along the left inguinal region up to the root of the scrotum, causing displacement of surrounding bowel loops and pushing the urinary bladder to the right. The radiological impression suggested a probable intraperitoneal lipoma (Figure 1).

Fine needle aspiration cytology was performed, revealing a few stromal fragments and plump spindle-shaped cells displaying mild pleomorphism, indicative of a spindle cell lesion.3 Subsequently, the patient underwent an exploratory laparotomy with excision of both intraperitoneal and extraperitoneal masses under general anesthesia. Intraoperatively, a large lipomatous mass was observed, occupying almost the entire peritoneal cavity. The mass infiltrated the large bowel and jejunoileal mesentery up to the retroperitoneum, extended into the pelvic cavity up to the sacral promontory, and protruded into the left hemiscrotum through the deep inguinal ring (Figure 2). This comprehensive surgical exploration aimed to address and remove the extensive lipomatous involvement identified during the procedure. In the histopathological examination (Figure 3), multiple sections reveal a highly pleomorphic and cellular tumor. The tumor cells exhibit large, round to spherical morphology with vacuolated cytoplasm. The nuclei are hyperchromatic, and in some areas, they appear bizarre. Notably, some tumor cells exhibit rhabdoid differentiation. Despite the cellular nature of the tumor, the mitotic count is very low. Based on these findings, the impression is consistent with well-differentiated adipocytic liposarcoma. This characterization provides crucial information about the nature and differentiation of the tumor, guiding further decisions regarding treatment and prognosis.

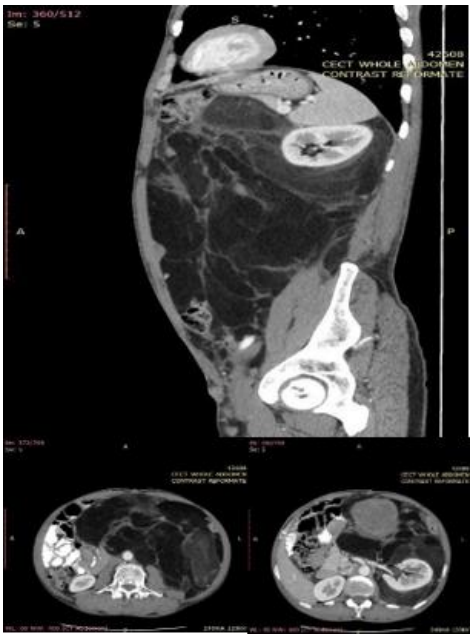

Figure 1: Pre- Op Contrast Enhanced Computed Tomography Whole Abdomen

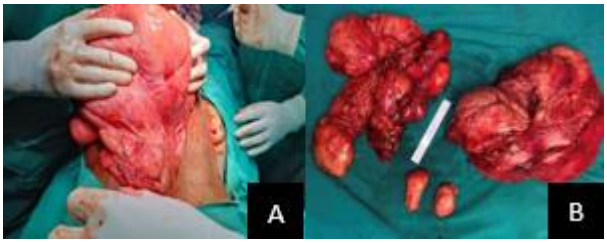

Figure 2: Intraoperative Images

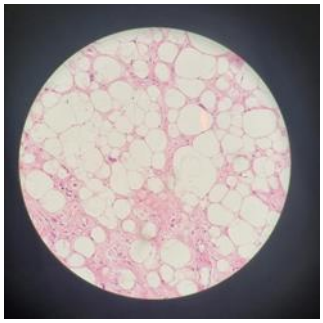

Figure 3: Post-Op HPE

III. Discussion

Retroperitoneal liposarcomas are characterized by their slow growth, and due to the expansive nature of the retroperitoneal space, these tumors often do not cause noticeable signs or symptoms early on. As a result, they have the potential to reach a significant size before detection, commonly manifesting through increased abdominal girth, the presence of a palpable lump, or the compression of nearby structures, leading to gastrointestinal, urologic, or neurological symptoms. Typically, retroperitoneal liposarcomas present with nonspecific symptoms such as abdominal discomfort, back pain, and alterations in bowel or urinary habits. The nature of these symptoms can make diagnosis challenging, and patients may not seek medical attention until the tumor has grown considerably. In many cases, retroperitoneal liposarcomas are discovered incidentally during routine physical examinations or through imaging studies conducted for unrelated reasons. This highlights the importance of thorough diagnostic investigations, especially in cases where nonspecific symptoms are present, to identify and address these tumors in a timely manner.4,5

Abdominal radiographs may provide indirect indications such as bowel displacement and signs of calcification in the tumoral mass, which could suggest teratoma. However, they are generally insufficient for a definitive diagnosis. Ultrasound is of limited value in the detailed assessment of retroperitoneal masses. The diagnostic modality of choice for comprehensive evaluation is contrast-enhanced computed tomography (CT) of the abdomen and pelvis.

Magnetic resonance imaging (MRI) is typically reserved for cases where there is suspicion of neurovascular and muscular invasion. Both MRI and ultrasound can be valuable complements to CT in characterizing indeterminate liver lesions.6-9 The combined use of CT and MRI aids in determining crucial aspects of the tumor, including its anatomical location, size, probable origin, relationship to adjacent visceral and neurovascular structures, potential compression or invasion, and the presence or absence of transperitoneal spread or metastasis to the liver or lungs. In cases where there is concern or a perceived risk of distant metastasis, further staging is recommended, often involving a contrast-enhanced CT scan of the chest. This comprehensive imaging approach is instrumental in achieving a thorough understanding of the tumor's characteristics, facilitating accurate diagnosis, and guiding appropriate treatment strategies.10

F-Fluorodeoxyglucose (FDG)-positron emission tomography (PET)-computed tomography (CT) scans serve as a valuable tool in distinguishing between high-grade and low-grade retroperitoneal liposarcomas. Beyond grading, this imaging modality plays a crucial role in staging by detecting potential metastases. Additionally, FDG-PET-CT aids in restaging and evaluating treatment responses, as well as in post-surgical follow-ups, where it can identify residual masses or recurrences following radical surgery for retroperitoneal liposarcomas.

Despite the significant contribution of FDG-PET-CT to preoperative assessment and postoperative surveillance, the definitive determination of the extent of tumor involvement with adjacent organs and structures is ultimately established during surgery. Surgical exploration remains a critical component in the comprehensive evaluation and management of retroperitoneal liposarcomas, providing a firsthand understanding of the tumor's relationship with surrounding structures and guiding appropriate surgical strategies.

Similarly, Bonvalot et al. reported that extended resection in 120 retroperitoneal liposarcoma patients led to a 3.3-fold lower local recurrence rate compared to 65 patients who had simple complete resection. The 3-year local recurrence rate was 10% for extended resection, significantly lower than the 47% observed for simple complete resection. These findings suggest that a more extensive surgical approach, involving the removal of adjacent organs and structures when implicated, may contribute to a reduced risk of local recurrence in retroperitoneal liposarcoma patients. United States critics pointed out that for both studies, overall survival was equivalent in patients that underwent extended versus standard resection. Longer follow-up data by Gronchi et al.11 did in fact show improvement in survival for low and intermediate but not high grade retroperitoneal liposarcoma. These findings along with the identification of microscopic organ invasion in the absence of a clear macroscopic involvement have led more recently to an agreed definition of the optimal extent of surgical resection in primary retroperitoneal liposarcoma: surgery should be aimed at achieving macroscopically complete resection

with a single specimen encompassing the tumor and involved contiguous organs while attempting to minimize microscopically positive margins.

In the management of retroperitoneal liposarcoma, surgical intervention remains the primary treatment, aiming for complete resection to maximize the chances of cure. However, recent research has delved into alternative strategies to enhance treatment outcomes and address challenges associated with this rare malignancy. Neoadjuvant radiation therapy has emerged as a promising approach, offering the advantage of minimizing radiation toxicity to abdominal viscera and vital structures by displacing them with the tumor.12 Studies, including the notable STRASS-1 trial, have investigated the impact of neoadjuvant radiation followed by surgery compared to surgery alone. The trial suggests potential benefits for certain histologic subtypes, such as well-differentiated liposarcoma (WDLPS) and low-grade de-differentiated liposarcoma (DDLPS), emphasizing the need for personalized treatment approaches based on tumor characteristics. These efforts represent a shift toward comprehensive strategies that not only address the challenges of surgical resection but also explore innovative modalities to improve the overall management of retroperitoneal liposarcoma.

Chemotherapy plays a nuanced role in the management of retroperitoneal liposarcoma. Adjuvant chemotherapy, while not significantly impacting recurrence rates, is selectively reserved for high-grade tumors with heightened metastatic potential. In the perioperative setting, chemotherapy serves the purpose of targeting micrometastatic disease and facilitating downsizing of tumors to increase the likelihood of achieving R0 resections.

Anthracycline-based chemotherapy, particularly with agents like Doxorubicin, takes precedence as the first-line treatment for advanced or metastatic liposarcoma.13 In the realm of second-line agents, Trabectedin demonstrates primary benefits, especially in the context of the myxoid liposarcoma histologic subtype. Eribulin has received approval for use in liposarcomas and leiomyosarcomas, broadening the therapeutic options.

The introduction of Palbociclib, a selective CDK4/CDK6 inhibitor, presents a notable advancement, showing favorable outcomes in terms of progression-free survival. However, the full spectrum of anticancer agents' efficacy in retroperitoneal liposarcoma treatment, including tyrosine kinase inhibitors or gemcitabine/docetaxel combinations, remains to be clarified.14 Ongoing research endeavors seek to elucidate the potential roles of these agents, contributing to a more comprehensive understanding of their place in the evolving landscape of retroperitoneal liposarcoma management.

Immunotherapy has emerged as a promising frontier in the treatment landscape for retroperitoneal liposarcoma (RPLPS), offering targeted approaches to disrupt key molecular pathways associated with the disease's growth and progression. The use of MDM2 inhibitors, capitalizing on the established role of MDM2 amplification and subsequent p53 inhibition, represents a therapeutic strategy to impede RPLPS development. Additionally, in the realm of CDK4 inhibitors, particularly in de-differentiated RPLPS, amplified sequences from the 12q13-15 chromosomal region containing CDK4 genes present opportunities for drugs like Palbociclib, Ribociclib, and Abemaciclib.

Aurora kinase inhibitors are explored due to altered AURKA expression in liposarcomas, offering a potentially encouraging avenue for therapeutic intervention. The focus on receptor tyrosine kinase (RTK) genes, amplified in a significant proportion of well-differentiated and de-differentiated retroperitoneal liposarcoma samples, underscores the relevance of inhibiting specific RTKs. Notably, multi-targeted tyrosine kinase inhibitors such as Ponatinib and Pazopanib are actively employed in this context.15 These immunotherapeutic strategies signal a shift towards precision medicine, aiming to tailor treatments based on the molecular characteristics of the tumor. Ongoing research endeavors are imperative to comprehensively understand the efficacy and potential of these immunotherapeutic interventions in the complex landscape of retroperitoneal liposarcoma.

In the realm of retroperitoneal liposarcoma, the exploration of alternative local treatments adds depth to the therapeutic landscape. Ablation techniques like cryotherapy and radiofrequency ablation offer targeted approaches for eliminating cancerous tissue, providing localized interventions. Similarly, embolization methods, including chemoembolization and radioembolization, focus on restricting the tumor's blood supply as part of a comprehensive treatment strategy.

Post-treatment, patients undergo a structured follow-up regimen involving regular physical examinations and imaging studies. The frequency of these assessments evolves over time, with intervals of every 3-6 months in the initial 2-3 years, followed by semi-annual checks for the subsequent 2 years, and eventually transitioning to yearly assessments. The prognosis for retroperitoneal liposarcoma is multifaceted, influenced by factors such as age, tumor characteristics (site, depth, size, resectability), histological subtype, grade, presence of nodal disease, and evidence of distant metastasis. The ongoing evaluation of these prognostic factors informs tailored treatment plans and guides long-term management strategies for individuals affected by this challenging malignancy.

IV. Conclusion

Effectively addressing the complexities of retroperitoneal liposarcomas requires a multidisciplinary approach, ideally implemented at specialized high-volume centres dedicated to managing these intricate malignancies. Current evidence underscores radical resection as the primary avenue for achieving a potential cure. In certain cases, the incorporation of chemotherapy and radiation therapy may contribute to a survival benefit, highlighting the significance of tailoring treatment strategies to the unique characteristics of both the tumor and the individual patient. This comprehensive and individualized approach is essential for optimizing outcomes and navigating the challenges posed by retroperitoneal liposarcomas. Immunotherapy has emerged as a promising frontier in the treatment of retroperitoneal liposarcoma (RPLPS), presenting targeted approaches to disrupt key molecular pathways associated with the growth and progression of the disease. This innovative therapeutic strategy holds the potential to enhance the precision and effectiveness of treatments for RPLPS, marking a significant advancement in the ongoing quest for improved outcomes in the management of this challenging malignancy.

Conflicts of Interest

The authors declare no conflicts of interest

Disclosures and Funding

None

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