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A CASE STUDY ON RETROPERITONEAL TUMOR: A RARE PATHOLOGY

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ABSTRACT

Introduction: Retroperitoneal tumors (RPTs) are extremely rare tumors, predominant in both sexes and can be malignant or benign. Due to the inaccessibility of the region and the asymptomatic or nonspecific presentation, these tumors most often reach a substantial size. The most frequent symptoms are abdominal pain, discomfort, or a painless palpable mass in the abdomen. In addition to the patient's clinical history, imaging tests are essential for diagnosis. Complete surgical resection is the only potential curative treatment modality for RPTs. Discussion: Benign RPTs are approximately four times less frequent than malignant tumors, although they often manifest clinically - which makes our patient's case even more atypical. The pathological stage is highly correlated with the prognosis of cancer and, for an accurate assessment, high-quality cross-sectional images play a fundamental role. Conclusion: The successful prognosis of RPTs is influenced by multiple factors. The grade of the tumor is of paramount importance, in addition to its intrinsic biology, together with the extent of surgical resection performed, playing crucial roles in determining prognosis and life expectancy. Therefore, performing detailed imaging exams and specific laboratory tests, when possible, are crucial.

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INTRODUCTION

Retroperitoneal Tumors (RPTs) are extremely rare tumors located in the retroperitoneal space, bounded anteriorly by the posterior parietal peritoneum and posteriorly by the transversalis fascia. These tumors can be benign or malignant. Malignant tumors originating from epithelial tissues are classified as cancers, while those arising from non-epithelial tissues are sarcomas, which are more prevalent. (CHOI and RO, 2020) Soft tissue sarcomas account for 1-2% of all solid malignant tumors, with 10% occurring in the peritoneum, marking their notable rarity. The most common types are liposarcomas (specifically well-differentiated and dedifferentiated subtypes) and leiomyosarcomas. (CHOI and RO, 2020) (SANTOS et al., 2005) (METTLIN, 1982)

The incidence of RPTs is approximately 0.3-0.4 cases per 100,000 inhabitants in the United States. While modest, this figure underscores the clinical relevance of these neoplasms, whose diagnosis and treatment often challenge the expertise of modern anatomists and pathologists. In Brazil, specific data on the incidence of retroperitoneal tumors are scarce, reflecting underreporting and a lack of systematic records. (METTLIN et al., 1982) Although not displaying a pronounced sex preference, retroperitoneal tumors predominantly occur in adults, typically between the fourth and seventh decades of life, possibly reflecting the accumulation of environmental and genetic factors over time. This age distribution suggests a complex interaction between aging and the biological events underlying the genesis of these neoplasms. (METTLIN et al., 1982)

The retroperitoneum is an anatomical region prone to tumor development due to its diverse tissue composition. (MOORE and DALEY, 2014) Although the etiology of RPTs remains unknown, genetic and environmental factors, radiation exposure, viral infections, and immunodeficiency may be associated with their development. (CHOI and RO, 2020) Pathologically, neoplastic cells change their regulatory mechanisms of multiplication and metabolism, acquire growth autonomy, and become independent of physiological stimuli. Consequently, cellular activities are deemed constructive, manifesting continuously and without regulation. (SANTOS et al., 2007)

RPTs originate primarily from soft tissues, including fat, muscles, nerves, lymph nodes, and blood or lymphatic vessels. They may involve retroperitoneal organs such as the kidneys, adrenal glands, pancreas, and pelvic organs (bladder, uterus, ovaries, prostate, etc.). Invasive RPTs can also affect major retroperitoneal structures, such as the abdominal aorta, inferior vena cava, and ureters. (CHOI and RO, 2020)

Due to the inaccessibility of the region and often asymptomatic or nonspecific presentation, these tumors usually reach substantial sizes. Thus, they are generally large at

the time of diagnosis. (STRAUSS et al., 2011) Nearly 50% are larger than 20 cm at diagnosis. Secondary symptoms of retroperitoneal lesions appear late in the disease course and are associated with organ displacement and obstructive phenomena. (CHOI and RO, 2020) Common symptoms include abdominal pain, discomfort, or a painless palpable abdominal mass. Moderate fever and mild leukocytosis may occur due to the central necrosis of large tumors. (SANTOS et al., 2007) Diagnosing retroperitoneal tumors is challenging due to overlapping morphological features with other tumor types and the increasing use of minimally invasive biopsy techniques, which provide limited tissue samples. (CHOI and RO, 2020)

In addition to the patient's clinical history, which may assist in identifying potential differential diagnoses (CHOI and RO, 2020), imaging studies are essential for diagnosis. Retroperitoneal tumors are best evaluated using high-quality cross-sectional imaging, and preoperative histology through core-needle biopsy is necessary when imaging is non-diagnostic. (STRAUSS et al., 2011) The imaging modality of choice is contrast-enhanced computed tomography (CT) of the chest, abdomen, and pelvis, which provides details about the size, location, relationship to adjacent organs, and the presence or absence of metastases. (STRAUSS et al., 2011) Magnetic resonance imaging (MRI) or PET scans may also be used. (SASSA, 2020)

Fat-containing tumors can be easily identified using MRI or CT; however, well-differentiated liposarcomas are indistinguishable from lipomas, appearing as well-defined lesions predominantly composed of fat with minimal soft-tissue attenuation. In such cases, needle biopsy is increasingly valuable for definitive diagnosis and for determining histological subtypes and classification. (SASSA, 2020)

CT-guided biopsy techniques allow for accurate and safe diagnosis of retroperitoneal tumors, despite the potential risk of tumor seeding at the biopsy site. The efficacy of imaging-guided fine-needle aspiration cytology (FNAC) has also been reported, with overall sensitivity, specificity, and diagnostic accuracy of 98.02%, 72.22%, and 94.12%, respectively. However, needle biopsy is recommended over FNAC by the European Society for Medical Oncology Clinical Practice Guidelines, the European Network for Rare Adult Solid Cancers, and the National Comprehensive Cancer Network Clinical Practice Guidelines. According to the Trans-Atlantic RPS Working Group, FNAC rarely provides diagnostic information, causes treatment delays, and should be avoided. (SASSA, 2020)

When determining histological type, diagnostic accuracy can be enhanced by genomic methods that identify specific mutations in individual sarcoma genes, miRNA differentiation, and translocation confirmation using fluorescence in situ hybridization,

irrespective of morphological diagnosis. Needle biopsy to confirm grading systems allows for estimating the prognosis of each RPS subtype and the cure rates associated with R0/R1 surgical resection. Benign RPTs do not require treatment if definitive tumor histology can be diagnosed preoperatively via biopsy and/or imaging. However, symptomatic tumors or those with rapid growth should undergo surgical resection to distinguish benign from malignant tumors. (SASSA, 2020)

In retroperitoneal sarcomas (RPSs), post-treatment tumor status is described by residual tumor classification (R). R0 resection indicates microscopic margin-negative resection with no macroscopic or microscopic tumor remaining at the primary RPS site. R1 resection involves removing all macroscopic diseases but with microscopic positive margins. R2 resection indicates residual gross disease with macroscopic tumor involvement. (SASSA, 2020) Complete surgical resection is often unachievable due to the vital structures involved. (CHOI and RO, 2020)

Complete surgical resection is the only potentially curative treatment modality for RPS, but local recurrence occurs in a significant proportion of patients, accounting for up to 75% of sarcoma-related deaths. Resection of adjacent organs is often necessary, with visceral resection rates reported between 34% and 93% in large series. (STRAUSS et al., 2011) Surgical strategies prioritize en bloc resection of the sarcoma and contiguous organs macroscopically involved or encased by the tumor to achieve complete macroscopic clearance. Organs merely adjacent to the tumor without involvement are spared. Commonly resected organs include the colon, kidney, pancreas, and spleen. (STRAUSS et al., 2011) Achieving R0/R1 surgical resection improves cure rates and prevents locoregional recurrence. Other factors influencing RPS prognosis include favorable tumor histology or biology, advancements in multimodal treatments, perioperative management, surgical techniques, and patient biology. (SASSA, 2020)

The use of radiotherapy in the treatment of RPS, alongside surgical treatment, has shown favorable results in a combined case-control study using data from the national oncology clinical database jointly managed by the American Cancer Society and the American College of Surgeons. However, specific irradiation methods, doses, and intervals were not detailed in the database. To assess the effectiveness of perioperative radiotherapy, a randomized multicenter controlled trial in Europe compared preoperative radiotherapy followed by surgery with isolated surgery for primary treatment. However, the results of the 2019 study did not demonstrate the utility of preoperative radiotherapy. Other clinical trials involving radiotherapy are ongoing. (SASSA, 2020)

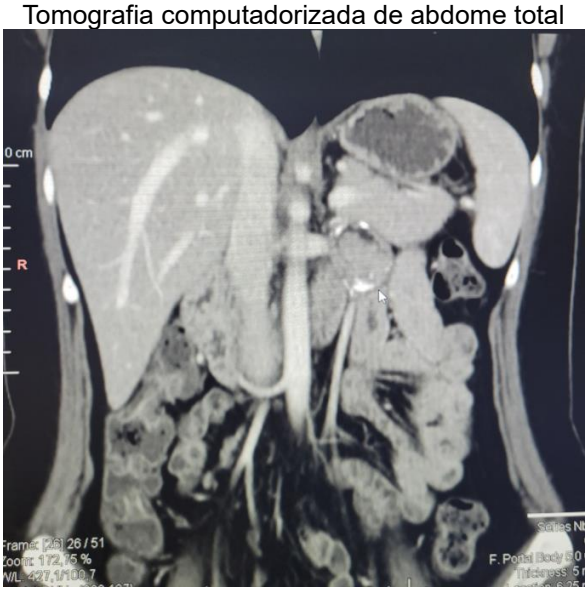
Regarding the use of systemic drugs in the treatment of RPS, doxorubicin has been a key medication in the systemic treatment of RPS for approximately 30 years. Ifosfamide and other drugs have also been used in combination with doxorubicin. Chemotherapy choices are used separately to prolong life and reduce preoperative tumor size. Adjuvant postoperative chemotherapy with doxorubicin and ifosfamide has not been recommended based on the results of a randomized clinical trial. (EORTC 62931) (SASSA, 2020)

In RPTs, the five-year survival rate for all subtypes is approximately 60%. Factors such as tumor grade, which reflects both aggressive metastatic behavior and increased local recurrence incidence in high-grade sarcomas, tumor biology, and the extent of resection impact survival. Surgical resection is the most important predictor of survival, with incomplete resection conferring a 70% higher risk of death. (FILHO, 2021)

CASE DESCRIPTION

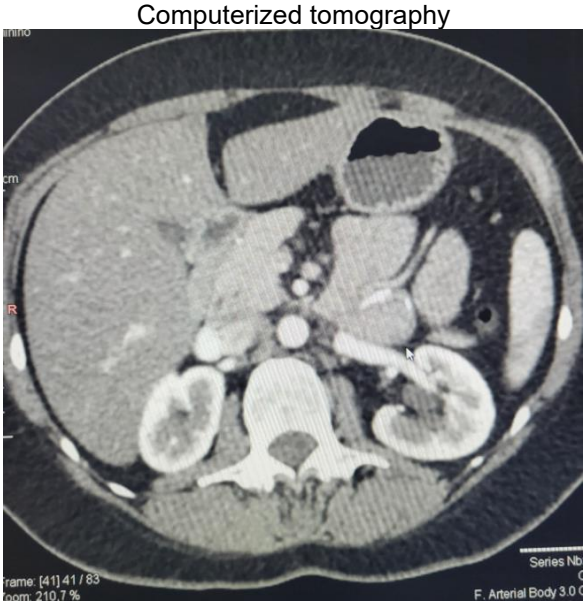
Patient C.N.F., female, 45 years old, smoker, with no comorbidities and asymptomatic. She reported having a fractured coccyx and that, during imaging tests, a lesion/tumor was found in the retroperitoneum, without being able to differentiate whether it was cancer or not. She was referred for evaluation by a gastroenterologist. She denied pain, nausea, vomiting, and/or malaise. The patient was admitted for an elective biopsy guided by computed tomography, but after discussing the case and due to difficult access, it was not possible to perform it. The patient was electively admitted for tumor excision due to suspected lymphoma. The stomach was released, accessing the abdominal retrocavity, without access to the lesion. A Mantoux maneuver was attempted, with the release of the left colon and access to the lesion from both planes. The abdominal cavity inventory revealed the presence of a retroperitoneal lesion below the pancreas with anterior limit: inferior mesenteric vein, superior mesenteric vein; upper limit: renal vein; and posterior limit: inferior vena cava. Laparotomy was performed with oncological retroperitoneal lymphadenectomy with lymph node dissection and omentectomy for better access to the lesion. The nodule in the retroperitoneum was resected and taken for biopsy, which resulted in a benign non-neoplastic cyst measuring 4.5/3.5/2.5 cm. The procedure was performed without complications. The patient evolved hemodynamically stable, without VAD, without nausea and vomiting, and with little pain. He was discharged after 4 days of hospitalization.

Tomografia computadorizada de abdome total



Images provided by Dr. Fernando Pereira de Almeida, Santa Casa Hospital

Computerized tomography



Images provided by Dr. Fernando Pereira de Almeida, Santa Casa Hospital.

DISCUSSION

RPTs have no gender predilection and mainly affect adults between the fourth and seventh decades of life, a period in which the patient falls right at the beginning. (METTLIN, 1982) The clinical picture presented by the patient was asymptomatic, which can occur in this type of tumor and contributes to the late discovery of the condition and with a mass that is already very evolved. However, routine imaging examination allowed the tumor to be found before it evolved to a substantial size, which occurs in half of the cases. (STRAUSS et al. 2011) Benign RPTs are approximately four times less frequent than malignant tumors, although they often manifest clinically - which makes the patient's case even more atypical. According to the literature, the largest number of benign RPTs was found in younger

patients, such as the case discussed. (SASSA, 2020) The pathological stage is highly correlated with the prognosis of cancer and, for an accurate assessment, high-quality cross-sectional images play a fundamental role. When tumor characteristics are not identified through imaging, preoperative histology through core needle biopsy is crucial to establish a definitive diagnosis. (STRAUSS et al. 2011) (NUSSBAUM et al. 2014) (FILHO, 2021) However, in the reported case, the patient had a lesion that was difficult to access, which prevented CT-guided biopsy, and, for this reason, surgery was chosen in this case. (SASSA, 2020) The diagnostic hypothesis was confirmed through POI tumor excision in the retroperitoneum, followed by retroperitoneal lymphadenectomy and omentectomy. With confirmation of the diagnosis, the medical team can safely proceed to planning an adequate prognosis.

Improved survival and prevention of locoregional recurrence can be achieved by performing surgery to ensure negative margins with wide and combined resection of some adjacent organs. The high volume of surgeons and specialized centers is associated with better outcomes for patients in major oncological surgeries, including hepatobiliary/pancreatic surgery, esophagogastric surgery, and surgical oncology, therefore, cooperation with a trained medical team composed of radiologists, pathologists, and medical oncologists in a centralized center positively reflects on the patient's prognosis. (STRAUSS et al. 2011) (NUSSBAUM et al. 2014)

METHODOLOGY

From a medical case that occurred at Santa Casa da Misericórdia de Presidente Prudente, together with its medical records, information was collected to carry out this case report. In addition, a search for information in databases was performed.

CONCLUSION

It is concluded that the success of the prognosis of retroperitoneal tumor is influenced by multiple interrelated factors. Tumor grade is of paramount importance, as it reflects not only the aggressive metastatic behavior but also the higher incidence of local recurrence observed in high-grade sarcomas. Furthermore, the intrinsic biology of the tumor, together with the extent of surgical resection performed, plays crucial roles in determining the prognosis and life expectancy of patients. Therefore, performing detailed imaging exams and specific laboratory tests are crucial, especially in at-risk populations; adjuncts to monitoring by a multidisciplinary medical team. These procedures are

fundamental for initial diagnosis and surgical planning, and the precise implementation of the steps is vital to optimize the chances of recovery.

CONFLICT OF INTEREST

The authors agree that there was no conflict of interest in the course of this case report.

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