Giant Retroperitoneal Liposarcoma: Case Report

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Lipossarcoma Retroperitoneal Gigante: Relato de Caso

Liposarcoma Retroperitoneal Gigante: Informe de Caso

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ABSTRACT

Introduction: Liposarcomas are rare malignant tumors of mesenchymal origin, from adipocyte precursors, which may occur in the retroperitoneum. Retroperitoneal liposarcomas account for 0.3% to 0.6% of malignant neoplasms and the main symptom is increased abdominal volume. Due to the slow and asymptomatic growth of the tumor, the condition is commonly associated with late diagnosis and indolent course. **Case report:** 70-year-old man with progressive abdominal enlargement for about two years. Magnetic resonance imaging demonstrated a formation in the right retroperitoneal topography, promoting significant deviation of the midline structures. Furthermore, there was a solid content above the structure described above. A xiphopubic laparotomy was performed with presentation of giant retroperitoneal mass occupying the entire abdomen, displacing the intestinal loops and the right kidney. En bloc tumor resection, diaphragmatic raffia and cholecystectomy were performed. Three months after the first surgical procedure, the patient presented a novel increase of the abdominal volume which imaging tests showed tumor recurrence. A new surgical procedure was performed, revealing a tumor with a ruptured capsule and multiple clots. After hospital discharge, the patient was submitted to complementary radiotherapy **Conclusion:** The fast return of the patient after the recurrence, common in these cases, was essential to reduce the residues of the second surgery, highlighting the importance of periodic examinations for the early recognition of local recurrence. In this case, radiotherapy sessions were also performed to prevent recurrence, however, unsuccessful.

Key words: liposarcoma; retroperitoneal space; neoplasms; laparotomy.

RESUMO

Introdução: Os lipossarcomas são tumores malignos raros de origem mesenquimal, a partir de precursores de adipócitos, podendo ocorrer no retroperitônio. Os lipossarcomas retroperitoneais representam de 0,3% a 0,6% das neoplasias malignas. Em virtude do crescimento lento e assintomático do tumor, tendo como principal manifestação clínica o aumento do volume abdominal, o quadro é comumente acompanhado de um diagnóstico tardio e curso indolente. **Relato do caso:** Homem, 70 anos de idade, com aumento progressivo abdominal há cerca de dois anos. A ressonância magnética demonstrou uma formação em topografia retroperitoneal à direita, promovendo importante desvio das estruturas da linha média, com presença de conteúdo sólido acima da estrutura supradescrita. Foi submetido à laparotomia xifopúbica, com apresentação de massa gigante retroperitoneal que ocupava todo abdome, com deslocamento das alças intestinais e rim direito. Foram realizadas ressecção do tumor em bloco, rafia diafragmática e colecistectomia. Após três meses do primeiro procedimento cirúrgico, o paciente apresentou novamente aumento do volume abdominal, com confirmação de recidiva tumoral após realização de exames de imagem. Um novo procedimento cirúrgico foi realizado, revelando tumor com cápsula rompida e múltiplos coágulos. Após alta hospitalar, foi submetido a sessões de radioterapia complementares à cirurgia. **Conclusão:** A rápida reapresentação do paciente após o surgimento da recidiva, comum nesses casos, foi essencial para a redução de resíduos na segunda cirurgia, evidenciando a importância de exames periódicos para o reconhecimento precoce da recorrência local. No presente caso, também foram realizadas sessões de radioterapia, com a finalidade de evitar a recidiva, sem sucesso.

Palavras-chave: lipossarcoma; espaço retroperitoneal; neoplasias; laparotomia.

RESUMEN

Introducción: Los liposarcomas son tumores malignos raros de origen mesenquimatoso, a partir de precursores de los adipocitos, y pueden presentarse en el retroperitoneo. Los liposarcomas retroperitoneales representan del 0,3% al 0,6% de las neoplasias malignas, siendo el síntoma principal el aumento de volumen abdominal. Debido al crecimiento lento y asintomático del tumor, la condición se acompaña comúnmente de un diagnóstico tardío y un curso indolente. **Informe del caso:** Varón, 70 años, con agrandamiento abdominal progresivo de unos 2 años de evolución. La resonancia magnética nuclear mostró una formación en la topografía retroperitoneal hacia la derecha, promoviendo una desviación significativa de las estructuras de la línea media. Además, se observó un contenido sólido por encima de la estructura descrita anteriormente. Considerando la principal hipótesis diagnóstica, liposarcoma de retroperitoneo, se realizó laparotomía, evidenciándose una masa retroperitoneal gigante que ocupaba todo el abdomen, que desplazaba las asas intestinales y el riñón derecho. Se realizó disección y resección del tumor en bloque para extirpar la masa tumoral, así como rafia diafragmática y colecistectomía. A los tres meses del primer acto quirúrgico, el paciente volvió a presentar aumento de volumen abdominal, en el que las pruebas de imagen mostraron recidiva tumoral. Se realizó un nuevo procedimiento quirúrgico que reveló un tumor con una cápsula rota y múltiples coágulos. **Conclusión:** El rápido retorno del paciente tras el inicio de la recidiva, frecuente en estos casos, fue fundamental para reducir los residuos en la segunda cirugía, destacando la importancia de las exploraciones periódicas para el reconocimiento precoz de la recidiva local. En este caso también se realizaron sesiones de radioterapia, con el objetivo de prevenir la recurrencia, sin éxito.

Palabras clave: liposarcoma; espacio retroperitoneal; neoplasias; laparotomia.

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INTRODUCTION

Liposarcomas are rare malignant tumors with mesenchymal origin and from adipocyte precursors. They are usually found in the retroperitoneum, approximately 30%, and in the extremities (hands, feet, and elbows). Retroperitoneal liposarcomas represent 0.3% to 0.6% of malignant neoplasms. Its main clinical manifestation is the increase in abdominal volume, usually painless, requiring imaging tests for diagnostic definition[1-3].

Tumor aggressiveness is determined by the degree of differentiation. The treatment of choice is surgery, aiming at resection of the tumor and with safety surgical margins. The local recurrence rate is 60% to 70% due to late diagnosis and the complexity of tumor localization[1,4]. This report presents a case of giant retroperitoneal liposarcoma, with emphasis on clinical history, diagnostic methods and surgical techniques performed.

CASE REPORT

A 70-year-old man sought consultation in a clinical oncology outpatient clinic at Beneficent Association Santa Casa de Campo Grande, Campo Grande-MS, in May 2020, complaining of progressive abdominal enlargement for two years, even when performing dietary measures. He denied abdominal pain complaints, changes in bowel habit, nausea, and vomiting. He reported daily headache with improvement after analgesia. Patient with no previous comorbidities. History of umbilical hernioplasty for about three years. Computed tomography (CT) of the abdomen showed a heterogeneous lesion on the right, measuring 40 x 27 x 29 cm, without invasion of adjacent structures, but with great displacement of them. In view of the clinical picture and the image suggestive of retroperitoneal liposarcoma, the patient was hospitalized for complementary tests for diagnostic confirmation and staging.

CT scan of the skull and chest without significant changes. Magnetic resonance imaging (MRI) of the abdomen showed lobulated formation, with components of soft tissues and fatty content, located in retroperitoneal topography on the right, having inaccurate dimensions, measuring as a whole about 14.0 x 9.9 x 24.0 cm (Figure 1), promoting an important deviation of the midline structures contralaterally. Solid content was also noted along the right flank, establishing contact with the structure described above, with minimum dimensions of 12.5 x 8.8 x 23.4 cm in its largest axes. He underwent xiphopubic laparotomy. A giant retroperitoneal mass was identified that occupied the entire abdomen with displacement of the intestinal loops and right kidney

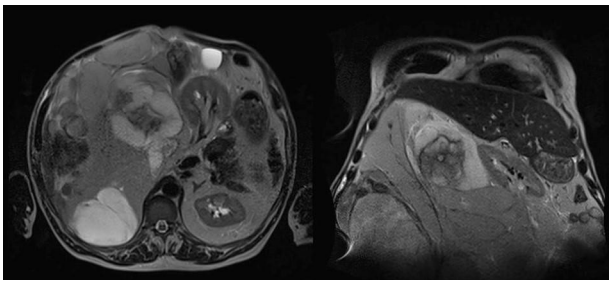

Figure 1. MRI showing lobulated formation located in retroperitoneal topography on the right

to the left flank. The tumor was resected in block, requiring diaphragmatic raffia, because the diaphragm was adhered to the tumor mass, followed by cholecystectomy. Abdominal and thoracic cavity drainage was performed (Figure 2). The patient remained in the hospital unit under follow-up by the multidisciplinary team, with good postoperative evolution, remained hemodynamically stable, and was discharged 11 days after surgery.

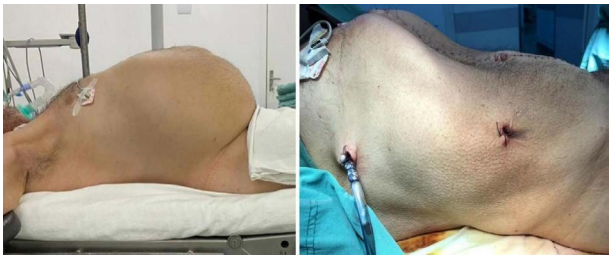

Figure 2. Pre- and postoperative period of the patient. After the first surgical procedure, tumor dissection and resection were performed

The anatomopathological of the surgical specimen confirmed the diagnosis of liposarcoma and described a bulky, well delimited, lobular tumor mass, weighing 15.330 kg and measuring 47.0 x 34.0 x 16.0 cm (Figure 3), presenting a degree of differentiation with score 2. Mitotic count presented score 2 (12 mitoses/10 CGA), necrosis, score 0 (absence of necrosis) and received grade 2 (total score equal to 4 or 5) and surgical margins free of neoplasia. Pathologic staging (pTNM) was classified into pT2b, pNX, and pMX.

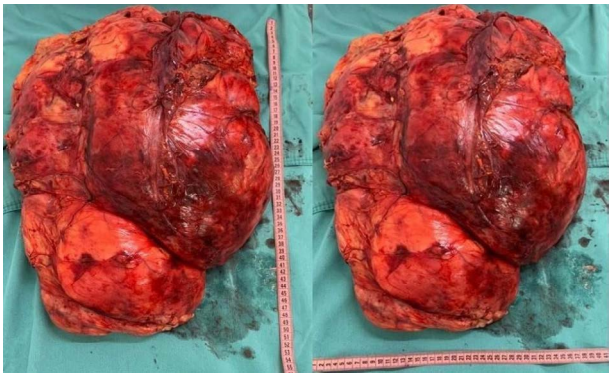

Figure 3. Bulky tumor mass removed after the first surgical procedure

Three months after surgery, the patient complained of increased abdominal volume, hyporexia and constipation. A new abdominal CT scan showed tumor recurrence, with an expansive retroperitoneal lesion on the right, determining great dilation of the collecting system, in addition to a lesion with similar characteristics infiltrating hepatic segments VI and VII, without lymph node involvement. The patient underwent a new surgical procedure. During the re-approach, a tumor with a ruptured capsule, multiple clots and secretion throughout the abdominal cavity were identified, with the presence of a mass adhered to the diaphragm. Cytoreduction was performed, however, due to the impossibility of excision of all tumor fragments, the surgery was considered R2 (surgery with macroscopic residual disease). The pathology of the surgical specimen showed recurrence. The patient presented hemodynamic instability in the trans operative period, requiring vasoactive drug and postoperative period in an Intensive Care Unit (ICU) bed. Because of the gradual improvement, and, after a week, being stable, oriented and without complaints, he was discharged from the hospital. Complementary therapy was chosen – postoperative use of radiotherapy.

This report began after submission and subsequent approval by the Research Ethics Committee (CEP) of the Beneficent Association Santa Casa de Campo Grande, MS, under the number of consolidated opinions 5.528.622 (CAAE: 59789522.4.0000.0134) in compliance with Resolution 466/12[5] of the National Health Council for research with human beings.

DISCUSSION

Retroperitoneal liposarcoma is a malignant tumor, of mesenchymal origin, which represents about 30% of retroperitoneal sarcomas, with a condition commonly accompanied by late diagnosis and indolent course, in view of the slow and asymptomatic growth of the tumor. It is difficult to manage due to the complex access to the retroperitoneal cavity[6,7]. The subdivision consists of three main subgroups: well-differentiated liposarcomas, myxoid, and pleomorphic.

The giant form of retroperitoneal liposarcoma has a very low occurrence, covering tumors of 15 to 20 kg and 20 to 25 cm in diameter. This rare manifestation causes the removal of several tissues and organs from the anatomical position, generating an expansive and heterogeneous lesion[8,9]. In the case presented, the patient followed the increase in abdominal volume for two years. There is still no description of the factors that corroborate the growth of the tumor mass in the proportions presented.

Retroperitoneal liposarcoma has a relatively limited diagnosis, since the displacement of organs from their anatomical positions and the involvement of large vessels make it difficult to determine the exact location of the lesion, pathological type, and extent of invasion[10]. Attention needs to be paid to the displacement of certain vessels and some of their branches. It is necessary to rule out any possibility that the lesion is originating from a retroperitoneal organ, in order to suggest a possible diagnosis of a primary retroperitoneal tumor[7,11]. In the case described, imaging tests contributed substantially to the visualization of the giant mass, involvement and displacement of adjacent structures, suggesting the diagnosis of retroperitoneal liposarcoma, confirmed only with laparotomy.

The International Union against Cancer (UICC) Classification of Malignant Tumors (TNM)[12], the most widely used staging method, has as evaluation criteria the size of the primary tumor, the presence of metastasis in regional lymph nodes and metastasis in distant organs, factors of greater relevance for the common prognosis for most cancers. In the case reported, the pathological evaluation was performed following the pTNM method, in which evidence obtained before treatment, together with that obtained during the surgical procedure and the histopathological examinations performed, provide the basis for classification[12,13]. The tumor mass received the pT2b classification, tumor with 1 to 2 mm thickness with ulceration, pNX, lymph nodes that could not be histologically evaluated, and pMX, with the presence of distant metastasis, but which could not be microscopically evaluated.

The main treatment of patients with retroperitoneal liposarcoma is radical surgery, with the main prognostic factor being total resection of the tumor, which is hampered by the size of the mass, anatomical restrictions of the retroperitoneal cavity and frequent involvement of neighboring structures. The block resection of adjacent organs and retroperitoneal tumor allows the achievement of higher rates of microscopically negative resection margins, in addition to allowing optimal control of the disease site[1]. The surgical management performed in this case report had its resection done in blocks, with the performance of a xiphopubic laparatomy to identify the giant mass, followed by diaphragmatic raffia, cholecystectomy and drainage in the abdomen and chest.

Longitudinal follow-up of the patient is important in view of the high rate of local recurrence, which is 60% to 70%, being higher in patients undergoing resection with a microscopically positive margin[1]. Thus, periodic examinations, such as CT and radiographs, are necessary for the early recognition of local recurrence, since time is decisive for increasing patient survival, as well as the interval between the first surgical procedure and local recurrence[7,15].

The use of postoperative radiotherapy for patients who did not receive it as part of the initial treatment presents beneficial evidence not yet directly correlated due to the difficulty of applying it on large beds in the retroperitoneal cavity and the lack of prospective studies[1,7,14]. In the present case, it was decided to perform radiotherapy sessions after the second surgery, as part of the complementary therapy.

CONCLUSION

The case of giant retroperitoneal liposarcoma presented demonstrated good evolution after the initial laparotomy performed. The rapid re-presentation of the patient after the appearance of recurrence was essential for the second intervention to reduce the residual points, removing the remaining neoplastic foci and the clots adhered to them.

In view of the rarity of this malignant tumor, the search for interventional or prophylactic means to prevent the onset of local recurrence, common in the disease, is of great importance. Based on the case presented, recurrent follow-up is suggested for the early diagnosis of a possible recurrence and prognostic evaluation based on the histopathological study of the tumor and the completeness of the resection performed, in addition to the search for metastases, despite being an unusual finding.

CONTRIBUTIONS

Giovanna Ricarte Granja Gomes, Fernanda Zem Rodrigues de Araujo Costa, Fabio Moraes de Jesus, and Gabriela Caroline de Paula Alcantara contributed substantially to the design and/or planning of the study; data collection, analysis and interpretation; writing and critical review. Diógenes Firmino do Nascimento Neto and Victor de Paula Fonseca contributed substantially to the collection, analysis and interpretation of data, in the writing and critical review. All authors approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interest to declare.

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