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CASE REPORT

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Giant Retroperitoneal Mass: Case Report in a Primary Hospital and Review of the Literature

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ABSTRACT

Retroperitoneal liposarcoma (RPLS) is a rare form of cancer that mostly affects adults. Symptoms may be undetected unless the tumor begins to grow. Diagnosis is made when the tumor is large and puts vital organs such as the pancreas, liver, kidneys at risk. Symptoms of a liposarcoma depend on tumor location. Liposarcoma in the arms, legs, particularly thighs may be painful, swollen, with patients citing weakness in the affected area; 15% of soft tissue sarcomas are retroperitoneal, with two of the most common types being the well-differentiated liposarcoma (WDLPS) and high-grade dedifferentiated liposarcoma (DDLPS). The tumor can weigh between 15 kg and 20 kg, with 20 to 25 cm in diameter (4). Surgery is the main form of treatment provided along with adjuvant therapies. We present the case of a 60-year-old woman who was referred to the Emergency Department with acute onset of abdominal distension, nausea, vomiting, right back pain, lipothymia and a giant abdominal mass.

Keywords: retroperitoneal sarcoma, liposarcoma, surgery.

INTRODUCTION

Retroperitoneal liposarcoma (RPLS) is a rare form of cancer that mostly affects adults. Symptoms may be undetected unless the tumor begins to grow. Diagnosis is made when the tumor is large and puts vital organs such as the pancreas, liver, kidneys at risk (1). Symptoms of a liposarcoma depend on the location of the tumor (2). Liposarcoma in the arms, legs, particularly thighs may be painful, swollen, with patients citing weakness in the affected area (3); 15% of soft tissue sarcomas are retroperitoneal, with two of the most common types being the well-differentiated liposarcoma (WDLPS) and high-grade dedifferentiated liposarcoma (DDLPS). The tumor can weigh be-

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tween 15 kg and 20 kg, with 20 to 25 cm in diameter (4). Surgery is the main form of treatment provided along with adjuvant therapies.

Well-differentiated liposarcomas are characterized as slow-growing tumors that may recur but have minimal capacity to metastasize. However, dedifferentiation and trans-differentiation may lead to the progression of WDLPS into high-grade tumors. On the other hand, DDLS may be either newly formed or progress from antecedent WDLPS and are characterized by high maturation and spread rate (5). Myxoid liposarcoma (MLPS) are rare in the retroperitoneum (6) and often spread to unusual areas such as bones and skin (5). Pleomorphic liposarcoma (PLPS) occurs mostly among the elderly and is characterized by aggressive tumors with high metastatic capacity. Diagnosis of patients is conducted through presurgical imaging such as MRI, ultrasonography, or CT scan. These imaging examinations also determine the surgical techniques to be applied by surgeons, either open or laparoscopic surgery for tumors less than 10 cm in size (7). The infrequency of retroperitoneal liposarcoma, anatomical location, and varying clinical genetic subtypes makes it difficult to deduce distinct retroperitoneal conclusions (6). However, despite its rarity, many surgeons, including general surgeons, can diagnose and provide treatment (2). We present the case of a 60-year-old woman who was referred to the Emergency Department with acute onset of abdominal distension and a giant non-tender mass. ❑

CASE REPORT

We present the case of a 60-year-old female patient who was referred to the Emergency Department with acute onset of, nausea, vomiting, abdominal distension. She had a medical history of essential hypertension and no previous surgery. On physical examination she had a good performance status, the arterial blood pressure was 120/70 mm Hg, pulse rate 80 b/min, and oxygen saturation 98%. Blood exams showed a lymphocyte number of $10x10^3/mm^3$ and hemoglobin 6 g/dL. Physical examination of the abdomen revealed a giant non-tender mass in the left and midline quadrants. The patient was transfused with three units of blood. Abdomen CT scan revealed a retroperitoneal mass of 30 cm in contact with surrounding viscera (stomach, spleen, vena cava) (Figure 1). The patient underwent a CT scan plus fine-needle-aspiration (FNA) which confirmed a WDLPS. The case was

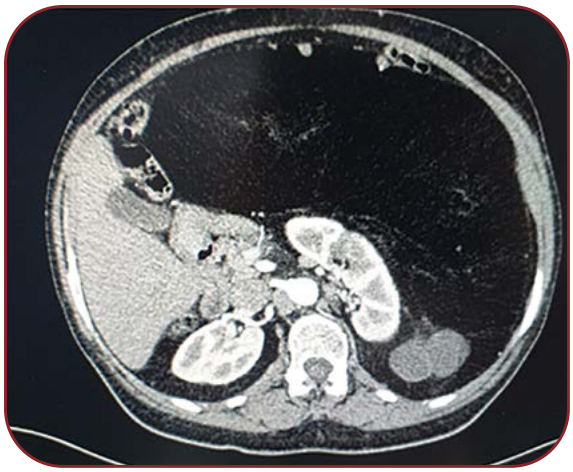

FIGURE 1. Abdomen CT scan revealed a retroperitoneal mass of 30 cm in contact with surrounding viscera (stomach, spleen, vena cava)

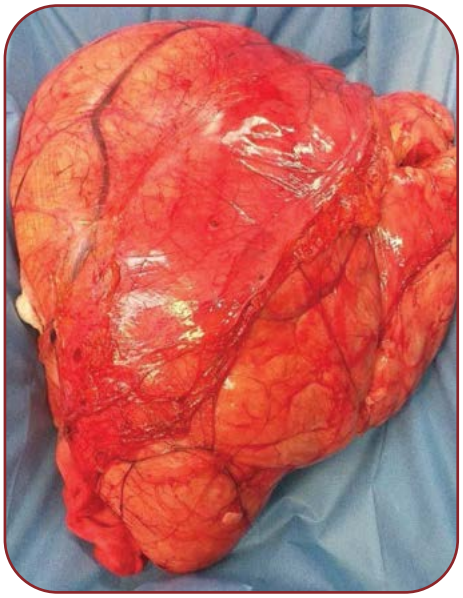

FIGURE 2. Total excision of retroperitoneal mass

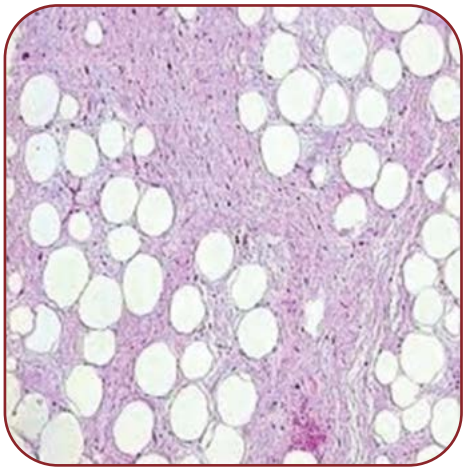

FIGURE 3. Hystopathological finding: well differentiated liposarcoma. French Federation of Cancer Centers Sarcoma Group: Grade 1 tumor differentiation:1; mitotic count:1; tumor necrosis:0

referred to a multidisciplinary team (MDT) composed of surgeons, pathologist, interventional radiologist and oncologist. The decision was to not carry out the neoadjuvant given but to perform up-front surgery. A laparotomy was performed. Xipho-pubic incision. Once the peritoneum is opened, a voluminous retroperitoneal mass of about 30 cm was highlighted extending from the left hypochondrium to the left iliac fossa and dislocate the left colon and small intestine medially; latero-medial colic along Toldt's fascia, take down of the splenic flexure and visualization of the entire mass. Mass dissection en bloc preserving left kidney, spleen, colon and ureter, with total excision of retroperitoneal mass without the need to remove other organs. Closure of the laparotomy (Figure 2). On the seventh day, the patient was in good clinical condition and she was discharged. Hystopathological finding: well-differentiated liposarcoma. French Federation of Cancer Centers Sarcoma Group: Grade 1 Tumor differentiation:1; Mitotic count:1; Tumor necrosis:0 (Figure 3). ❑

DISCUSSION

Analysis of patient records diagnosed with RPLS shows different histological subtypes of the tumors which vary in terms of size, growth, and metastatic potentials. Patients would complain of lower abdominal pain. As per the reviewed literature, patients' age ranged from the late forties to mid-fifties, with equal gender distribution. The retroperitoneum has no bony boundaries; therefore, no immediate symptoms were highlighted (8). For non-metastatic RPLS, surgery is the mainstay form of treatment (4). The peculiarities of the case highlight that surgery aims at complete resection as this is the only chance for a cure. Ensuring a negative microscopic margin (R0) is ideal as it reduces the risk of recurrence; however, it may be difficult to achieve it due to the size of the RPLS. Therefore, complete resection defines optimal macroscopic margins at negative (R0) or positive (R1) microscopic margins (9).

Patients with recurrent RPLS may still be considered for surgery, although case discussion is critical. Non-surgical therapies such as radiation therapy are offered as well. Studies show that radiotherapy along with surgery improved the survival rates compared to just surgery alone. Patients would receive pre- (62% survival) and post-operative (60% survival) radiation therapy compared to 54% and 52% survival rate with surgery only. However, patients with RPLS would prefer pre-operative radiation as it limits the damage caused to adjacent organs. Chances of recurrence when using non-surgical treatment remain uncertain (9). On the other hand, chemotherapy evaluates the response to therapy, providing a surgical advantage when the tumor size reduces. However, patients' response to chemotherapy differs depending on the RPLS subtype and anatomical location.

According to the available literature, WDLP and DDLPS recorded poor responses to systemic therapy (8). Adjuvant therapies are provided to patients as per the recommendations of surgeons and oncologists. Each cycle of radiation therapy and chemotherapy requires a clinical follow-up to monitor recurrence. Patients are expected to adhere to after-treatment guidelines such as lifestyle changes, no tobacco use or drinking alcohol, and weight control, and ensure that they attend the screening as per instruction. Non-surgical treatments are considered controversial by researchers due to the negative effects they manifest in patients (3); hence, surgery is considered to be the most effective treatment for liposarcoma. Radiation treatment is provided for patients with R1 and R2 microscopic margins. Adjuvant therapies enable surgeons to monitor patients' response and reduce tumor size, providing a surgical advantage. This, in turn, enables surgeons to achieve a negative microscopic margin of R0. Surgery often results in the complete removal of the tumor and adjacent structures (1). For patients with RPLS, tumor invasion leads to organ dislocation, which can contribute to uncontrollable bowel movements. However, since the viscera may fall into the resection bed, postoperative therapies are essential (5). Though clinical follow-up is recommended for non-surgical patients, it may be uncertain as patients tend to be asymptomatic with unspecified symptoms. This also leads to misinterpretation of local recurrence (5). Liposarcomas are the most common type of sarcomas, with 12-40% of cases arising in the retroperitoneum. However, they occur where there is fat. They are associated with genetic anomalies with different behavioral scales. Progression and development range from well-differentiated tumors that do not spread to

more aggressive dedifferentiated tumors. It is observed that differentiation most likely occurs in the retroperitoneum, although researchers have yet to ascertain the reasons behind this progression (10). Survival and recurrence are associated with complete resection with negative microscopic margins. Surgery remains the optimal treatment since surgeons providing non-surgical treatments gain experience by conducting them on patients. This may result in potentially substandard outcomes. Therefore, it is critical for surgeons and oncologists to have multidisciplinary case discussions to determine the most effective therapeutic approach for their patients. Patients and healthcare givers ensure that there is prior engagement before commencing adjuvant therapies. This was witnessed with the presence of written consent from the patients after discussions with the multidisciplinary board. Researchers further suggest trials of novel systemic therapies based on the biological understanding of a given histological subtype. This means that the efficiency in treatment would heavily rely on the subtype (8). ❑

CONCLUSIONS

The available literature shows that RPLS can be cured by integrating surgical and non-surgical treatments. However, data on recurrence and survival remains unclear when the two therapeutic approaches are applied together. Research should focus more on highlighting the statistics in this regard to enable sarcoma centers to implement and promote these treatments. Management of RPLS is challenging, but institutional collaboration would be vital in developing strategies that promote the best clinical practice to understand RPLS. Finally, sarcoma centers should work alongside researchers on mechanisms that would enable them to detect RPLS before tumor growth and escalation of the disease. Since treatment can only commence once the tumors are sizeable for surgery, perhaps a shift in focus in this regard would help lower the chances of local recurrence and increase survival rates. ❑

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