ISSN (P) : 2589-9120 / (O) : 2589-9139
PubMed-National Library of Medicine - ID: 101773527

International Journal of Medical Science and Applied Research (IJMSAR)

Available Online at: https://www.ijmsar.com

Volume – 6, Issue – 2, March – 2023, Page No. : 16 – 21

Giant Dedifferentiated Retroperitoneal Liposarcoma a Multidisciplinary Approach in the Diagnosis and Management in a Patient with a History of Multiple Tumors: A Case Report

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Citation of this Article: Niño S, Castañeda I, Principe J, Jose A, Pasha A, "Giant Dedifferentiated Retroperitoneal Liposarcoma a Multidisciplinary Approach in the Diagnosis and Management in a Patient with a History of Multiple Tumors: A Case Report,'' IJMSAR – March – 2023, Vol. – 6, Issue - 2, Page No. 16-21.

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Type of Publication: A Case Report

Conflicts of Interest: Nil

ABSTRACT

Background

Liposarcomas are defined as malignant adipocytic differentiation tumors. They are mainly divided into three morphological subgroups, with a wide diversity of biological behaviors, ranging from well-differentiated tumors with low metastatic potential to pleomorphic subtypes that tend to be high-grade and with a high rate of distant metastasis, like dedifferentiated subtype. The prognosis of retroperitoneal liposarcomas is poor compared to other retroperitoneal sarcomas, with complete surgical resection being the gold standard in management and the most important prognostic factor. However, in well-differentiated histological subtypes where the margins are not well defined, it may require resection of contiguous organs and combined management with radiotherapy and chemotherapy as an adjuvant or neoadjuvant treatment to prevent future recurrences.

Case Presentation

In this report, we present the surgical excision of a retroperitoneal liposarcoma without the involvement of contiguous structures and its possible genetic or syndromic relationship in a patient with a history of thymoma and pulmonary hamartoma.

Conclusion

Retroperitoneal liposarcomas should be entities approached by a multidisciplinary group, in order to carry out an early diagnosis and a safe treatment.

Keywords

Dedifferentiated liposarcoma, multidisciplinary approach, retroperitoneal sarcomas, surgical treatment.

INTRODUCTION

Retroperitoneal sarcomas (RPS) are a rare heterogeneous group of mesenchymal neoplasms which is divided into liposarcomas, leiomyosarcomas, solitary fibrous tumors, malignant peripheral nerve sheath tumors and undifferentiated pleomorphic sarcoma. Liposarcomas (LPSs) are malignant tumors of adipocytic differentiation . They are among the most common soft tissue sarcoma (STS) subtypes, accounting for approximately 15% to 20% of all STSs [1]. Dedifferentiated liposarcoma is one of the histologic subtypes of liposarcoma, and it is characterized by a high-grade and aggressive disease. They are rarely in the retroperitoneum as a primary location and are known for local and metastatic recurrence [2]. Most of the cases are associated with the amplification of the chromosome segment MDM2, CDK4, and HMGA2. MDM2 is responsible for encoding a protein that induces the degradation of p53 (a tumor suppressor gene), while CDK4 encodes an oncoprotein that promotes G1/S progression of the cell cycle [3].

The most common clinical presentation is that of a gradually enlarging, painless mass. Less commonly, they suffer from pressure symptoms like bowel obstruction from being compressed by the mass. Chest, abdomen, and pelvis computed tomography (CT) scan with IV contrast are the images of choice for characterizing the lesion and locating possible metastases [2].

A routinely recommended diagnostic procedure is a percutaneous core needle biopsy, which helps grade the tumor and consequently guides the treatment plan. Nowadays, there are three types of treatments for RPS. Although the cornerstone is surgery, some patients are candidates for radiotherapy or chemotherapy, depending on the grade and type of RPS [2]. The aim of the surgery is to perform a complete macroscopic resection [4]. Preoperative radiotherapy is preferred to postoperative radiotherapy, and can be beneficial for low to intermediate grade RPS. On the other hand, the response of dedifferentiated liposarcomas to chemotherapy is about 21% [2].

CASE PRESENTATION

We present the case of a 70-year-old Cuban man referred to our surgery outpatient clinic for evaluation of an enlarging abdominal mass found during a primary care visit. He complained of abdominal discomfort, constipation, and shortness of breath on moderate exertion. On physical examination, the patient was well and in no apparent distress, with a visibly distended abdomen in correlation with a centrally situated variegated, non-tender mass of 20 x 17 cm occupying the bilateral iliac, lumbar, umbilical, and hypogastric regions of the abdomen. Bowel sounds were present, and there were no other significant physical findings.

The patient is a former smoker with a past medical history of hypertension, obesity class III (BMI 35.7 kg/m2), dyslipidemia, aortic atherosclerosis, cardiac arrhythmia, and emphysema. Surgical history is remarkable for a left robotic assisted thoracoscopy for local resection of lower left hamartoma, mediastinal

resection of a thymoma, and a colonic polypectomy. There is no known history of allergies, and the family history is unremarkable.

Lab investigations revealed elevated white blood count (12,900/mL), platelets (583,000/mL), glucose (146 mg/dL), potassium(5.6 mEq/L), alkaline phosphatase (170 U/L), and ALT (61 U/L), and decreased hemoglobin (11 g/dL). A CT abdominal/pelvic scan without contrast demonstrated interval development of a large heterogeneous, possibly multicystic mass in the lower abdominal cavity measuring 20 x 17 x 18 cm of unclear origin situated retroperitoneally (Figures 1A, 1B).

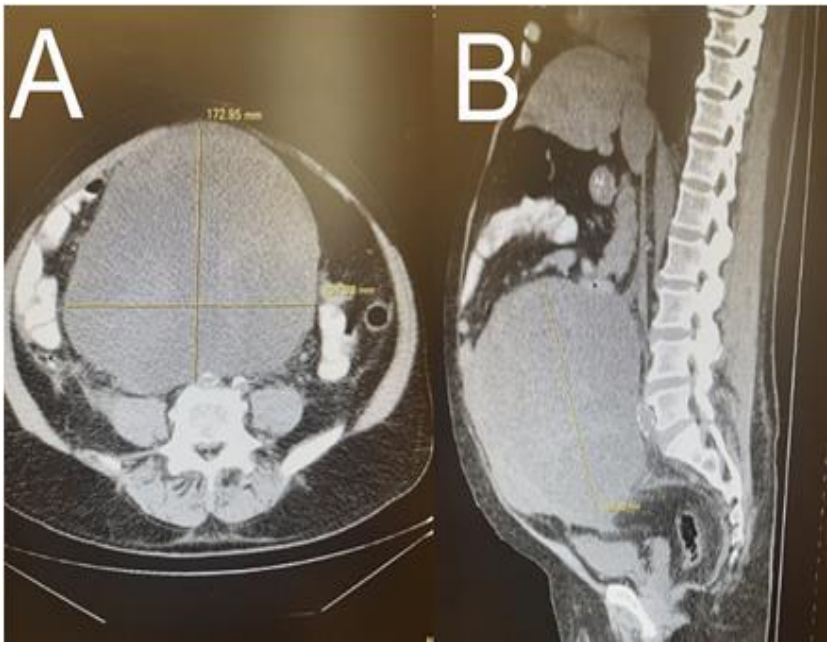

Figure 1: Preoperative CT scan of the abdomen and pelvis. (A) Axial plane of the retroperitoneal mass with measurements of 172.95mm x 197.28mm (yellow lines) and (B) sagittal plane with a length of 164.46mm (yellow line).

The initial surgical plan was to carry out a diagnostic laparoscopy, but it was converted into an open resection of the abdominal mass and inspection of the peritoneum. Upon opening the abdominal cavity, the mass was incidentally open with leaking of clear fluid, blood, and a gelatinous substance. The mass was in the middle and lower abdominal cavity; it was adherent to the intestinal mesentery. All internal organs were displaced upward, and free fluid was found. The intraoperative planes were obscured by the high vascularity of the mass, and bleeding control was performed with posterior excision of the mass and pedicle, with a total weight of 4596 grams (Figure *2*). A JP drain was sutured in place. Post-operatively the patient was admitted into the intensive care unit for further management. Pathology of surgical report demonstrated a retroperitoneal dedifferentiated liposarcoma grade II with margins involved by sarcoma, with immunostains positive for MDM2.

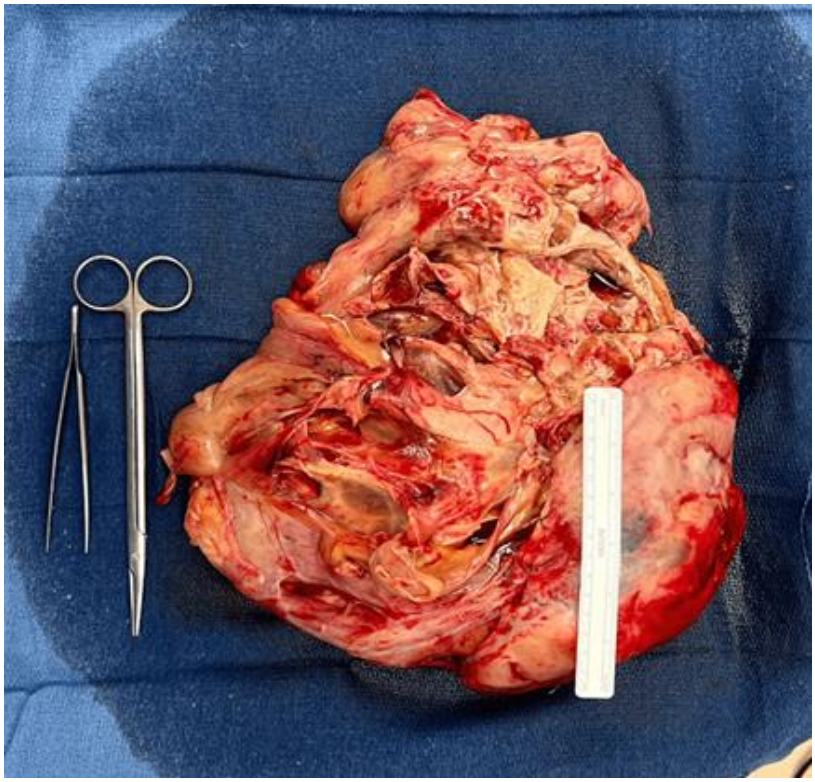

Figure 2: Macroscopic view of the tumor. Gross view of the liposarcoma: 30×28×13 cm in size.

DISCUSSION

Soft tissue sarcomas are an extremely rare entity with a prevalence of less than 1%. Within the multiple histological subtypes, liposarcomas are the most common tumor in 15 to 20% of cases, the most frequent location being the extremities in 48%, followed by a retroperitoneal location in 13% of cases [1-2] .

Retroperitoneal liposarcomas are mesenchymal tumors with a slow and often asymptomatic clinical evolution due to the characteristics of the abdominal and retroperitoneal cavities, which provide them with adequate conditions for their development [3-4]. These tumors, in most cases, are incidental findings since the patients who present any symptoms are at a stage that usually suggests that the mass has reached a large enough size to compress adjacent structures, mostly vascular and possibly infiltrated adjacent organs, a case similar to what occurred with the patient presented in this case report [5].

Dedifferentiated liposarcoma is one of the histologic subtypes of liposarcoma. It is characterized by a high-grade and aggressive disease that can be located within the retroperitoneum and is associated with high rates of local and metastatic recurrence and disease-specific mortality. Recent research in molecular genetics has found the genes implicated in liposarcomas. Among these is the murine double minute 2 (MDM2) and CDK4 genes [6].

Currently, the gold standard for the preoperative evaluation of liposarcomas is the CT scan of the abdomen since it provides us with the dimensions of the mass and additionally gives us the tools to plan the surgical approach. Today, surgery is the only definitive management, in which a radical and

complete resection is the best way to reduce recurrence rates [5] since the main factors that affect the prognosis of the disease continue to be the tumor grade and degree of surgical resection. However, complete resections on multiple occasions are often strenuous due to the difficulty in identifying the margins and the involvement of adjacent organs such as ureters, kidneys, great vessels, and intestinal vasculature. For this reason, when the margins are difficult to demarcate or in the presence of neighboring structures being compromised, the removal of the involved structure is recommended as it can result in recurrences if left behind.

However, today new treatments have appeared together with surgery, this includes neoadjuvant radiotherapy or radiochemotherapy. According to the STRASS study, preoperative radiation and surgery did not increase patients' chances of living without recurrence compared to surgery alone but instead made their adverse effects worse. [7]. On the other hand, in a retrospective case series of patients with dedifferentiated liposarcomas in which radiochemotherapy and surgery were used together, it was shown that out of 10 patients, tumor recurrence was observed in 3 of 4 patients (75%) without neoadjuvant treatment, and 2 of 6 patients (33%), with neoadjuvant radiochemotherapy in a 2-year follow-up [8-9]. Therefore, studies and results are insufficient to define the effectiveness and efficacy of neoadjuvant therapy and surgery [9].

CONCLUSION

We present a case of dedifferentiated retroperitoneal liposarcoma. Taking into account that this patient has a past history of other tumors (hamartoma, thymoma, and a colonic polyp), we consider that this patient may be genetically predisposed to develop tumors, so we highly recommend he should go under genetic testing. However, we don't rule out the possibility that this tumor has appeared de novo. This demonstrates the importance of an extended multidisciplinary group, in order to provide an early and adequate diagnosis, surgical planning through diagnostic images, and the concomitant help of pathology for both preoperative and postoperative treatment, as well as other specialties such as genetics, in specific cases.

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