CASE REPORT

Mixed-type Retroperitoneal Liposarcoma—A Combination of the Well-differentiated and Myxoid Types: Case Report and Literature Review

Orlando Favio Arevalo Guerra[1], Alden Pool Gomez[2], Cristian Daniel Arevalo Guerra[3], Adriana Marcela Zuñiga[4]

Received on: 25 February 2023; *Accepted on:* 11 May 2023; *Published on:* 20 July 2023

Abstract

Introduction: Soft tissue sarcomas represent less than 1.5% of malignant tumors. They are characterized by destructive growth, recurrence, and distant metastases to the lungs. The sites more frequently affected are the extremities in 40% and the retroperitoneum in 15–20% of the cases.

Case description: A 57-year-old female presented reporting a history of pain for 12 months and a progressively growing abdominal mass. The initial histological analysis indicated a diagnosis of lipoma in the mesentery, but a nuclear magnetic resonance of the abdomen showed two retroperitoneal masses. After laparoscopic resection, immunohistochemistry confirmed a well-differentiated liposarcoma and myxoid liposarcoma. One year later, the patient had a second surgical intervention due to tumor relapse, with pathology showing a well-differentiated liposarcoma.

Discussion: Mixed-type liposarcoma is the most uncommon histotype. It is still an unknown entity. It shows the combined features of myxoid/round and liposarcoma well-differentiated/dedifferentiated. Also, it can show the characteristics of the myxoid subtype/round or pleomorphic liposarcoma. All these different presentations generate conceptual conflicts when establishing the protocols for management and treatment.

Conclusion: Mixed-type liposarcoma represents the most uncommon subtype of liposarcoma. Complete surgical resection with negative margins remains the standard of treatment. In order to achieve negative margins, en bloc multivisceral resection and neoadjuvant therapies (chemotherapy or radiotherapy) are often necessary in selected cases.

Keywords: Case report, Leiomyosarcoma, Myxoid liposarcoma, Retroperitoneal liposarcoma, Surgery, Well-differentiated liposarcoma.

AMEI's Current Trends in Diagnosis & Treatment (2023): 10.5005/jp-journals-10055-0160

Introduction

Soft tissue sarcomas are a heterogeneous group of rare malignant tumors, with an annual incidence of approximately 5 per 100,000 individuals, representing less than 1.5% of all human malignancies.[1] They are characterized by destructive growth, recurrence, and distant metastases to the lungs. The sites more frequently affected are the extremities in 40% and the retroperitoneum in 15–20% of the cases.[2]

The most common histotypes that tend to appear in the retroperitoneum are represented by liposarcomas (LS), and leiomyosarcomas in 75% of cases.[3]

Liposarcomas are rare malignant adipocytic tumors that constitute approximately 20% of all soft tissue sarcomas and 60–70% of retroperitoneal sarcomas. The most frequent histotypes of retroperitoneal sarcoma in their order of frequency are well-differentiated liposarcoma, dedifferentiated liposarcoma, then myxoid, round cell, pleomorphic, and mixed-type.[4]

Mixed-type LS are an uncertain entity. The term refers to the combination of morphological subtypes of a liposarcoma, most often a mixture of well-differentiated LS with myxoid, dedifferentiated, myxoid/round cell, or pleomorphic liposarcoma.[4]

The heterogeneity of histological subtypes and the absence in the literature with specific guidelines for the management of this type of neoplasms encouraged us to carry out this review of the literature. This case report was written in accordance with the Surgical Case Report (SCARE) criteria.[5]

[1,2,4]Cirugía General, Hospital Universitario del Valle "Evaristo Garcia", Cali, Valle del Cauca, Colombia

[3]Department of Medicina, Cali, Valle del Cauca, Colombia

Corresponding Author: Orlando Favio Arevalo Guerra, Cirugía General, Hospital Universitario del Valle "Evaristo Garcia", Cali, Valle del Cauca, Colombia, Phone: +57 3123754728, e-mail: orlan916@gmail.com

How to cite this article: Guerra OFA, Gomez AP, Guerra CDA, *et al*. Mixed-type Retroperitoneal Liposarcoma—A Combination of the Well-differentiated and Myxoid Types: Case Report and Literature Review. AMEI's Curr Trends Diagn Treat 2023;7(1):17–20.

Source of support: Nil

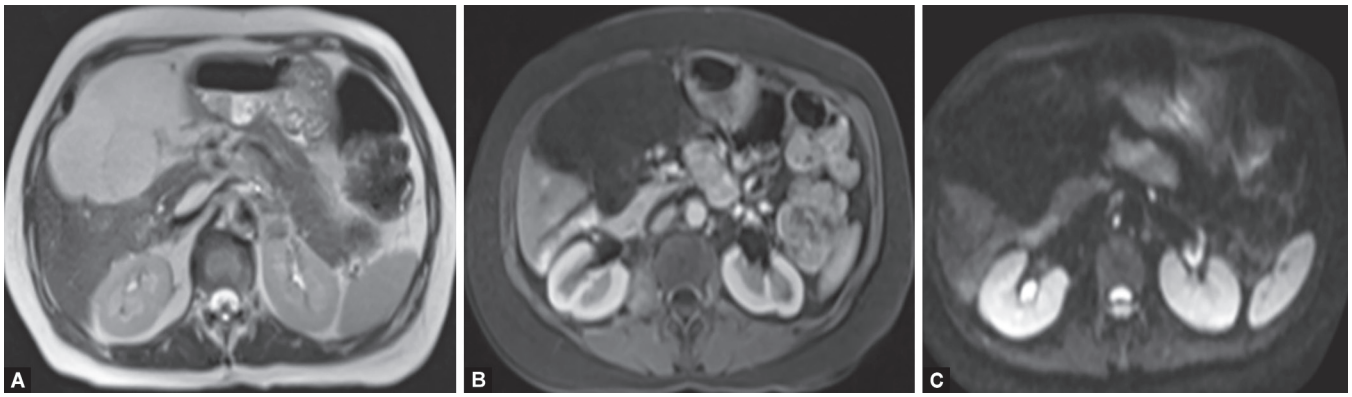
Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

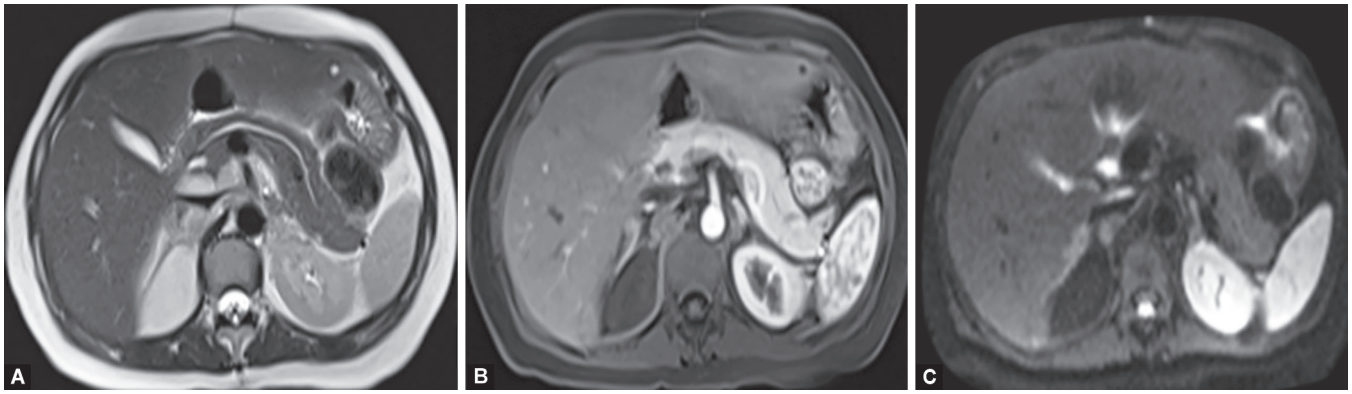
Case Description

A 57-year-old female patient presented reporting a mass in the lower left quadrant of the abdomen, with progressive growth, that appeared a year ago. The mass was associated with constipation, abdominal distension, non-radiated oppressive type pain, concomitant heartburn, and occasional emesis.

Relevant medical history includes non-insulin-dependent diabetes mellitus, well-controlled for 18 years, and right



Figs 1A to C: (A) Preoperative nuclear magnetic resonance imaging (MRI) shows a mass in the subhepatic location with homogeneous signal intensity and similar to adipose tissue in the T2-weighted image; (B) In the acquisition of T1 with fat saturation after the administration of contrast medium, no areas of enhancement were observed; and in (C) Diffusion-weighted imaging (DWI) images, there is no restriction



Figs 2A to C: Mass with similar characteristics in the right perirenal space

hemicolectomy due to tumor resection in the distal ileum and right colon. The data were found in the medical history from 2006, and no results for histology analysis were reported.

An initial abdominal computerized tomography (CT) was performed, showing a 7 × 6 × 6 cm lesion with a lipomatous appearance in the mesentery, adjacent to the gallbladder fossa and the right hepatic rim. Also, an upper gastrointestinal endoscopy was completed, reporting a type I hiatal hernia (3 cm) and erythematous gastropathy. The total colonoscopy showed normal results.

The abdominal lesion was taken to a Tru-Cut biopsy and histological analysis, indicating that findings were consistent with lipoma.

Due to the persistence of intermittent symptoms, a simple and contrasted nuclear magnetic resonance (NMR) of the abdomen was performed, showing a 12 × 7 × 10 cm anterior subhepatic soft tissue mass in the right lobe and a 3 × 1 × 2 cm kidney lesion in the lower left pole (Fig. 1). After these results, the patient was taken to laparoscopic surgery where the lesion and lower pole of the left kidney were resected. Microscopic examination showed two patterns: the first part showed cells similar to mature fat cells with occasional multivacuolated lip blasts in the well-differentiated liposarcoma with positive immunohistochemistry for S-100, the second part showed myxoid matrix, plexiform vascular pattern, undifferentiated cells, and atypical lip blasts, with adipocyte maturation in the periphery of territory with the characteristics of myxoid liposarcoma, negative for S-100. The definitive diagnosis revealed well-differentiated liposarcoma and myxoid liposarcoma, with non-evaluable surgical margins. (Fig. 1).

She received adjuvant management with ifosfamide and doxorubicin for 12 months. One year after surgery, an abdominal MRI was performed with two lesions in the right posterior perirenal space and a solid nodular lesion about the ipsilateral adrenal gland, suggestive of a recurrence tumor (Fig. 2).

Fifteen months after the index surgery, she was taken again for resection of a retroperitoneal mass where a right nephrectomy was performed, with well-differentiated liposarcoma histology study. The tumor has not presented recurrence during 2 years of follow-up to the current date. The renal function before the first surgery presented creatinine of 0.8 mg/dL; after radical nephrotomy on the right and partial left, it presented creatinine of 1.1 mg/dL without requiring dialysis support.

The reported case represents a rare example of mixed-type liposarcoma arising in the soft tissue of the retroperitoneum in an adult patient, in which both tumor components are present in a multifocal manner, highlighting the exotic nature of this presentation.

Discussion

Sarcomas are soft tissue tumors derived from mesenchymal tissue. They are the most uncommon solid tumors (<1% in patients older

than 50 years). There are more than 50 histological types, and the most frequent locations include 40% extremities, 22% viscera, 16% retroperitoneum, 10% thorax, and 12% other sites.[3]

Primary retroperitoneal tumors represent 0.2% of all malignant neoplasms. Within these, liposarcoma constitutes approximately 20% of all soft tissue sarcomas and 60–70% of retroperitoneal sarcomas in adults, being the most common sarcoma from the retroperitoneum.[6] These originate in the adipocyte and are divided into morphological subtypes, such as well-differentiated, dedifferentiated, myxoid, round cell, pleomorphic, and mixed-type liposarcoma.[4,6]

Mixed-type LS are rare, accounting for 13.8% of all retroperitoneal LS,[6] and are characterized by the combined properties of myxoid/round cell and well-differentiated/dedifferentiated liposarcoma or features of the myxoid/round cell/pleomorphic subtype. The several different presentations mentioned above generate conceptual problems in management protocols.

The morphological heterogeneity of mixed-type LS has been explained by different levels of maturation within a single developmental pathway of adipogenesis.[1]

The diagnosis commonly arises as an incidental finding on images performed on asymptomatic patients with an abdominal mass. The average size at the time of diagnosis is 15 cm is found, of which 10% have metastases.[7]

The clinical features often appear with vague and indolent symptoms that present a diagnostic challenge. The compression or invasion of neighboring structures could guide us. As a result of this, we can see edema in the extremities, neurological symptoms in the lower limbs, early satiety and intestinal obstruction when the gastrointestinal tract is compromised, ascites due to compression of the portal vein, catarrh-like symptoms in high-grade sarcomas, and rapid growth, and paraneoplastic hypoglycemia in leiomyosarcoma producing Insulin-like growth factor 2 (IGF-2).[2]

Regarding the differential diagnosis, it should be remembered that 2/3 of the neoplasms of the retroperitoneum are not sarcomas, and each case must be individualized, that is, in a patient with a retroperitoneal mass evidenced by images that are accompanied by fever, diaphoresis, weight loss, adenopathy, one should be thinking on lymphomas as the first diagnosis to rule out.

When finding a retroperitoneal mass with mass in the testicles, the first diagnosis to be ruled out is a germ cell tumor. Also, when there is evidence of a retroperitoneal mass with hematuria, one should consider kidney cancer.[2,8]

The The National Institute for Health and Care Excellence (NICE), National Comprehensive Cancer Network (NCCN), and UK guidelines indicate intravenous contrast-enhanced tomography of the thorax, abdomen, and pelvis as the standard for staging.[9–11] The European Society for Medical Oncology (ESMO) guidelines recommend the NMR be used especially for pelvic tumors to assess specific aspects of tumor extent, in allergies to intravenous contrast, as well as in cases where the evaluation of spinal foramina, posterior vertebrae, nerves, and/or muscles are necessary.[12]

Although it was previously believed that LS biopsy could lead to inadvertent tumor tract seeding, the literature shows that the incidence of tumor tract seeding is negligible (0.4% risk).[2] Thus, it is advised to perform a biopsy before treatment for pathological diagnosis, except in the case of well-differentiated liposarcoma, in which the histological diagnosis is achieved by NMR, and pre-resection biopsy is not necessary.[10]

Due to the high local recurrence rather than distant metastasis, complete surgical resection with negative margins remains the standard of treatment for non-metastatic LS and is the only possibility of cure.[13] To achieve negative margins, en bloc multivisceral resection is often necessary. The use of neoadjuvant therapies (chemotherapy, radiotherapy) is indicated in selected cases of initial unresectability.[14–16]

In general, the 5-year survival rate in myxoid and well-differentiated liposarcoma exceeds 70%.[1]

Conclusion

The mixed-type liposarcoma represents the rarest subtype of liposarcoma, with only a few cases reported in the literature. In this article, a rare case of mixed-type retroperitoneal liposarcoma has been reported, highlighting the broad spectrum of lipogenic malignancies and the need for more extensive and detailed studies of large mesenchymal neoplasms.

Consent

Written informed consent was obtained from the patient and his family members for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Orcid

Orlando Favio Arevalo Guerra https://orcid.org/0000-0003-3615-7336
Alden Pool Gomez https://orcid.org/0000-0002-1830-4577

References

1. Fletcher CDM. The evolving classification of soft tissue tumors – an update based on the new 2013 WHO classification. Vol. 64, Histopathology. John Wiley & Sons, Ltd; 2014. p. 2–11. DOI: https://doi.org/10.1111/his.12267.
2. Quagliuolo V, Gronchi A, editors. Current treatment of retroperitoneal sarcomas. Milano: Springer Milan; 2019.
3. Brennan MF, Antonescu CR, Alektiar KM, et al. General Description. In: Management of Soft Tissue Sarcoma. Springer International Publishing; 2016. p. 3–17. DOI: https://doi.org/10.1007/978-3-319-41906-0_1.
4. Kindblom LG. Lipomatous tumors – How we have reached our present views, what controversies remain and why we still face diagnostic problems: A tribute to Dr. Franz Enzinger. Adv Anat Pathol 2006;13(6):279–285. DOI: 10.1097/01.pap.0000213053.00060.5.
5. Agha RA, Franchi T, Sohrabi C, et al. The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines Int J Surg 2020;84:226–230. DOI: 10.1016/j.ijsu.2020.10.034.
6. Ohyabu Y, Sameshima H, Nakayama M, et al. The retroperitoneal liposarcoma: a case report. Hinyokika Kiyo 1989;35(2):307–313. Available from: http://hdl.handle.net/2433/116430.
7. Dumitra S, Gronchi A. The Diagnosis and Management of Retroperitoneal Sarcoma. Oncology (Williston Park). 2018;32(9):464–469.
8. Mota MMDS, Bezerra ROF, Garcia MRT. Practical approach to primary retroperitoneal masses in adults. Rad Bras 2018;51(6):391–400. DOI: 10.1590/0100-3984.2017.0179.
9. Sarcoma NICE quality standard 78, 2015. Available from: https://www.nice.org.uk/guidance/qs78.
10. Benjamin RS, Bui MM, Choy E, et al. NCCN Guidelines Version 2.2021 Soft Tissue Sarcoma.
11. Dangoor A, Seddon B, Gerrand C, et al. UK guidelines for the management of soft tissue sarcomas. Clin Sarcoma Res 2016;6:20. DOI: 10.1186/s13569-016-0060-4.
12. Casali PG, Abecassis N, Bauer S, et al. Soft tissue and visceral sarcomas: ESMO–EURACAN Clinical Practice Guidelines for diagnosis,

treatment, and follow-up. ESMO Guidel Comm 2018;29(4):iv51–iv67. DOI: 10.1093/annonc/mdy096.
13. de Vreeze RSA, de Jong D, Koops W, et al. Oncogenesis and classification of mixed-type liposarcoma: A radiological, histopathological and molecular biological analysis. Int J Cancer 2011;128(4):778–786. DOI: 10.1002/ijc.25390.
14. Brennan MF, Antonescu CR, Maki RG, et al. Natural history: importance of size, site, and histopathology. In: management of soft tissue sarcoma. New York: Springer; 2013. p. 19–35.
15. Mullinax JE, Zager JS, Gonzalez RJ. Current diagnosis and management of retroperitoneal sarcoma. Cancer Contro 2011;18(3):177–187. DOI: 10.1177/107327481101800305.
16. Dumitra S, Gronchi A. The Diagnosis and Management of Retroperitoneal Sarcoma. Oncology (Williston Park). 2018;32(9): 464–469. PMID: 30248168.