Leiomyosarcoma of inferior vena cava wall in an old woman

Leiomiosarcoma de la pared de la vena cava inferior en una paciente anciana

doi: 10.61997/bjm.v14i1.441

Vitorino Modesto dos Santos[1], Taciana Arruda Modesto Sugai[2], Lister Arruda Modesto dos Santos[3]

Dear Editor

Leiomyosarcoma (LMS) is a malignant tumor first described in 1871 by Leopold Perl and Rudolph Virchow, more often affecting women in their fifth and sixth decades of life and a median age of 59 years, and rarely developing in the inferior vena cava (IVC) wall.[1-6] The authors of a literature review in 2024 estimated that vascular LMS represents 5% of soft tissue ones, and 50% have origin in the IVC or veins of lower limbs, while the incidence in autopsies is 1/7000-34,000, and less than 400 cases have been described.[3] Recently, we read Diaz Riverol's excellent didactic Radiographic Quiz focusing on the main characteristics of LMS developed in the IVC wall of a 78-year-old female patient.[2] The retroperitoneal tumor presented necrotic areas and caused compression of the bile and pancreatic ducts, besides hydronephrosis, that were corrected by surgical procedures.[2] The author emphasized that only 5% of LMSs are exclusively intravascular, and may have metastases at diagnosis in 23% of patients, mainly to the lungs, liver, and peritoneum.[2] Aiming to highlight this very illustrative case study published in this Journal,[2] some additional comments on literature data from 2024 about LMS of IVC are presented.[1,3-6]

A 68-year-old man with a diagnosis of LMS of the IVC associated with a tumor thrombus affecting the right atrium, underwent surgical resection of the tumor in addition to the infrarenal IVC, right kidney, and adrenal, after the neoadjuvant chemoradiation (combined 28 cycles of radiation and dose-escalated of Pazopanib up to 600 mg/ day).[1] The LMS measured 16.1×14.6×20.7 cm and the pattern was intermediate grade 2/3. The post-operative course was marked by chylous ascites managed by Denver shunt; metastasis to C7 treated by radiotherapy and trabectedin; and metastasis to the lungs and liver controlled with Eribulin and Lenvatinib; he is followed by the oncology specialists. The authors stressed the management strategy for LMS at first considered inoperable.[1] The case series including five women with a median age of 53 (51-74) years at diagnosis of LMS in the IVC was described, and either their presentation symptoms were nonspecific or the malignancy was incidentally detected during a routine examination.[4] All the patients underwent surgical resections (three had involvement of suprarenal IVC, and two had LMS involving the infrarenal segment); cholecystectomy and partial liver resection were needed in one case because of metastases present in the left liver lobe.[4] The LMS growth was intraluminal in three cases and mixed in two, the median size was 5 cm, four patients had intermediate (grade 2) or high-grade (grade 3) tumors, the patient who had liver metastases died 19 months after surgery, the other evolve asymptomatic.[4]

A 67-year-old woman diagnosed with LMS of the IVC presented a lesion extending up to the right atrium, besides multiple hepatic, osseous, and lymph node metastases.[5] Chemotherapy was the best option for the unresectable LMS and scattered metastases; nevertheless, with a poor general health, she did not tolerate it and died within 3 weeks.[5] The authors also emphasized the LMS resistance to chemotherapy and radiation therapy.

A retrospective study of LMS of IVC from 2014 to 2023 included 13 cases (10 women and 3 men) with a median

[1]MD, PhD. Department of Medicine, Armed Forces Hospital and Catholic University of Brasília-DF, Brazil. Correspondence: vitorinomodesto@gmail.com ORCID: https://orcid.org/0000-0002-7033-6074
[2]MD, Specialist. American Society of Neurophysiology, and Dermatologist of Brasília-DF, Brazil. ORCID: https://orcid.org/0000-0002-4397-3254
[3]MD, Preceptor. Department of General Surgery of State Worker's Hospital, São Paulo-SP, Brazil. ORCID: https://orcid.org/0000-0003-4647-4044

age of 59 years and a median tumor size of 7.0 cm; 9 had microscopically negative surgical margins, and no patient had lymph node metastasis.[6] Seven right nephrectomies and three left renal vein ligations were needed, and the disease-specific survival was 93%; and the authors concluded that IVC management can be safe by ligation, primary repair, or patch angioplasty, depending on the vein patency.[6]

REFERENCES

1. Castellanos LD, Tabbara MM, Livingstone AS, Salerno TA, Gonzalez J, Ciancio G. Unresectable leiomyosarcoma of the inferior vena cava with right atrium tumor thrombus: when to deem this tumor inoperable? A case report and literature review. Frontiers Oncol. 2024; 13:1331896. doi: 10.3389/fonc.2023.1331896
2. Diaz Riverol JM. Radiographic Quiz. Belize J Med. 2024; 13(2). doi: 10.61997/bjm.v13i2.430
3. Gama JM, Almeida R, Oliveira RC, Casanova J. When vessels and sarcomas combine: A review of the inferior vena cava leiomyosarcoma. J Vasc Dis. 2024; 3(1):34-48. doi: 10.3390/jvd3010003
4. Lazar D, Stefan D, Marko D, Zlatanovic P, Sladojevic M, Ilijas C, et al. Case series of the inferior vena cava primary leiomyosarcoma treatment. J Surg Case Rep. 2024; 2024(6):rjad546. doi: 10.1093/jscr/rjad546
5. Mandava A, Koppula V, Kandati M, Reddy AK, Abubacker ZA. Primary leiomyosarcoma of suprahepatic inferior vena cava with metastases. Indian J Nuclear Med. 2024; 39(1):63-65. doi: 10.4103/ijnm.ijnm_130_23
6. Shafique HS, Commander SJ, Blazer DG 3rd, Kim Y, Southerland KW, Williams ZF. Surgical outcomes of patients with inferior vena cava leiomyosarcoma. J Vasc Surg Venous Lymphat Disord. 2024; 101885. doi: 10.1016/j.jvsv.2024.101885

Keywords: Diagnosis; Inferior vena cava; Leiomyosarcoma; Treatment

Palabras clave: Diagnóstico; Vena cava inferior; Leiomiosarcoma; Tratamiento

Funding sources
This work did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Disclosure
No conflicts of interests to disclaim.

Authorship
Vitorino Modesto dos Santos, Taciana Arruda Modesto Sugai, and Lister Arruda Modesto dos Santos made equally substantial contributions to: 1) the conception and design of the study, acquisition of data, and analysis and interpretation of data; 2) the drafting the article and revising it critically for important intellectual content; and 3) the final approval of the version to be submitted.