Primary Leiomyosarcoma of Suprahepatic Inferior Vena Cava with Metastases

Abstract

A 67-year-old female presented with shortness of breath, weight loss, abdomen, and back pain for 2 months. Ultrasound of the abdomen revealed multiple focal liver lesions. ^{18}F-Fluorodeoxyglucose whole-body positron emission tomography/computed tomography revealed a hypermetabolic lesion in the suprahepatic inferior vena cava extending into the right atrium. Multiple hypermetabolic lesions were seen in liver, bones, and abdominal lymph nodes, suggestive of metastases. Histopathology and immunohistochemistry of the lesions revealed it to be metastatic leiomyosarcoma.

Keywords: *Inferior vena cava, leiomyosarcoma, metastases, positron emission tomography/computed tomography, right atrium*

Anitha Mandava, Veeraiah Koppula, Meghana Kandati, Arvind Kumar Reddy, Zakir Ali Abubacker[1]

Departments of Radiodiagnosis and [1]Nuclear Medicine, Basavatarakam Indo American Cancer Hospital and Research Institute, Hyderabad, Telangana, India

^{18}F-Fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) images of a 67-year-old female with histologically confirmed diagnosis of primary leiomyosarcoma of inferior vena cava (IVC) revealed a suprahepatic IVC lesion extending into the right atrium of the heart along with multiple hypermetabolic hepatic, skeletal, and lymph nodal metastases [Figures 1 and 2]. Histopathology of the lesions exhibited round to ovoid smooth muscle cells containing eosinophilic granular cytoplasm with hyperchromatism and nuclear atypia [Figure 3]. Immunohistochemistry was positive for desmin, caldesmon, and smooth muscle actin (SMA) confirming the diagnosis of leiomyosarcoma. Chemotherapy was recommended as the patient had unresectable tumor with multiple metastases. The patient did not tolerate chemotherapy due to her poor general condition and expired within 3 weeks.

Vascular leiomyosarcomas are invasive soft tissue tumors of mesenchymal origin arising from the smooth muscles of blood vessels, more commonly from the veins than arteries. Primary IVC leiomyosarcomas are uncommon vascular neoplasms arising from the tunica media of IVC. They usually present in the fifth to sixth decades with nonspecific symptoms and exhibit female preponderance. The common presenting symptoms are abdominal pain, abdominal distension, back pain, lower extremity edema, Budd–Chiari syndrome, pulmonary thromboembolism, and rarely can lead to sudden death.[1] As these tumors are slow growing, systemic spread and metastases usually occur late in the course of disease. Metastases and recurrences are seen in about 50% of cases.[2] Unlike other soft tissue sarcomas that commonly metastasize to lungs, leiomyosarcomas have a tendency to metastasize to the liver.[3] Metastases to bone and lymph nodes are rare.[3] The location and size of the neoplasm and the presence of metastases determine the resectability of vascular leiomyosarcomas. Based on the site of origin and the relationship to hepatic and renal vessels, IVC leiomyosarcomas are classified into three types: lower segment (involving the IVC below the renal veins), middle segment (renal veins to hepatic veins), and upper segment (hepatic veins to the right atrium).[4] Upper segment leiomyosarcomas involving the suprahepatic IVC and the heart are extremely rare and are considered malignant and unresectable by virtue of their location.[4] Ultrasound is inadequate for complete evaluation of IVC; hence, contrast-enhanced CT and/or magnetic resonance imaging of the abdomen and chest are needed for the evaluation of IVC neoplasms.[5] PET/CT is recommended for detecting the metastatic lesions, defining the

How to cite this article: Mandava A, Koppula V, Kandati M, Reddy AK, Abubacker ZA. Primary leiomyosarcoma of suprahepatic inferior vena cava with metastases. Indian J Nucl Med 2024;39:63-5.

Address for correspondence:
Dr. Anitha Mandava, 1-7-139/75, S. R. K. Nagar, Risalagadda, Musheerabad, Hyderabad - 500 020, Telangana, India.
E-mail: kanisri@gmail.com

Received: 23-11-2023
Revised: 26-12-2023
Accepted: 26-12-2023
Published: 27-03-2024

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.ijnm_130_23

Quick Response Code:

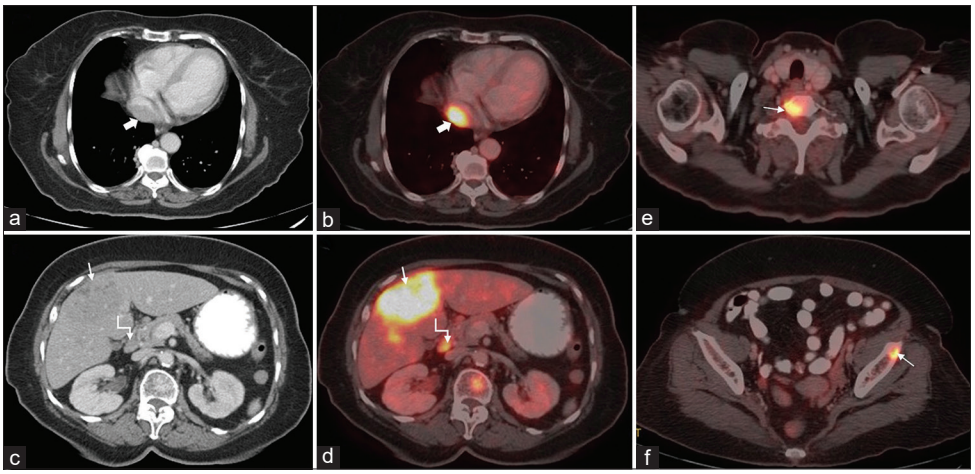

Figure 1: (a-d) Axial contrast-enhanced computed tomography and corresponding positron emission tomography/computed tomography (PET/CT) fusion images of chest and abdomen show mildly enhancing hypoattenuating hypermetabolic lesion in the inferior cavoatrial junction (thick arrows), metastases in liver (arrows), and enlarged precaval lymph node (elbow arrows). (e and f) Axial PET/CT fusion images of the neck and pelvis show bone metastases in vertebral body and left iliac bone (arrows).

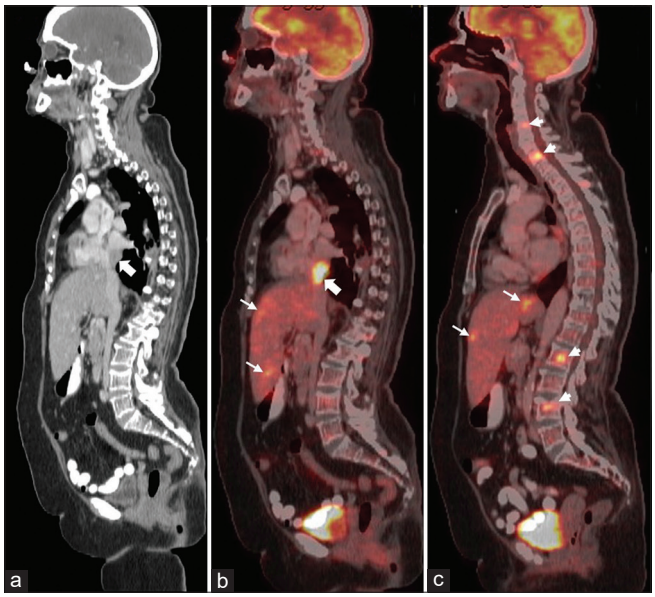

Figure 2: (a-c) Sagittal whole-body computed tomography (CT) and positron emission tomography/CT fusion images show hypermetabolic lesion in the suprahepatic inferior vena cava and right atrium (thick arrow), metastases in liver (arrows) and spine (arrow heads)

extent of lesion and staging of the malignancy.[6] Whole-body PET/CT is useful in differentiating primary IVC leiomyosarcoma from a bland thrombus and is also helpful in identifying tumor thrombus extending from adjacent organ malignancies.[6]

Leiomyosarcomas involving the right atrium are associated with adverse outcomes as cardiac involvement drastically reduces the survival rate.[7] Accurate early diagnosis and *en bloc* surgical resection with venous reconstruction is the only curative option in the management of IVC leiomyosarcomas.[8] Vascular leiomyosarcomas are relatively resistant to chemotherapy and radiation therapy; hence, surgically unresectable metastatic leiomyosarcomas such as in this case have poor prognosis.

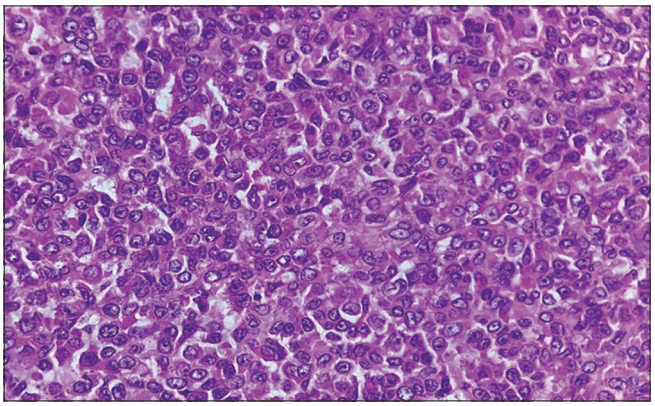

Figure 3: H and E × 40: Round to ovoid tumor cells with eosinophilic granular cytoplasm and irregularly shaped nuclei of leiomyosarcoma

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent Patient identity is not revealed in the manuscript and images.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Wachtel H, Gupta M, Bartlett EK, Jackson BM, Kelz RR, Karakousis GC, *et al.* Outcomes after resection of leiomyosarcomas of the inferior vena cava: A pooled data analysis of 377 cases. Surg Oncol 2015;24:21-7.
2. Abisi S, Morris-Stiff GJ, Scott-Coombes D, Williams IM, Douglas-Jones AG, Puntis MC. Leiomyosarcoma of the inferior

vena cava: Clinical experience with four cases. World J Surg Oncol 2006;4:1.

3. Amin M, Edge SB, Greene F, editors. AJCC Cancer Staging Manual. 8th ed. Chicago: Springer International Publishing, American Joint Commission on Cancer; 2017. p. 517-21.
4. Kulaylat MN, Karakousis CP, Doerr RJ, Karamanoukian HL, O'Brien J, Peer R. Leiomyosarcoma of the inferior vena cava: A clinicopathologic review and report of three cases. J Surg Oncol 1997;65:205-17.
5. Sephien A, Mousa MS, Bui MM, Kedar R, Thomas K. Leiomyosarcoma of the inferior vena cava with hepatic and pulmonary metastases: Case report. J Radiol Case Rep 2019;13:30-40.
6. Ronchi B, Peña GA, Sacchi C. PET/MR: Primary inferior vena cava leiomyosarcoma. Eur J Hybrid Imaging 2022;6:24.
7. Fujita S, Takahashi H, Kanzaki Y, Fujisaka T, Takeda Y, Ozawa H. Primary leiomyosarcoma in the inferior vena cava extended to the right atrium: A case report and review of the literature. Case Rep Oncol 2017;9:599-609.
8. Teixeira FJ Jr., do Couto Netto SD, Perina AL, Torricelli FC, Ragazzo Teixeira L, Zerati AE, *et al.* Leiomyosarcoma of the inferior vena cava: Survival rate following radical resection. Oncol Lett 2017;14:3909-16.