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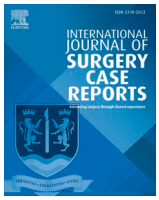

Case report

Leiomyosarcoma of the inferior vena cava presenting with bilateral lower extremity edema with comorbid sarcoidosis: A case report

Dallas Hampton [a,*], Elie Zayyat [d], Francisco I. Macedo [c], Sharon Hook [b]

[a] Medical Student, Eastern Virginia Medical School, Norfolk, VA 23507, USA
[b] Department of Pathology, North Florida Regional Medical Center, Gainesville, FL 32605, USA
[c] Department of General Surgery, North Florida Regional Medical Center, Gainesville, FL 32605, USA
[d] Department of Vascular Surgery, North Florida Regional Medical Center, Gainesville, FL 32605, USA

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ABSTRACT

We present a case of a 70-year-old female with Leiomyosarcoma (LMS) of the inferior vena cava (IVC). Although this is an extremely rare entity, in contradistinction, it is also the most common primary malignancy of the IVC [5]. The patient has a history of sarcoidosis, hypertension, diabetes mellitus type two, and chronic obstructive pulmonary disease (COPD). She presented with a complaint of bilateral lower extremity edema and was admitted where a computerized tomography (CT) scan of the abdomen and pelvis showed a large mass filling the IVC, a finding confirmed by magnetic resonance imaging. Radical resection of the retroperitoneal tumor was carried out including portions of the inferior vena cava with en bloc radical right nephrectomy and right adrenalectomy. The pathologic diagnosis of inferior venal caval leiomyosarcoma (IVC LMS) was made with positive immunostains for desmin, vimentin and smooth muscle actin. The rarity of this entity, clinical presentation along with concomitant sarcoidosis makes this an interesting case.

1. Introduction

Leiomyosarcoma (LMS) is rare tumor comprising around 0.5 % of all adult soft tissue sarcomas and when present in the IVC, it arises from smooth muscle cells of the tunica media within the blood vessel [1]. While LMS is an exceedingly rare tumor, it is the most common primary tumor associated with the IVC [5], typically presenting in females around fifty- to sixty-years of age [6]. There are only a few hundred reported cases of IVC LMS in the literature [4] and little is known in regard to prognosis and treatment for these tumors due to the scarcity of data; however, a study with a pooled data analysis of 377 cases revealed a very high likelihood of relapse though long-term survival is possible [9].

Our patient had an unusual presenting complaint as well as sarcoidosis. The most common symptom seen on presentation is long standing pain in the right flank, 97 % of patients in the above study, while those with lower extremity edema, as in this patient, were considered an inconsistent finding as it was seen in only 32 % of patients in the same study [5]. A search of the literature for patients with sarcoidosis who develop sarcoma showed one case series of eight patients that was published in 2018 [3]. Inclusive of case studies appearing in the literature, our case appears to be the only case of sarcoidosis associated with IVC LMS.

2. Case report

The patient is a 70-year-old female with a history of sarcoidosis, hypertension, diabetes mellitus type two, and chronic obstructive pulmonary disease (COPD) who presented with abdominal pain along with bilateral lower extremity edema. This case was related in accord with SCARE Guidelines [7]. A CT scan with contrast was performed showing a soft tissue mass filling the IVC spanning an area from below the level of the renal veins up toward the hepatic confluence measuring approximately 10 cm in length with extension bilaterally into the renal veins. Additional findings were consistent with deep vein thrombosis (DVT) of the right lower extremity. There were enlarged nodes identified in the mediastinum. An MRI confirmed dilation of the IVC and the left renal vein, which was consistent with thrombosis, along with some peripheral enhancement within the dilated segment of the IVC. Endobronchial ultrasound with fine needle aspiration of the subcarinal and hilar

* Corresponding author at: 2001 SW 117th street 32607, USA.
E-mail addresses: hamptodj@evms.edu (D. Hampton), Elie.Zayyat@HCAhealthcare.com (E. Zayyat), Francisco.Macedo@HCAhealthcare.com (F.I. Macedo), Sharon.Hook@HCAhealthcare.com (S. Hook).

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adenopathy showed non-necrotizing granulomata compatible with sarcoidosis with no evidence of malignancy.

The patient underwent radical resection of the retroperitoneal tumor including resection of a portion of the IVC, en bloc radical right nephrectomy (Fig. 1) and right adrenalectomy. Pathology returned with high grade pathological stage pT3N0 LMS of the IVC (13.0 × 6.8 × 5.5 cm mass) (Fig. 2) and non-necrotizing granulomatous lymphadenitis compatible with sarcoidosis (Figs. 4). Histochemical stains of portal caval lymph nodes were negative for fungus (GMS) and mycobacteria (AFB) and are nonreactive with appropriate control. Tumor extended into the right and left renal vein with involvement of the right kidney. LMS abutted the adrenal without direct extension into the parenchyma. The IVC was reconstructed just inferior to the right hepatic vein with total cross-clamp time of 39 min. Tumor diagnosis was confirmed by positive immunostaining with desmin (Fig. 3), vimentin and smooth muscle actin, while CD117, pankeratin, and GATA3 were non-immunoreactive. Final margins were determined to be clinically negative. A single regional lymph node was submitted and was negative for metastasis. The postoperative course was complicated by an upgrade to ICU due to respiratory distress with subsequent chest tube placement on day eight, but she was returned to the progressive care unit by day 10 after remaining hemodynamically stable during her ICU stay. She was eventually discharged to a rehabilitation center with no post-surgical oncologic treatment. The patient was discussed twice at a multidisciplinary tumor board.

3. Discussion

This is an unusual case for a few reasons, first of all, IVC LMS is a rare tumor with only a few hundred documented cases. Secondly, this tumor most often presents as vague flank pain. Our case presented with bilateral lower extremity edema secondary to tumor thrombus in the IVC, an uncommon symptom but adding the possibility of tumor to this

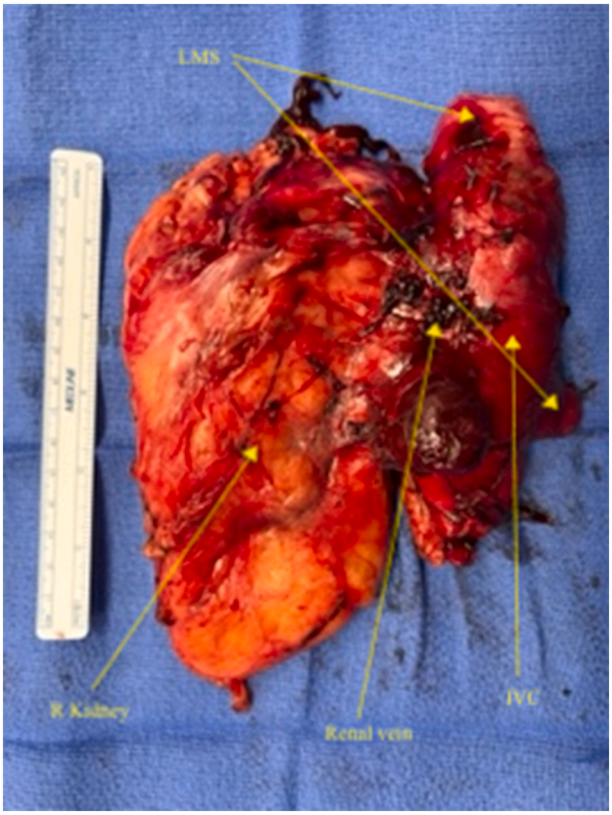

Fig. 1. Gross specimen of leiomyosarcoma of the inferior vena cava including portion of the IVC, left renal vein, and right kidney after resection.

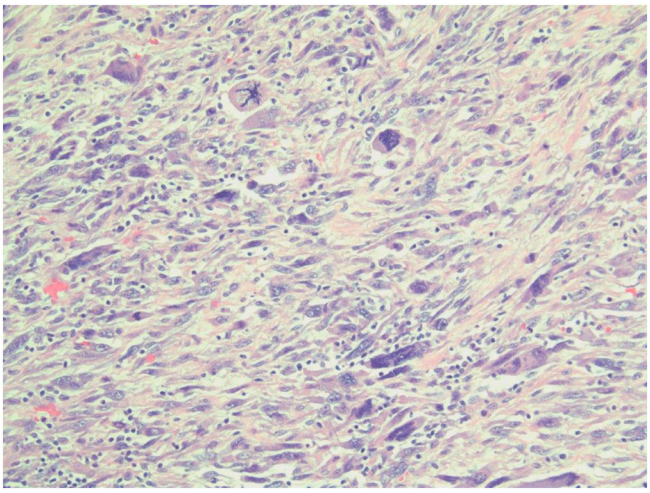

Fig. 2. Hematoxylin and Eosin stain of high grade leiomysarcoma of inferior vena cava, 200× magnification.

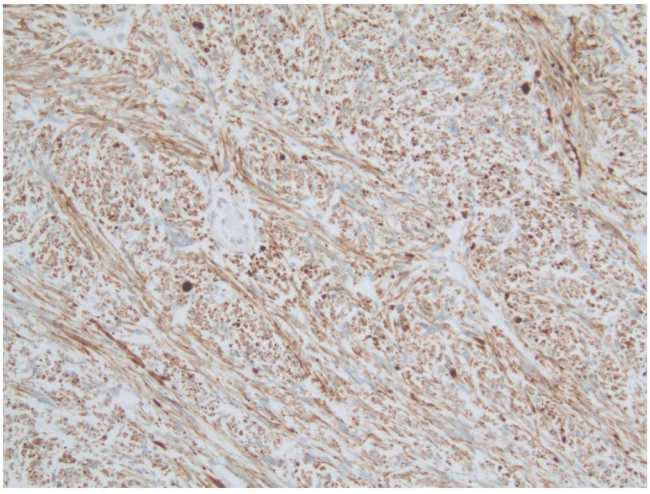

Fig. 3. Positive desmin immunohistochemical stain, 200× magnification.

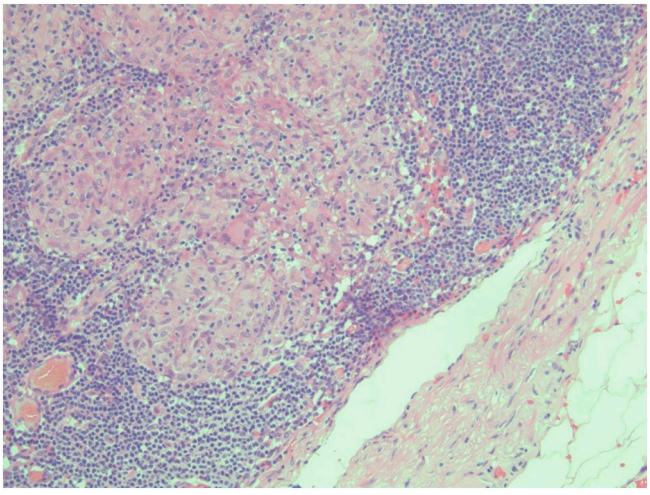

Fig. 4. Hematoxylin and Eosin stain of non-necrotizing granulomatous lymphadenitis, 200× magnification, portal caval lymph node.

differential diagnosis is paramount. A last factor is the newly diagnosed sarcoidosis. Found incidentally upon biopsies that were performed in an attempt to exclude metastasis of the IVC tumor within lymph nodes. There are few references citing sarcoidosis in conjunction with sarcoma yet no cases that specifically show IVC LMS. It has been postulated that long standing inflammation including sarcoidosis can lead to the development of malignant tumors, specifically carcinoma [8]. However, there are several case reports and a limited series that have suggested a non-random association between sarcoma and sarcoidosis. The link between sarcoidosis and sarcoma has yet to be determined but the importance of recognizing this possible connection is that the presence of sarcoidosis within lymph nodes, as in our case, can mistakenly lead to higher tumor stage due to these nodes being interpreted clinically as metastasis [3] thus highlighting the necessity of pathologic diagnosis in these cases.

4. Conclusion

We report a rare case of leiomyosarcoma of the inferior vena cava that presented with bilateral lower extremity edema with a comorbid diagnosis of sarcoidosis. This unique presentation adds an additional case of a rare tumor to the medical literature that can assist in development of future clinical treatment and diagnosis. The association of sarcoidosis with sarcoma is currently tenuous at best but when present should be considered as the etiology for nodal enlargement and require a diagnostic biopsy for stage determination.

Ethics statement including patient consent statement

Written informed consent was obtained from the patient for 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.

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Guarantor

Dr. Sharon Hook.

Consent

Written informed consent was obtained from the patient for 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.

Credit authorship contribution statement

Dallas Hampton- writing the manuscript.

Sharon Hook- writing the manuscript, supervised and validated the case data, and pathologist who read the case.

Elie Zayyat-participated as vascular surgeon on the case, conceptualization.

Francisco I Macedo- participated as general surgeon on the case, conceptualization.

Declaration of competing interest

To the authors knowledge there are no competing personal or financial relationships that could appear to influence the paper.

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