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ORIGINAL ARTICLE

An Imaging-Based Assessment of Inferior Vena Cava Leiomyosarcoma: A Case Series

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Abstract
Primary inferior vena cava(IVC) leiomyosarcoma (LMS) is an extremely rare retroperitoneal mesenchymal tumour with < 500 cases reported in literature so far. Patients always present with nonspecific complaints, hence delaying the actual diagnosis, and they are associated with poor oncological outcomes. Differentiating primary leiomyosarcoma from other pathologies affecting IVC-like bland thrombus, retroperitoneal liposarcomas and other tumours infiltrating IVC secondarily may be difficult at times. We present here our experience with five cases of IVC leiomyosarcoma, who presented to us during a 6-year period.

Keywords Leiomyosarcoma · Inferior vena cava · Diagnostic imaging

Introduction

Leiomyosarcoma is a malignant neoplasm originating from mesenchymal tissue and most frequently occurs in the uterus and gastrointestinal system. Primary inferior vena cava leiomyosarcoma (IVC LMS) is extremely rare. It usually presents as large tumour at the time of diagnosis due to delayed diagnosis. Prognosis is typically poor, and definitive treatment is surgical resection with clear margins [1]. The 5-year survival rate ranges between 31 and 66.7% for patients with IVC LMS following complete macroscopic resection [2]. However, recurrence rates are as high as 50%, and thus, surgery may simply be providing palliation in most of the cases [3].

Case Reports

Case 1

A 69-year-old lady presented with a short-term history of vague abdominal pain and weakness. Ultrasonography (USG) of the abdomen revealed an 8 × 9-cm mass, which was suspected to arise from the right kidney. Computed tomography (CT) scan was performed to further characterise the origin of the mass, to demonstrate the extent and to look for any metastases. It revealed a predominantly hypoenhancing mass arising from the vena cava and extending into the intrahepatic segment of IVC (Fig. 1). Histological examination revealed it to be high-grade leiomyosarcoma composed of spindle cells with positive immunohistochemistry staining for SMA (smooth muscle actin), Desmin and HHF35 (muscle-specific actin). Surgical en bloc excision was done, and IVC reconstruction was done using an 18-mm Dacron prosthetic graft. Within 8 months, she returned with similar complaints, and a CT scan revealed a recurrence of the tumor at the post-operative site. Currently, the patient is on chemotherapy.

Case 2

A 38-year-old gentleman presented with the chief complaints of vague chest pain and abdominal discomfort. Chest X-ray revealed few well circumscribed random nodules in the left lung which were suspicious for metastases. Hence, CECT of the chest and abdomen was performed which revealed random solid nodules in the lung along with an 8.3 × 7.4 × 6-cm multilobulated heterogeneous mass in the right hemiabdomen encasing the IVC and infiltrating into the retroperitoneal space (Fig. 2). CT guided biopsy was performed which revealed it to be high grade LMS. The neoplastic cells were positive for smooth muscle actin, desmin and caldesmon. The patient was started on chemotherapy

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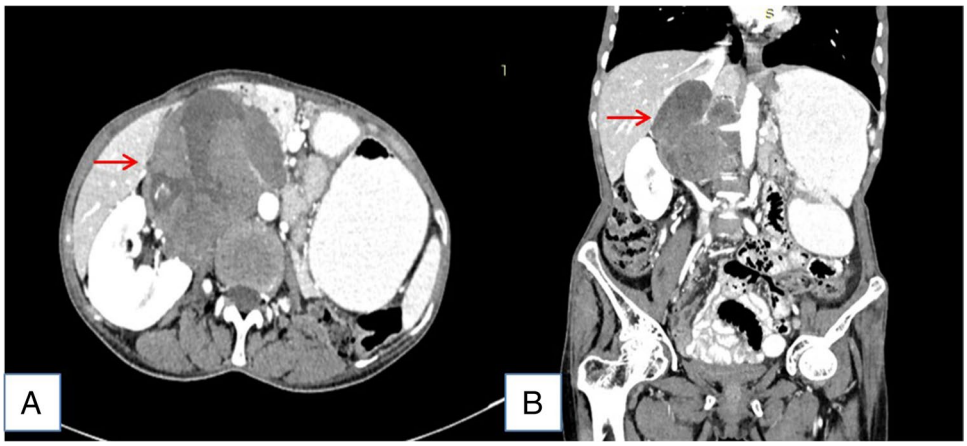

Fig. 1 CT of 69-year-old female patient of biopsy proven IVC leiomyosarcoma. CECT axial and coronal reformatted images (**A**, **B**) show a heterogeneous mass in aortocaval location involving the left wall of infrahepatic IVC

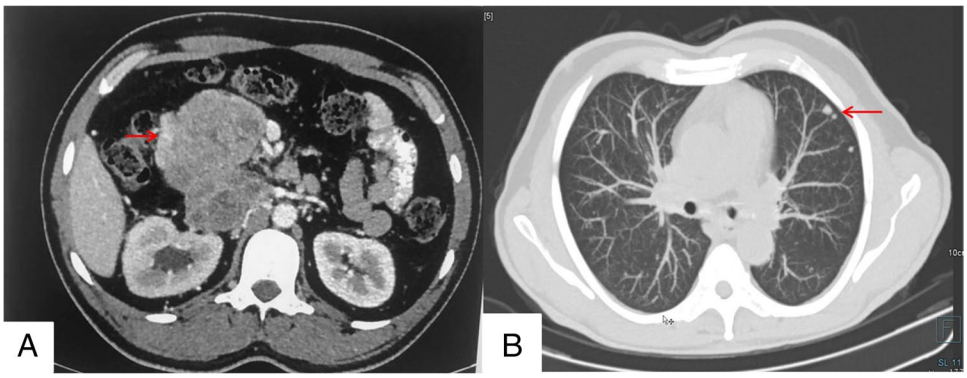

Fig. 2 CT of 38-year-old male patient of IVC LMS. CECT axial abdomen image (**A**) shows a heterogenous retreoperitoneal mass involving IVC. Chest image (**B**) shows left lung metastasis

thereafter; however, he succumbed to the disease within 6 months of initiating treatment.

Case 3

A 65-year-old lady presented with complaints of abdominal discomfort which persisted for 5 months, and bilateral symmetrical pedal oedema for half a month. Laboratory parameters revealed raised serum lactate dehydrogenase and CA-125 level. Ultrasonography was suggestive of a solid mass in the retroperitoneum which was suspected to be arising from the vena cava. The patient underwent dual-phase CT which revealed the lesion to be heterogeneous mass within the infrahepatic IVC reaching inferiorly till bilateral common iliac veins along with deposits in the right pleural cavity (Fig. 3). Hence, the disease was found to be unresectable, and the patient was started on chemotherapy.

Case 4

A 33-year-old gentleman presented with a history of progressive abdominal pain mainly in the right flank, nausea and bilateral pedal oedema. Ultrasound of the abdomen revealed a $7.3 \times 5.4 \times 6.3$-cm solid heterogeneous mass in the retroperitoneum with significant internal vascularity. Contrast CT of the chest and abdomen showed a heterogeneous intraluminal mass involving the infrarenal segment of IVC (Fig. 4). CT-guided biopsy was performed which revealed it to be LMS, following which the patient was started on chemotherapy.

Case 5

A 46-year-old man presented with OPD with complaints of right flank pain. Physical examination was essentially normal. X-ray of the KUB region was performed to rule out renal calculus. USG was also done which revealed right-sided mild hydronephrosis with dilated upper ureter and a soft tissue encasing the mid ureter. Further, the patient underwent CECT of the abdomen which revealed a heterogeneous mass in the infrarenal IVC with intra as well as extraluminal component which was seen to encase the abdominal aorta (Fig. 5). CT-guided biopsy of the lesion revealed it to be LMS, and the patient was started on chemotherapy thereafter. The clinic-radiological demographics of all five cases have been summarised in Table 1.

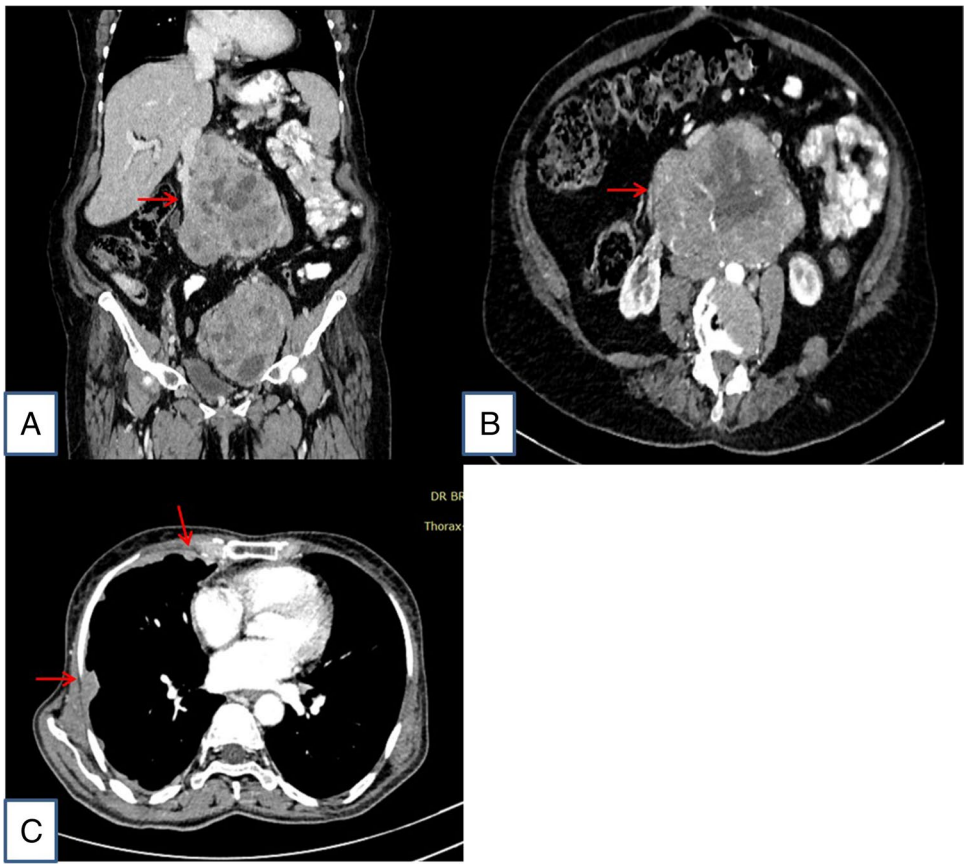

Fig. 3 CT of 65-year-old male patient of IVC LMS. CECT images of abdomen (**A**, **B**) show heterogeneous extraluminal mass extending along IVC from its intrahepatic location to bifurcation into iliac veins. Chest images (**C**) show deposits along right pleura

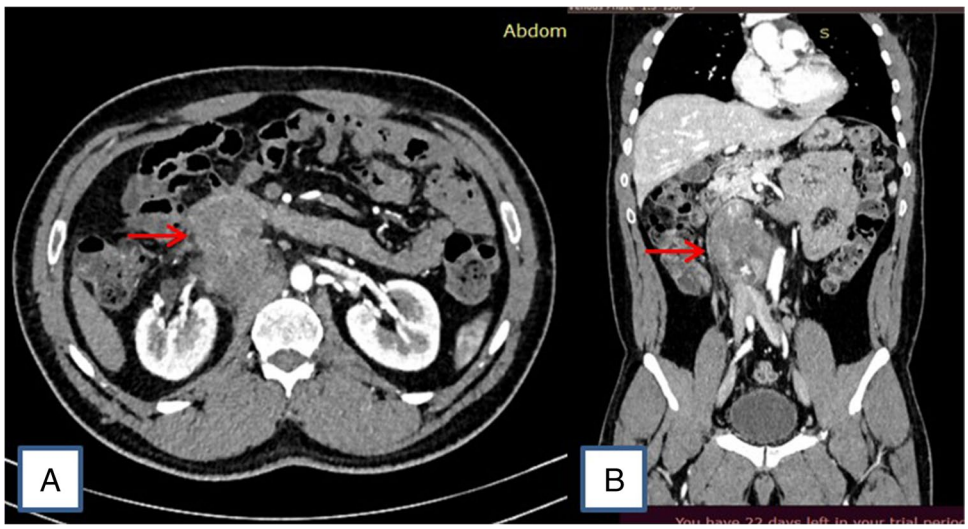

Fig. 4 CT of 33-year-old male patient of IVC LMS. CECT axial and coronal reformatted images shows heterogeneous solid mass in the infrahepatic segment of IVC

Discussion

Both primary and secondary malignancies can involve the IVC and often have similar imaging appearance. Most of the IVC masses are either direct extensions from tumours like hepatocellular carcinoma (HCC), renal cell carcinoma (RCC) and adrenocortical carcinoma or intraluminal thromboembolization. Leiomyosarcoma of the IVC, though rare, is the most common primary tumour of the IVC. It arises from the smooth muscle cells in the vessel wall. IVC leiomyosarcoma predominantly occurs in females with a female/male ratio of approximately 3:1

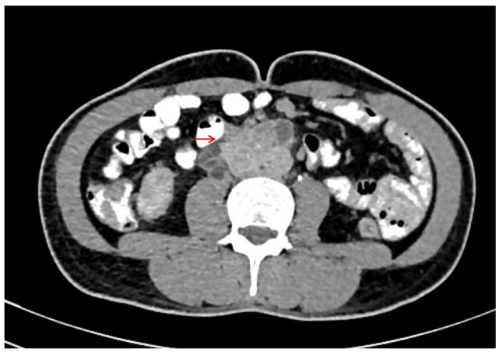

Fig. 5 CT of 46-year-old male patient of IVC LMS. CECT axial and coronal reformatted images shows heterogeneous solid mass in the infrahepatic segment of IVC with both extra and intraluminal component

[4]. Although it can occur in any age, it is more commonly seen in middle-aged females in their fifth to sixth decades of life.

Three patterns of IVC leiomyosarcoma growth have been identified: extraluminal, intraluminal and mixed, with extraluminal pattern being the most common form [5]. Four out the five patients in our study showed mixed pattern of growth with both intra and extraluminal component. It is also being classified into three types based on the segment involved: segment I—lower, infrarenal (34% of tumours) till iliac bifurcation, segment II—middle, inter- and suprarenal up to but not including the main hepatic veins (42% of tumours) and segment III—upper, from the hepatic veins to the right atrium (least common location, 24% of tumours) [6].

Clinical manifestation which mainly depends on the location of the tumour appeared to be nonspecific, including abdominal pain, back ache, lower limb oedema/pain, renal hypertension and dyspnoea. Tumour extension into the right atrium can cause atrial fibrillation. Rarely, some cases present with Budd chiari syndrome as tender hepatomegaly, jaundice and ascites. Metastatic disease occurs in up to half of the cases at presentation, most commonly involving liver followed by lung and lymph nodes [7]. In our case series, one patient had lung metastases and another had multiple pleural deposits. The pleural deposits in inferior vena cava (IVC) leiomyosarcoma are quite rare. The rarity of the tumour and absence of specific signs and symptoms make diagnosis of IVC leiomyosarcoma challenging. Correct identification of the exact longitudinal extent of the IVC involvement is important for staging and determining surgical approach and intervention. Role of radiologist is important in establishing diagnosis, demonstrating the anatomy and extent of the disease as well as to perform image-guided biopsies. Key imaging modalities used in diagnosis, staging and surveillance of the disease include computed tomography (CT) and magnetic resonance imaging (MRI) either along or in conjunction with cavography, ultrasound and PET-CT.

Sonographic features of IVC leiomyosarcoma are nonspecific. Due to retroperitoneal location, large body habitus and operature dependence, many a times, it is difficult to evaluate the lesion. Doppler evaluation of the lower limb veins may show abnormal reversal of waveform due to upstream obstruction.

CT is the workhorse imaging modality in oncology due to its excellent spatial resolution, easy availability and cost. To our knowledge, there is no specific protocol in the literature for imaging IVC tumours. At our institute, a dual-phase CT is routinely done in all suspected cases of IVC leiomyosarcoma consisting of arterial and venous phase. A delayed scan is acquired on case to case basis. Arterial phase (20–25 s post contrast injection) helps in better assessment of the relation of the tumour to adjacent major arterial structures for pre-operative planning and also in detection of hypervascular liver metastases. Portal venous phase (70–90 s post contrast injection)

Table 1 Clinico-radiological demographics of the patients

Case	Age	Sex	Presenting symptoms/duration	Metastases	Segment involved	Outcome	Management	Biopsy performed
1	69	Female	Vague abdominal pain, 2 months	No	Segments II and III	Recurrence in retroperitoneum	Surgical resection	Yes
2	38	Male	Chest pain and abdominal discomfort, 6 months	Lung metastases	Segments I, II and III	Succumbed	Chemotherapy	Yes
3	65	female	Abdominal discomfort and bilateral pedal oedema, 3 months	Present in right pleural cavity	Segments II and III	On treatment	Chemotherapy	Yes
4	33	Male	Abdominal pain and pedal edema, 4.5 months	No	Segments II and III	On treatment	Chemotherapy	Yes
5	46	Male	Right flank pain, 9 months	No	Segment III	On treatment	Chemotherapy	Yes

demonstrates better opacification of the IVC and the tumour. Common CT findings include tumour thrombus within the IVC, expansion of the lumen and lack of direct continuity between the tumour thrombus and any other adjacent primary retroperitoneal structures. Prominent collaterals may be seen around the lesion, especially in slow-growing masses. Usually, they are not associated with regional or distant lymphadenopathy.

MRI provides superior contrast resolution, allowing improved characterisation of tumour composition. In contrast to bland thrombus, intraluminal leiomyosarcoma expands the lumen, appears iso to hyperintense on T2-weighted images and enhances on post-contrast T1-weighted images. Because of increased cellularity, it may show diffusion restriction on DWI images and low apparent diffusion coefficient. In addition, MR venography provides accurate anatomical details and blood flow dynamics which are essential for planning surgery.

PET CT can help differentiate between IVC tumour and bland thrombus as bland thrombus will show low FDG uptake while leiomyosarcoma will have higher uptake. Furthermore, for patients receiving therapy, PET-CT can offer crucial data on treatment response and recurrence surveillance.

To avoid unnecessary diagnoses, it is crucial to recognize common pitfalls in imaging the inferior vena cava (IVC). A notable pitfall is flow-related phenomenon at the level of renal veins, where contrast mixing with unenhanced blood may mimic thrombosis on contrast CT. Such artefacts can also been seen in cases of right heart failure or when contrast injection rate exceeds 3 ml/s. Another rare and recently described phenomena, "Pseudolipoma," is a partial volume artefact from pericaval fat above the caudate lobe, which is frequently observed in cirrhotic patients. Differentiating IVC leiomyosarcoma from other retroperitoneal malignancies still remains challenging for the radiologists. Few signs like positive embedded organ sign have been found to be useful in identifying masses arising from plastic organs like vessel and bowel. According to Webb et al. [8], the most useful sign in the identification of a retroperitoneal mass as an IVC leiomyosarcoma was an imperceptible caval lumen. Angiosarcoma of right atrium can sometimes extend into IVC and mimic IVC leiomyosarcoma. However, it is often associated with regional lymphadenopathy, and the epicentre of the mass in is heart, differentiating it from in IVC LMS. Another common differential is intravenous leiomyomatosis which usually is associated with uterine leiomyomas.

Because of the variation in the anatomical segment involved, different approach and surgical intervention techniques are usually employed for specific case. The management of these tumours has been, where possible, by means of radical en-bloc resection with an aim of achieving clear margins. Although adjuvant radiation therapy may decrease the risk of local recurrence, it does not improve the overall survival [9]. Surgical options include complete resection and graft placement, partial resection and primary cavoplasty and ligation of IVC.

Factors associated with worse prognosis in IVC leiomyosarcoma include the presence of lower limb oedema, tumour involvement of the upper segment, intracardiac extension, Budd Chiari syndrome and residual postsurgical macroscopic disease. On the other hand, complete radical resection, the absence of a palpable mass and the presence of abdominal pain are associated with better prognosis [10].

Conclusion

In conclusion, LMS of IVC is a very rare aggressive retroperitoneal sarcoma with no specific clinical complaints. A heterogeneous enhancing mass along the IVC should be a warning and possibility of IVC LMS should always be considered. However, histopathology may be required for definitive diagnosis. Radical en-bloc surgery is the only curative option available with a view to obtaining negative resection margins.

Data Availability On request.

Declarations

Conflict of Interest The authors declare no competing interests.

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