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CASE SERIES

Imaging of Leiomyosarcoma of the Inferior Vena Cava: A Comparison of 3 Cases

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Abstract

Leiomyosarcoma of the inferior vena cava is a rare tumour arising from the smooth muscle fibres of the media with a mean size at diagnosis generally around 12cm (range 2-38cm). Leiomyosarcoma of the inferior vena cava is a rare clinical entity that has been described in fewer than 300 patients in the literature[1]. Leiomyosarcoma is the most common primary tumour of the inferior vena cava and the second most frequent retroperitoneal neoplasm in adults. This study compares 3 cases of Leiomyosarcoma of the inferior vena cava discovered incidentally and with symptomatic late-stage disease.

Keywords: Inferior Vena Cava, Leiomyosarcoma of the inferior Vena Cava, Magnetic Resonance Imaging, Mesenchymal Tumour, Multidetector Computed Tomography

1. Introduction

Leiomyosarcoma (LMS) of the Inferior Vena Cava (IVC) represents an exceptionally rare malignant neoplasm, constituting less than 1% of all soft tissue sarcomas. Despite its rarity, it is recognised as the most prevalent primary tumour affecting the IVC. This mesenchymal tumour arises from the smooth muscle layer (tunica media) of the venous wall and predominantly presents in female patients, most commonly during their sixth decade of life.

The clinical and radiologic manifestations of IVC LMS are influenced by the tumour's anatomical site of origin and the direction of its growth, which may be intraluminal, extraluminal, or both.

Surgical resection with clear (tumour-free) margins via *en bloc* excision remains the cornerstone of treatment. However, complete surgical removal is often challenging due to delays in diagnosis, which are attributed to the tumour's indolent progression and its typical retroperitoneal location—an area where clinical symptoms are often vague or nonspecific in the early stages.

Imaging studies play a pivotal role in diagnosis and preoperative planning. The imaging characteristics of three distinct LMS cases: one involving a small, incidentally discovered tumour measuring 6cm detected by Multidetector Computed Tomography (MDCT), and two others involving larger, symptomatic tumours, one diagnosed using both MDCT and Magnetic Resonance Imaging (MRI) and the other with MDCT.

2. Aim and Objectives

To compare the different clinical and radiological presentations of leiomyosarcoma of the inferior vena cava

3. Review of Literature

Leiomyosarcoma of the IVC is a rare and aggressive vascular tumour originating from the smooth muscle cells of the IVC wall, specifically the tunica media. While soft tissue sarcomas themselves are uncommon, LMS of vascular origin is rarer still, with the IVC being the most commonly affected blood vessel. The first known description of this entity dates back to Perl in 1871, and since then, fewer than 500 cases have been

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reported in the international literature, highlighting its rarity and the importance of cumulative published experience. IVC LMS occurs more frequently in middle-aged individuals, with a noted female preponderance (approximately 70% in multiple series). The condition is typically diagnosed between the ages of 50 and 60 years. Risk factors remain unclear, although there are hypotheses involving hormonal influences and chronic vascular injury; causality is not firmly established. Mingoli *et al*. classified IVC LMS based on its location relative to hepatic and renal veins, dividing the tumour into: Segment I (lower) – below the renal veins, Segment II (middle) – between the renal and hepatic veins, Segment III (upper) – involving the suprahepatic IVC to the right atrium[2].

This anatomical classification is not only descriptive but also prognostically and surgically significant. The tumour's location influences both symptomatology and resectability. Segment I tumours may present with Deep Vein Thrombosis (DVT), leg oedema, or pelvic mass. Segment II involvement often causes vague abdominal or back discomfort. Segment III tumours, due to proximity to the heart and hepatic vasculature, may result in Budd-Chiari syndrome, hepatomegaly, or even cardiac symptoms. Due to the retroperitoneal location of the IVC and the tumour's slow, indolent growth, symptoms are usually nonspecific and occur late. This contributes to diagnosis typically being made at an advanced stage. Imaging plays a central role in both diagnosis and surgical planning. MDCT and MRI are the primary modalities, offering high-resolution cross-sectional assessment of the tumour's size, extent, intravascular involvement, and relationships with adjacent organs. MRI is particularly useful for defining tumour margins, vascular invasion, and soft-tissue characterisation, often showing heterogeneous enhancement, necrotic areas, and extraluminal expansion. Histologically, LMS is composed of intersecting fascicles of spindle cells with eosinophilic cytoplasm, elongated nuclei, and high mitotic activity. Immunohistochemistry is critical for definitive diagnosis, showing positive staining for Smooth Muscle Actin (SMA), desmin, and h-caldesmon, while being negative for epithelial and neural markers.

The cornerstone of treatment remains radical surgical resection with clear margins (R0). Preoperative biopsy is often avoided due to bleeding risk and the potential for tumour seeding unless radiologically feasible and necessary. Several surgical techniques have been documented, including: *En bloc* tumour resection, IVC reconstruction with grafts, Polytetrafluoroethylene (PTFE) or autologous, and multivisceral resections when adjacent organs are involved (e.g., kidney, liver, adrenal gland). In the series by Kieffer *et al*. (2006) involving 22 patients, radical surgical resection achieved a 5-year survival of 57%, although recurrence remained common. Notably, resection of suprarenal or suprahepatic tumours carries increased surgical complexity and higher perioperative risk[3,4].

Despite complete resection, prognosis remains guarded due to high local recurrence and occasional distant metastasis, particularly to the lungs and liver. Recurrence rates are reported as high as 50–70% at 5 years, with median survival ranging between 25–50 months depending on the series. In Mingoli *et al*.'s registry review (1992) of 218 cases of IVC LMS, patients with complete resection had significantly better outcomes than those with residual disease. Pawlik *et al*. in a single-centre Armed Forces Institute of Pathology (AFIP) study showed a 5-year survival approaching 60% in patients undergoing complete surgical excision. The presence of macroscopic residual disease, segment III tumours, and non-reconstructible vascular involvement was identified as a key factor negatively affecting survival.

The role of adjuvant therapies (chemotherapy and radiotherapy) remains controversial and not well standardised. While Doxorubicin and Ifosfamide-based regimens have been used in soft tissue sarcoma protocols, evidence for their effectiveness in IVC LMS is anecdotal or based on retrospective small series. Radiotherapy may be considered in cases with positive margins or recurrence, although risks of damage to adjacent vital structures are significant[4].

Recent case reports and small cohort studies describe the use of targeted therapies, such as Pazopanib, for unresectable or metastatic LMS. Immunotherapy checkpoints (e.g., PD-1 inhibitors), though efficacy data remain limited, *ex vivo* liver resections and autotransplantation in centres with advanced surgical expertise

Such innovative strategies are promising yet remain investigational and technically demanding. Support

from a multidisciplinary tumour board involving oncologists, liver/vascular surgeons, and interventional radiologists is essential for optimal care.

4. Materials and Methods

Three cases of IVC leiomyosarcoma at the Department of Radiodiagnosis of this institution were evaluated.

Two cases underwent triple-phase contrast-enhanced MDCT using a 128-slice GE computed tomography scanner. 100 – 120ml of non-ionic iodinated contrast (Omnipaque) was administered at a rate of 3-5 ml/s using a power injector. Arterial phase, portal venous phase and delayed phase images were taken at 35-45 seconds, 60-75 seconds and 2-5 minutes after contrast administration, respectively.

Another one with both MDCT and MRI, including Half-Fourier-Acquired Single-shot Turbo Spin Echo (HASTE) with sequential acquisition of high-resolution T2-weighted images, True Fast Imaging with Steady-State Free Precession(TRUFI), Time of flight and Diffusion Weighted Imaging (DWI) and Apparent Diffusion Coefficient (ADC) with High b-values (800s/mm^2) sequences were performed.

5. Results (Including Observations)

3 cases of Leiomyosarcoma of the inferior vena were observed. First case, a 45-year-old female who was a known case of ureteric calculi came for follow-up imaging with no specific complaints. Contrast-Enhanced Computed Tomography (CECT) performed on a 128-slice scanner showed an incidentally detected, rounded, solid retroperitoneal mass (6cm) with lobulated margins, which involved the space between the pancreas, aorta and right kidney. The lesion showed heterogeneous contrast enhancement on arterial, porto-venous and venous phase with a filling defect in the IVC (Figures 1 and 2). Postoperative excision histopathology showed a moderately circumscribed, encapsulated fibrous tumour with a pattern of intersecting sharply marginated groups of spindle cells, with palisaded arrangement. The second case was clinically presented with right hypochondrial pain with loss of weight. CECT showed a little larger lesion completely replacing the infrahepatic IVC, measuring 9cm in its largest diameter, with areas of necrosis, and

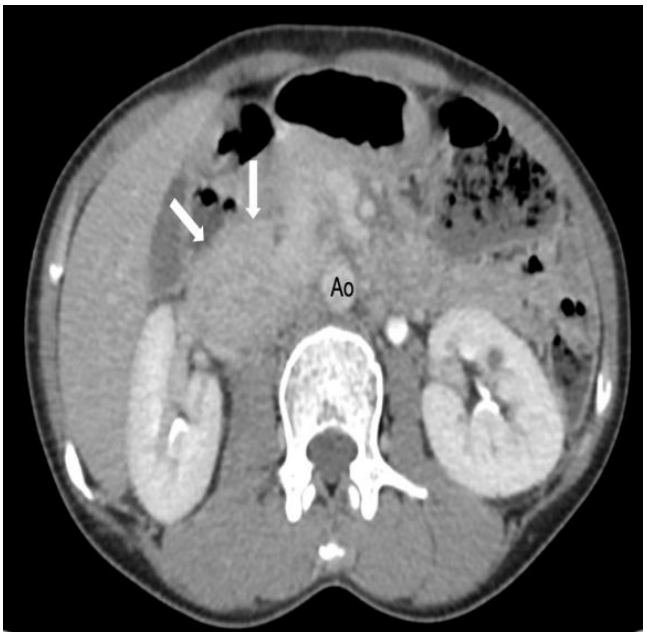

Figure 1. Venous phase CT shows a rounded, predominantly homogenously enhancing solid retroperitoneal mass (6cm) with lobulated margins(arrow), which involves the space between the pancreas, aorta and right kidney.

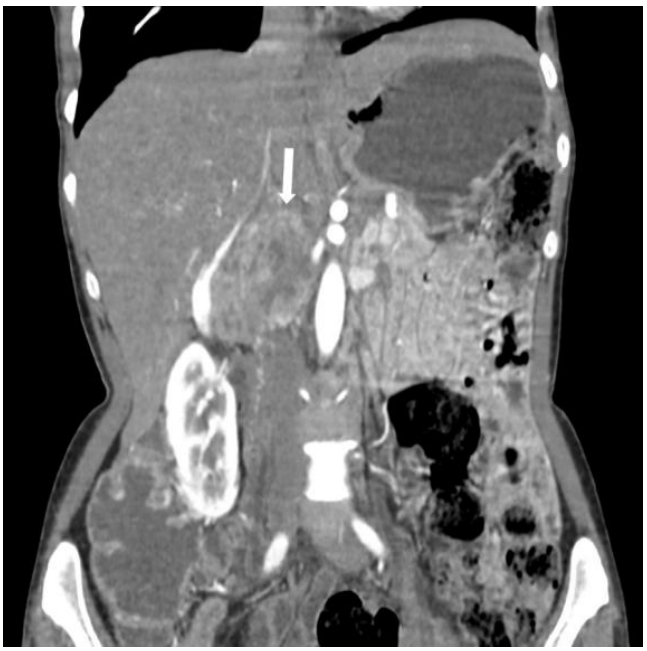

Figure 2. Arterial phase CT shows a heterogenously enhancing solid retroperitoneal mass with lobulated margins(arrow), displacing the right renal artery.

MRI showed a similar lesion arising from the IVC with an intraluminal, extraluminal component and diffusion restriction. The Time of Flight (TOF) image

showed minimal eccentric flow in the IVC (Figures 3 to 10). The third case was presented with abdominal distension with firm palpable swelling in the right hypochondrial region. Triple phase CECT performed showed a large lobulated heterogeneously enhancing lesion involving the right hypochondrium and pelvis

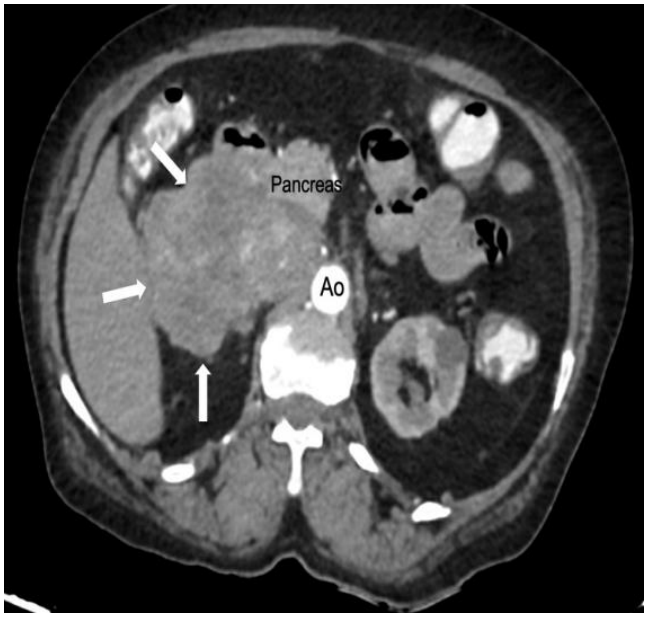

Figure 3. Arterial phase CECT shows a heterogenously enhancing mass lesion with lobulated margins and areas of necrosis seen arising from IVC(arrow), adjacent to the head of pancreas and aorta compressing the second part of the duodenum.

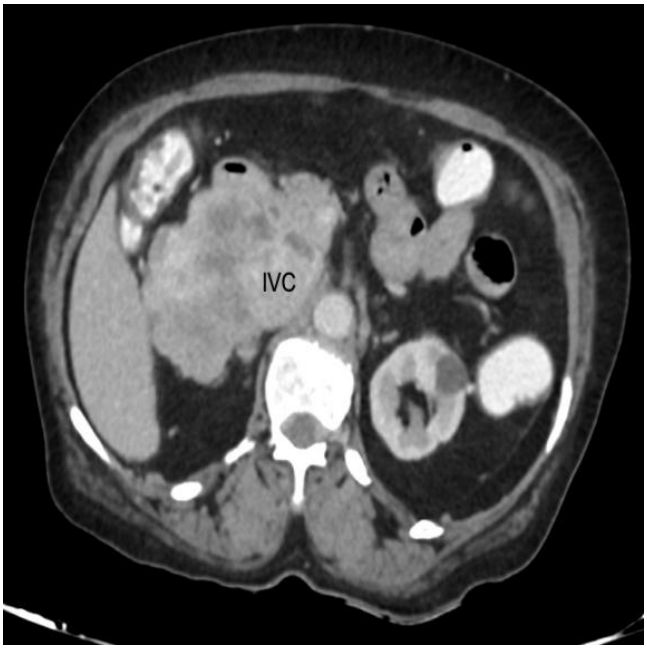

Figure 4. Venous phase CECT shows a heterogeneously enhancing mass lesion with areas of necrosis seen arising from the IVC, showing an organ-embedded sign.

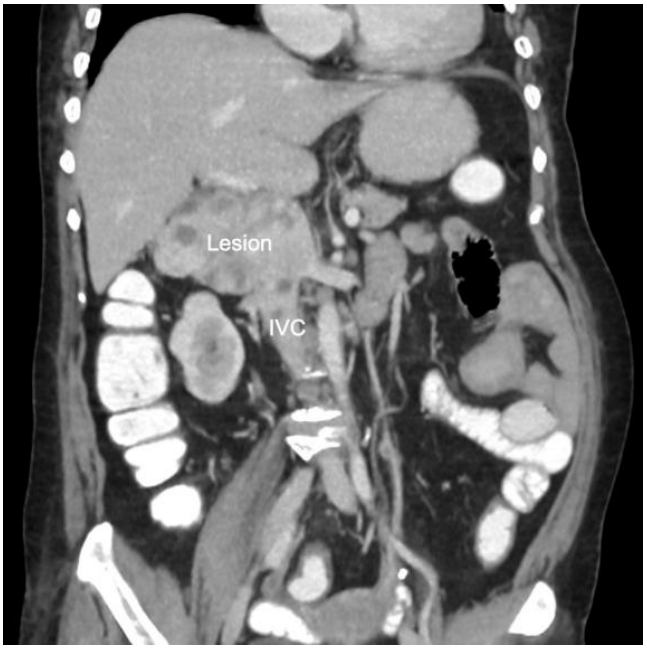

Figure 5. Venous phase coronal section CECT shows a heterogenously enhancing mass lesion with areas of necrosis seen arising from the IVC.

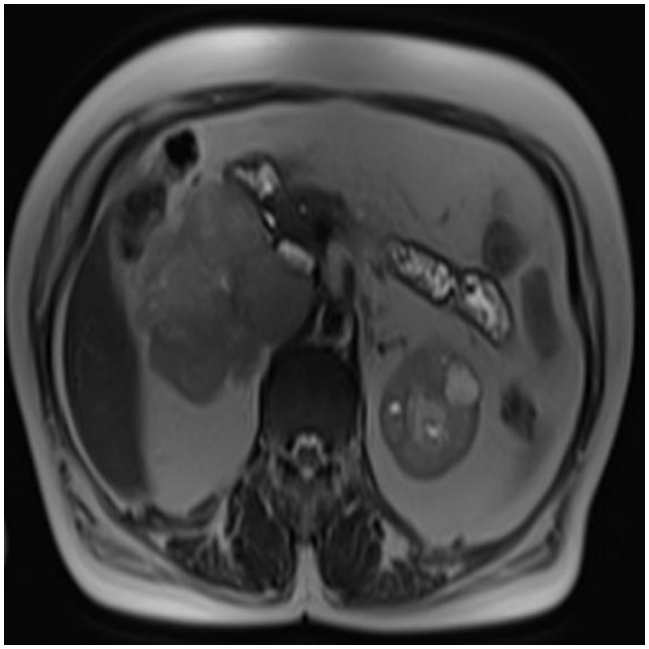

Figure 6. T2 haste shows a heterointense mass lesion with lobulated margins arising from the IVC, adjacent to the head of the pancreas and aorta, compressing the second part of the duodenum.

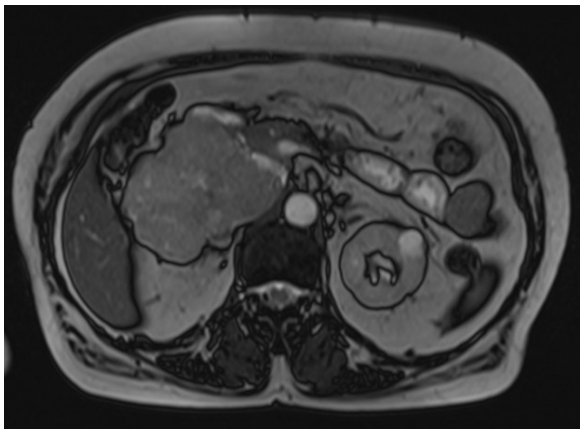

Figure 7. T2 trufi axial shows the seen lesion arising from the IVC.

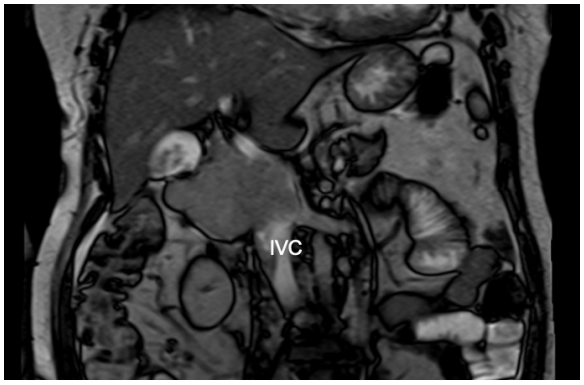

Figure 8. T2 trufi coronal shows a lesion arising from the IVC with an intraluminal component and a filling defect.

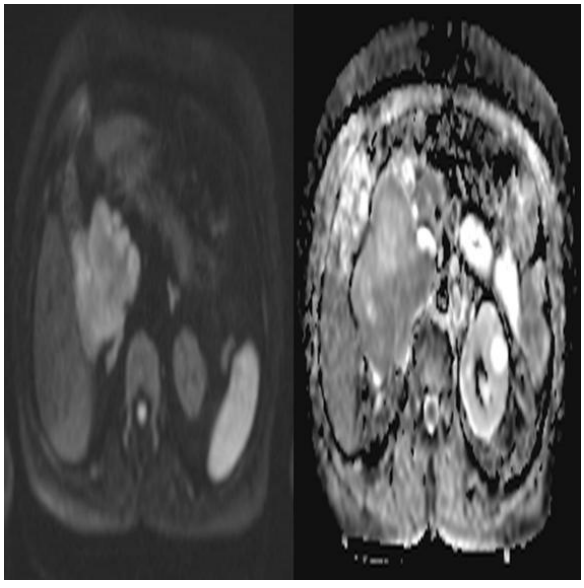

Figure 9. Diffusion weighted images show significant diffusion restriction with a drop in ADC.

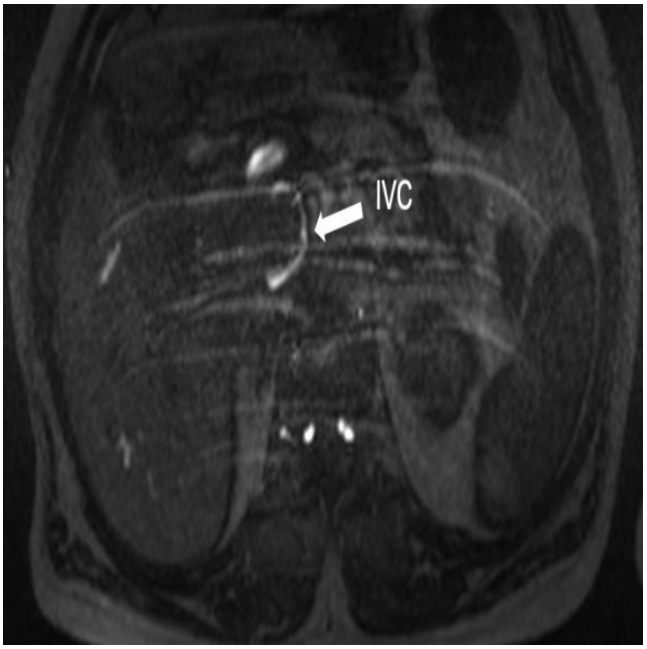

Figure 10. TOF axial shows an intraluminal component and a filling defect with eccentric flow in the IVC.

with areas of necrosis. The lesion measured 22.5(CC) x16.1(AP)x15.2(TR)cm. Superiorly, it extended up to the inferior surface of the liver and displaced the bowel loops superolaterally. Medially, the lesion crossed the midline (Figures 11 to 13). Histopathology showed a malignant spindle cell neoplasm.

6. Discussion

LMS of the IVC is one of the rarest vascular tumours, constituting less than 1% of all soft tissue sarcomas and representing the most common primary malignancy of the IVC. The clinical course is typically insidious due to the tumour's retroperitoneal location and gradual onset, often resulting in delayed diagnosis. This case series, comprising three female patients, further underscores the heterogeneous clinical presentations, radiologic patterns, and complex diagnostic challenges associated with this rare entity. The first case involved a 45-year-old female who presented asymptomatically during follow-up evaluation for ureteric calculi. This case highlights a potentially incidental detection, which is particularly unusual given the generally asymptomatic nature of retroperitoneal LMSs until they reach considerable size. Imaging revealed a 6cm lobulated retroperitoneal mass with heterogeneous enhancement

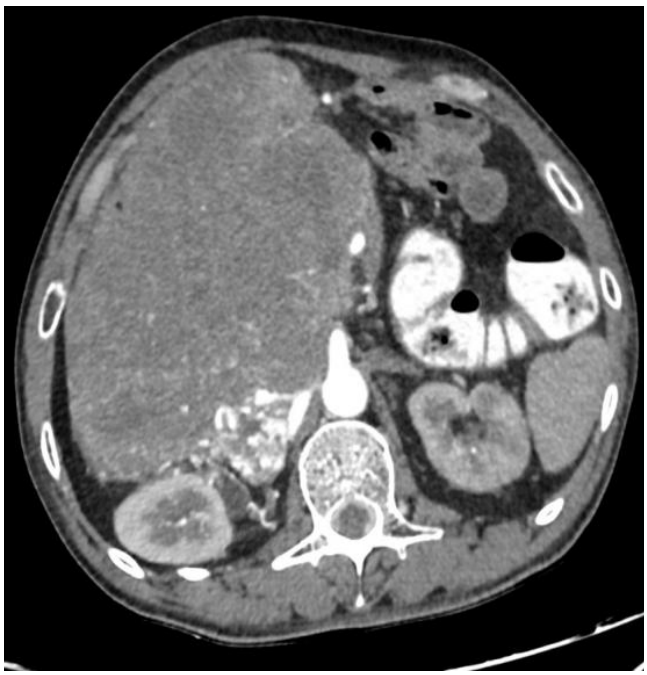

Figure 11. Arterial phase CECT shows a large heterogeneously enhancing mass lesion involving the right hypochondrium and pelvis with lobulated margins and areas of necrosis seen arising from the IVC with neoangiogenesis. The lesion crossed the midline medially.

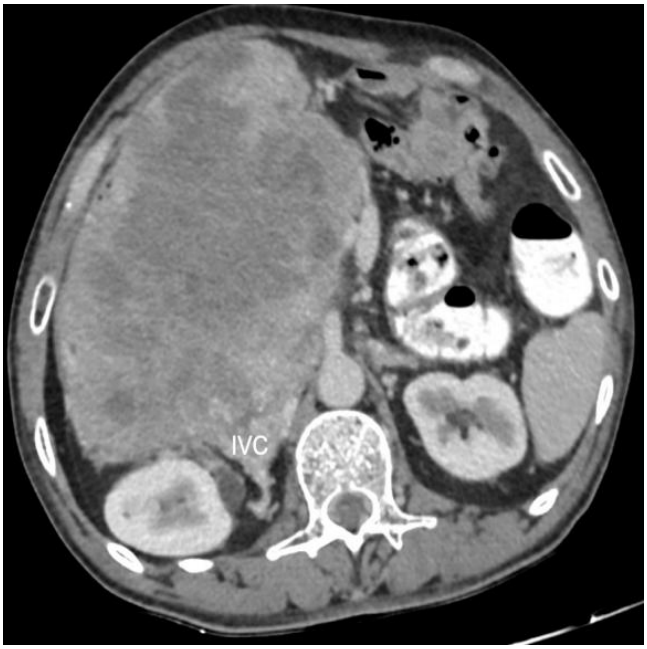

Figure 12. Venous phase CECT shows a large heterogenously enhancing mass lesion with lobulated margins and areas of necrosis seen arising from the IVC, involving the right hypochondrium and pelvis.

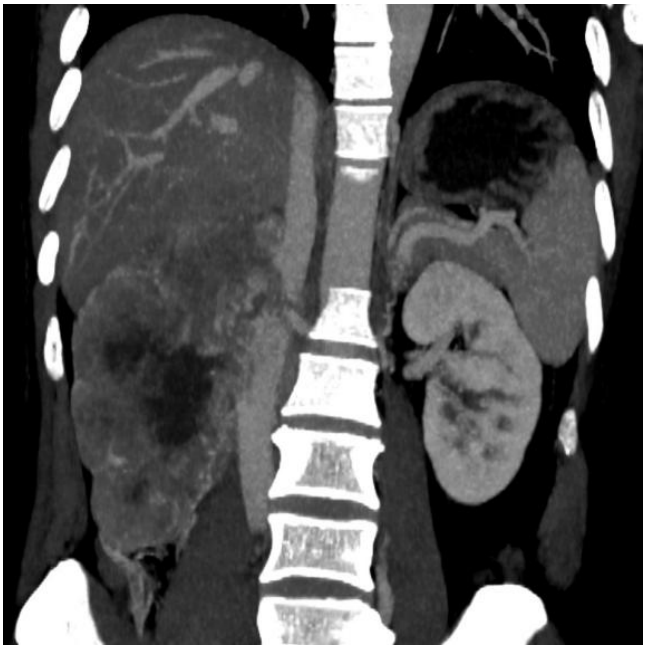

Figure 13. The Coronal MIP image shows a large heterogeneously enhancing mass lesion seen arising from the IVC with neoangiogenesis. Inferiorly, the lesion extends up to the right iliac fossa.

and an associated filling defect in the IVC, suggesting intraluminal involvement[5]. This finding aligns with descriptions in the literature, where early truncal LMSs may show partial venous obstruction without overt vascular or systemic signs (Mingoli *et al.*; Kieffer *et al*)[1,2].

Histopathologic confirmation showed spindle cell morphology organised in palisading fascicles with moderate encapsulation, characteristic of LMS[6.] Notably, the early-stage detection in this case potentially allowed for more favourable surgical excision and prognosis.

In comparison, the second case presented with more overt symptoms—right hypochondrial pain and unexplained weight loss, leading to advanced radiologic imaging. CECT showed a larger lesion measuring 9cm, significantly compromising the infrahepatic IVC with intraluminal and extraluminal extension as confirmed on MRI, which also demonstrated diffusion restriction—a typical signal property of high-grade sarcomas. The TOF image revealed eccentric residual flow, indicating near-total venous occlusion, a complication whose progression could explain the symptomatic presentation. This radiological pattern is in accordance with prior studies by Pawlik *et al.*, which

noted that segment II IVC tumours often produce nonspecific systemic symptoms and tend to be detected later.

The third case demonstrated the most advanced pathology, presenting with gross abdominal distension and palpable mass. Imaging revealed an extensive lobulated mass (measuring 22.5 × 16.1× 15.2cm) extending from the right hypochondrium to the pelvis, with displacement of adjacent bowel loops and superior extension up to the liver. This case illustrates classic features of large retroperitoneal sarcomas, including organ displacement, heterogeneous post-contrast enhancement, and areas of central necrosis, reflective of aggressive biology and tumour burden. The mass's sheer size would likely complicate surgical intervention and increase the risk of incomplete resection, a key negative prognostic factor identified in the literature (Kieffer *et al.*; Wachtel *et al.*)[2]. Across all three cases, several key points emerge:

Varied Presentation: While one tumour was discovered incidentally, the others presented with either localised pain or a conspicuous abdominal mass. This variability emphasises the importance of clinical suspicion, especially in middle-aged women presenting with vague abdominal symptoms or non-specific systemic complaints.

Imaging Role: Multiphase CT and MRI, particularly with contrast enhancement and diffusion-weighted sequences, offered critical diagnostic clues. Features such as heterogeneous enhancement, necrotic central zones, intraluminal extension, and adjacent structure displacement are considered highly suspicious for malignant retroperitoneal tumours with vascular involvement[7].

Growth Patterns: These cases highlight LMS's tendency for both extraluminal and intraluminal spread. The second and third tumours clearly demonstrate the complex growth pattern across vascular compartments, which increases surgical challenge and risk for vascular compromise.

Histologic Features: Though only one histopathological report was described in detail, the typical LMS morphology of palisading spindle cells within a fibrous capsule was evident. This morphology is consistent with the diagnostic standard for smooth muscle sarcoma and forms the basis for distinguishing LMS from mimickers like malignant peripheral nerve sheath tumours or Gastrointestinal Stromal Tumours (GIST).

7. Summary and Conclusion

Leiomyosarcoma of the IVC is an exceedingly rare but formidable malignancy requiring early recognition, advanced imaging, and aggressive surgical intervention. Although complete surgical resection offers the best curative potential, delayed diagnosis and high recurrence rates limit long-term outcomes. Continued reporting through case series, institutional registries, and multi-centre collaborations is vital to build evidence and potentially establish guidelines. Future research should aim to explore the molecular profile of these tumours to identify therapeutic targets and improve patient survival through personalised medicine.

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