Submission-Ready Case Report (Manuscript)

IMAGING OF THE PRIMARY IVC LEIOMYOSARCOMA AS A RARE ENTITY: A CASE REPORT ON DIAGNOSTIC FEATURES ON CONTRAST-ENHANCED MRI AND POST-OPERATIVE IMAGING

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Abstract

Primary leiomyosarcoma of the inferior vena cava (IVC-LMS) is an exceptionally rare vascular malignancy that often poses diagnostic and surgical challenges. We report the case of a 65-year-old woman who presented with a large infra-renal IVC mass. Contrast-enhanced MRI demonstrated a lobulated lesion with heterogeneous enhancement, central necrosis, and restricted diffusion in the solid components. The presence of an “IVC claw sign” confirmed its vascular origin. The mass closely abutted the liver, pancreas, and right kidney without evidence of invasion, and both renal veins were preserved. The patient underwent complete en bloc tumor excision with IVC reconstruction using a collagen-coated polyester (INTERGARD) graft following Cattell–Braasch exposure. Postoperative CT imaging confirmed total tumor removal and graft patency. This case underscores the crucial role of MRI in defining the vascular origin and extent of the tumor, facilitating precise surgical planning, and demonstrates the feasibility of successful IVC reconstruction using synthetic vascular grafts.

Keywords: Inferior vena cava (IVC) leiomyosarcoma; Contrast-enhanced MRI; IVC reconstruction; Vascular graft (INTERGARD); Retroperitoneal tumor; Rare vascular malignancy

Highlights

- Primary leiomyosarcoma of the IVC is a rare vascular malignancy.
- MRI demonstrates hallmark features including heterogeneous enhancement, central necrosis, restricted diffusion, and the "IVC claw sign."
- Complete en bloc resection with IVC reconstruction offers optimal local control.
- This case demonstrates successful graft reconstruction using a collagen-coated polyester (INTERGARD) graft.
- Postoperative CT confirmed complete excision and graft patency

Introduction

Leiomyosarcoma of the IVC arises from smooth muscle of the tunica media and accounts for <2% of retroperitoneal sarcomas [1]. Clinical manifestations are nonspecific, often related to mass effect. MRI is the imaging modality of choice for diagnosis, as it delineates vessel involvement, signal features, and resectability [2,3]. Complete surgical excision with caval reconstruction is the mainstay of treatment, with graft options including PTFE, Dacron, or collagen-coated polyester [4–6]. We present a case of infra-renal IVC leiomyosarcoma with detailed MRI characterization and surgical reconstruction, compared with prior literature.

Case Presentation

A 65-year-old woman presented with a ten-year history of intermittent upper abdominal pain. The pain was insidious, non-progressive, non-radiating, and relieved intermittently by analgesics. She also reported anorexia but denied weight loss, fever, jaundice, hematemesis, melena, or vomiting.

On examination, her condition was stable with no icterus or lymphadenopathy. Abdominal palpation revealed a firm, hard epigastric mass not moving with respiration.

PET-CT demonstrated a retroperitoneal mass measuring 12 × 10 × 16 cm, involving the second and third portions of the duodenum, the pancreatic head, and the right kidney. No metastases or nodal uptake were seen. Biopsy confirmed smooth muscle origin, consistent with leiomyosarcoma.

Imaging Findings

MRI demonstrated a large lobulated mass expanding the infra-renal segment of the inferior vena cava (IVC), exhibiting both intraluminal and extraluminal components. On T1-weighted images, the lesion appeared predominantly hypointense, while T2-weighted images showed heterogeneous hyperintensity with areas of central necrosis. Post-contrast T1-weighted sequences revealed heterogeneous enhancement with cystic and necrotic regions. Diffusion-weighted imaging (DWI) demonstrated restricted diffusion within the solid components, with no restriction in the necrotic areas. A characteristic "IVC claw sign" was noted, with an imperceptible vessel wall at the tumor origin, confirming its vascular source. The mass abutted the undersurface of the liver, pancreatic head, and

right kidney; however, intervening fat planes were preserved, and there was no evidence of direct invasion. Both renal veins were intact and uninvolved.

IMAGES:

Figure 1: Axial T1WI (A) and axial T2WI (B) showing signal characteristics, where the mass appears as low signal intensity on T1WI, consistent with its soft tissue nature and shows high signal intensity on T2WI, reflecting internal necrosis and fluid content (white arrows)

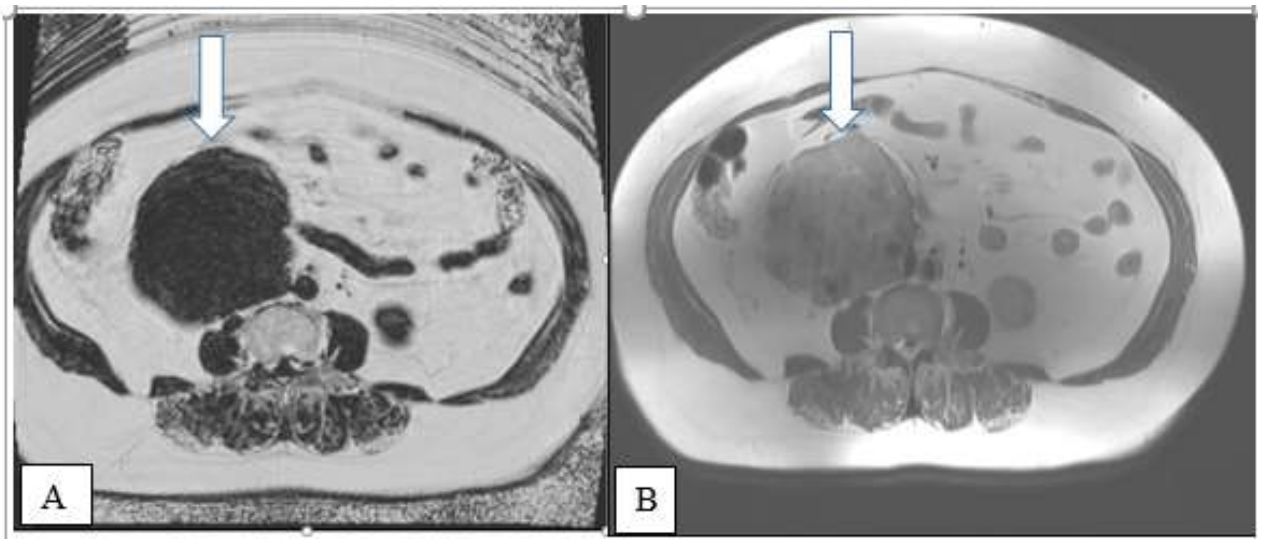


Figure 2: Post-contrast MRI axial (A), sagittal (B), and coronal (C) T1-weighted MRI showing heterogeneous enhancement with necrosis. The mass expands the IVC lumen, with both intraluminal and extraluminal components. Imperceptibility of the IVC wall at maximal tumor contact and the "IVC claw sign" support a vascular origin consistent with primary IVC leiomyosarcoma (white arrows

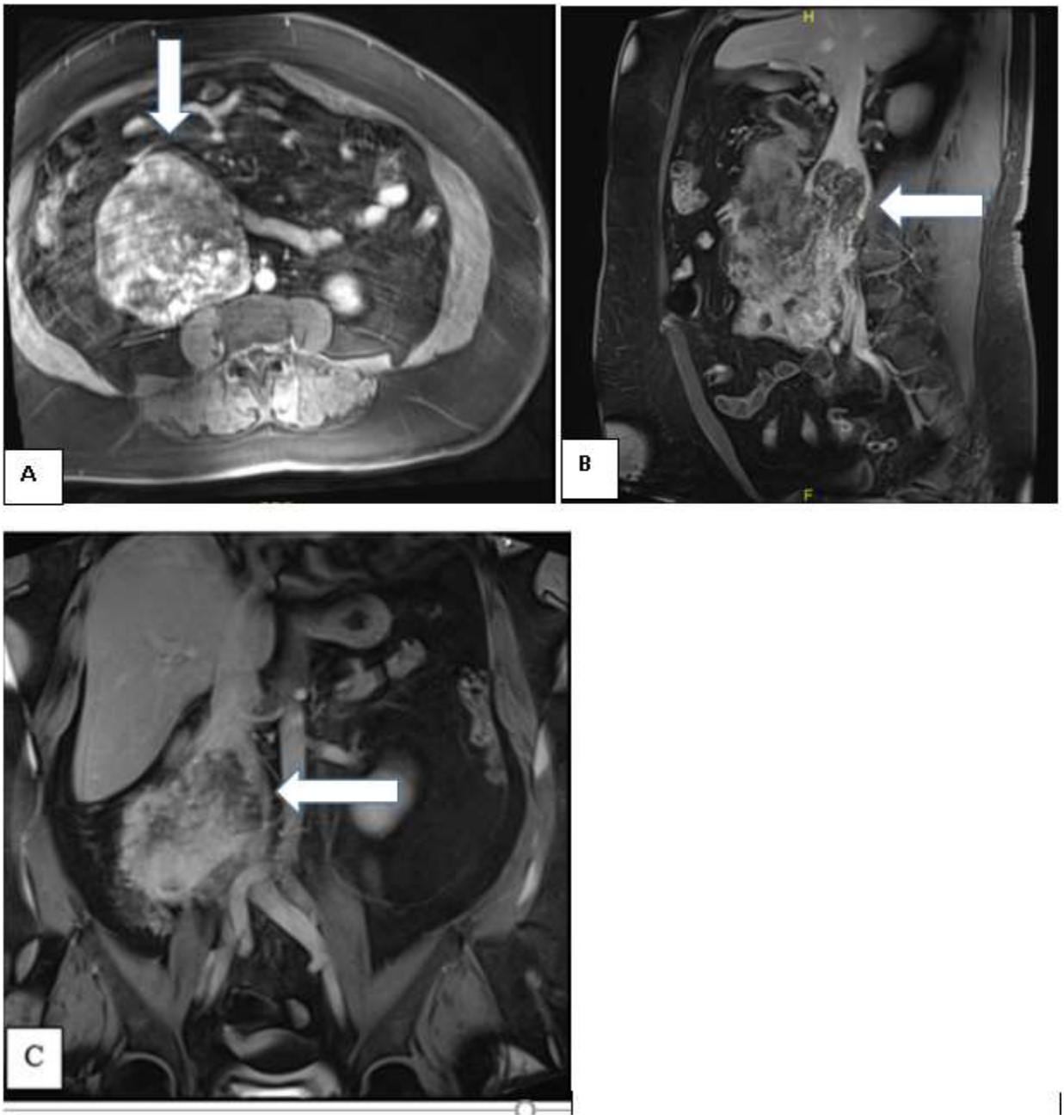


Figure 3: MRI axial ADC /DWI images showing restricted diffusion in solid tumor portions.

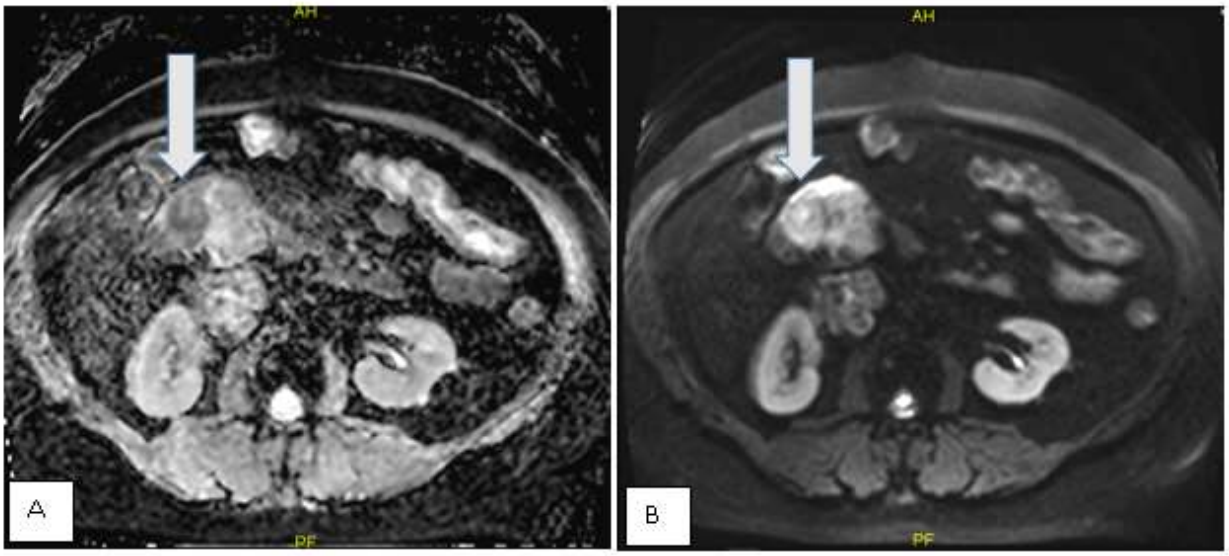
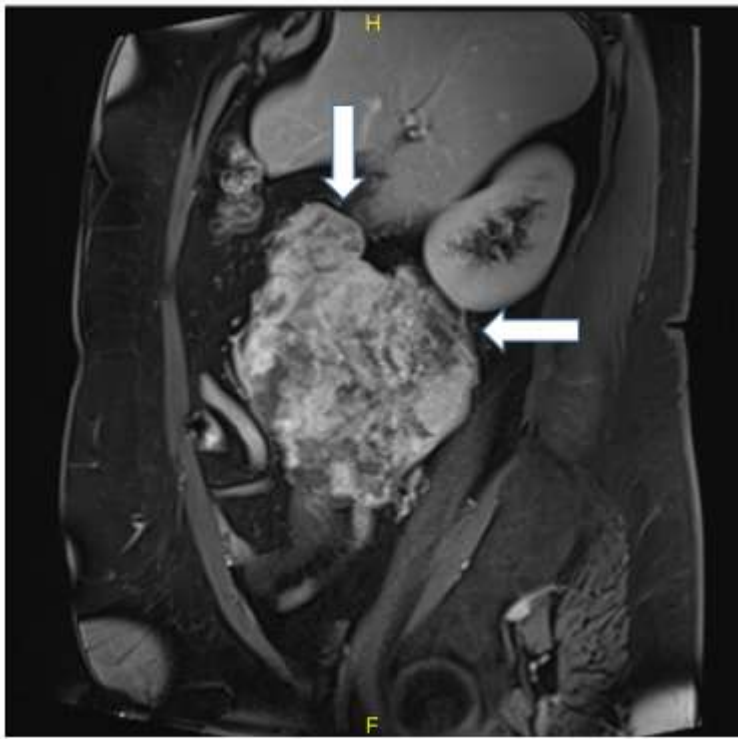


Figure 4: Showing tumor abutment with the inferior surface of the liver and the right kidney with reserved fat planes and intact organ morphology indicate no direct invasion (white arrows)



Operative Technique

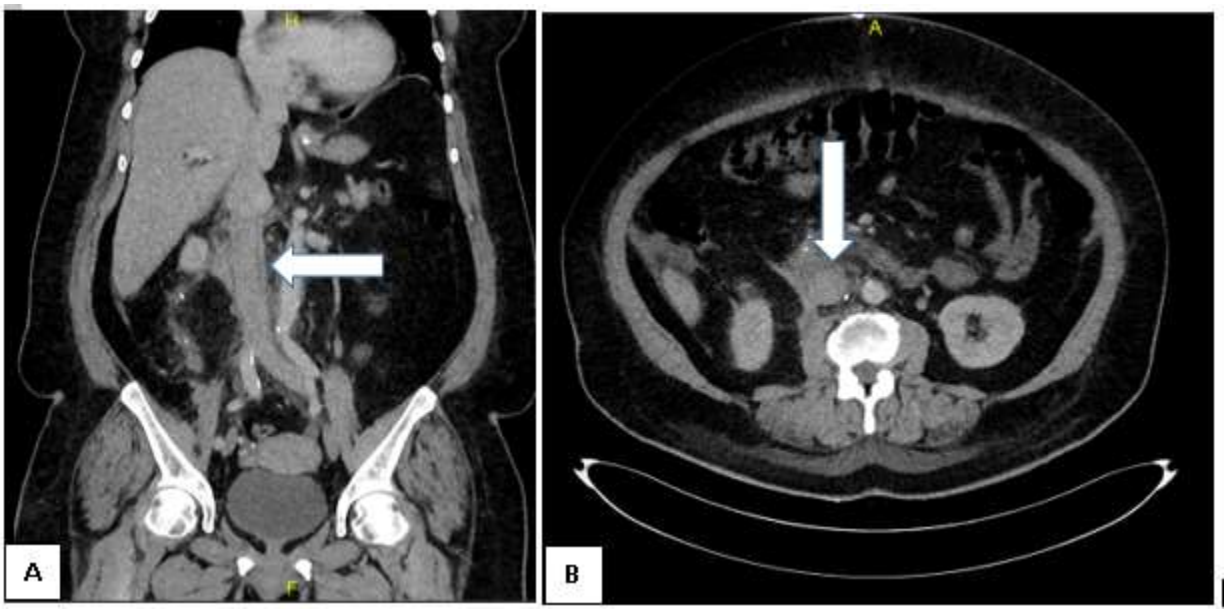
Through a midline laparotomy, a right medial visceral rotation (Cattell–Braasch maneuver) and extended Kocher were performed to expose the infra-renal IVC, duodenum (D2/D3), and right renal hilum. The tumor, arising from the anterior wall of the infra-renal IVC and densely adherent to the right ureter and duodenum, was mobilized. After systemic heparinization and vascular control (infra-renal and supra-iliac IVC clamping), the IVC segment was excised en bloc with the tumor (cross-clamp time: 40 minutes). The duodenal serosa was reinforced with interrupted 5-0 Prolene sutures.

Reconstruction was achieved with a collagen-coated polyester interposition graft (INTERGARD, 300 mm × 22 mm, ~15 cm used), sutured with continuous 4-0 Prolene. Hemostasis was secured and a pelvic drain placed. The postoperative course was uneventful.

This approach mirrors techniques reported in literature for IVC tumor excision with prosthetic graft reconstruction [4–6].

Postoperative plain CT scan images coronal and axial confirmed complete excision and successful IVC graft reconstruction.

Figure 5: Postoperative plain CT, coronal and axial images (A & B) - showing graft reconstruction and patency.



Discussion

Our case demonstrates classical MRI findings of IVC-LMS, which typically include low T1 and high T2 signal intensity, heterogeneous enhancement, central necrosis, and restricted diffusion in solid components [2,3]. The “IVC claw sign” and imperceptible vessel wall are critical imaging features that help differentiate primary vascular tumors from retroperitoneal masses. Such features, combined with the lesion’s intraluminal and extraluminal components, strongly suggest a vascular origin rather than secondary compression or invasion [1,2].

Primary leiomyosarcoma of the inferior vena cava (IVC) is an exceptionally rare malignant smooth muscle tumor, representing fewer than 2% of all soft tissue sarcomas and approximately 0.5% of malignancies affecting the venous system [1,4, 8]. It predominantly affects middle-aged to elderly women, with a predilection for the infra-renal and inter-renal portions of the vessel [1,8].

Clinical presentation of IVC-LMC varies depending on tumor location, ranging from nonspecific abdominal pain to lower limb edema or renal dysfunction due to venous obstruction [1,5, 9]. In our case, the tumor involved the infra-renal segment, which generally allows safer surgical resection and vascular reconstruction compared to more proximal lesions [4,6, 10].

Magnetic resonance imaging (MRI) serves as the imaging modality of choice, offering excellent soft-tissue contrast and multiplanar assessment. Typical findings include a lobulated mass with heterogeneous T2 hyperintensity, heterogeneous enhancement, central necrosis, and restricted diffusion in solid components [2,3]. Cine MRI may further evaluate adhesion to the vessel wall and assess dynamic flow characteristics [3]. MRI is particularly valuable in differentiating IVC-LMS from other retroperitoneal masses, such as adrenal cortical carcinoma, renal cell carcinoma, or paraganglioma, by demonstrating the continuity of the mass with the venous wall (“IVC claw sign”) [2, 9].

Complete **en bloc resection** with prosthetic graft reconstruction remains the treatment of choice, as incomplete excision is associated with high rates of local recurrence and poor survival [4–6, 8]. The Cattell–Braasch and extended Kocher maneuvers allow safe mobilization of the duodenum, pancreas, and ureters, enabling adequate vascular control [7]. Vascular graft options include PTFE, Dacron, and collagen-coated polyester; INTERGARD grafts, as used in our patient, offering superior durability, infection resistance, and long-term patency [5,6].

Postoperative complications can include graft thrombosis, bleeding, infection, and transient renal dysfunction, highlighting the need for meticulous surgical technique and vigilant postoperative monitoring [4,6,7]. Long-term prognosis depends on complete tumor excision and absence of metastasis; adjuvant therapies have limited proven benefit [1,5, 8, 10]. Postoperative imaging is essential to confirm graft patency and detect early recurrence, which typically occurs within 2–3 years [6, 8]. In our patient, postoperative CT confirmed complete tumor removal and patent graft, without early complications.

This case underscores the pivotal role of MRI in preoperative evaluation, providing critical information on tumor extent, vessel wall involvement, and surgical planning. Multidisciplinary coordination among radiology, vascular surgery, and oncology teams is essential for achieving optimal outcomes in patients with this rare but potentially curable vascular malignancy [1,4–6, 9, 10].

Conclusion

Primary IVC leiomyosarcoma is a rare but surgically treatable malignancy. MRI plays a pivotal role in diagnosis and operative planning by demonstrating vessel wall origin, enhancement pattern, and extent. Complete en bloc excision with IVC reconstruction, as in our case, offers the best chance for local control.

Declarations

- **Ethics Approval and Consent to Participate:** Not required; this report describes anonymized radiological data.
- **Consent for Publication:** Not applicable; no identifiable patient information is included.
- **Competing Interests:** The authors declare no competing interests.
- **Funding:** None.
- **Authors' Contributions:** Dr. Sylvia Bedas Nsato conceived the case, performed imaging analysis, conducted literature review, and drafted the manuscript. Supervisors and co-authors contributed to imaging interpretation, critical revision, and manuscript approval. All authors reviewed and approved the final manuscript.

Figure Legends

- **Figure 1:** Axial T1- and T2-weighted MRI showing low T1 and high T2 signal intensity.
- **Figure 2:** Post-contrast axial and coronal MRI demonstrating heterogeneous enhancement and central necrosis.
- **Figure 3:** DWI/ADC maps showing restricted diffusion in solid components.
- **Figure 4:** Post contrast T1W sagittal showing intact organ morphology the liver and the right kidney indicate no direct invasion.
- **Figure 5:** Postoperative CT confirming IVC graft reconstruction.

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