Case Report

# Intracardiac Extension of Intravenous Leiomyomatosis: Incremental Value of Multimodality Imaging and Intravascular Ultrasound

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## Abstract

Introduction: Intravenous leiomyomatosis (IVL) is a rare benign smooth muscle neoplasm characterized by progressive intravascular growth that may extend through the inferior vena cava (IVC) into the right heart. Despite its benign histology, intracardiac extension may cause life-threatening complications and often poses significant diagnostic and surgical challenges.

Case summary: A 37-year-old woman with a history of uterine leiomyomatosis treated with subtotal hysterectomy underwent routine transthoracic echocardiography, which unexpectedly revealed a mobile right atrial mass arising from the IVC. Multimodality imaging – including echocardiography, computed tomography (CT), cardiac magnetic resonance (CMR), positron emission tomography/computed tomography (PET/CT), and intravascular ultrasound (IVUS) – demonstrated continuous tumor extension from the right gonadal vein to the right atrium. IVUS provided unique intraluminal information by confirming the absence of vessel wall invasion and associated thrombus, thereby facilitating surgical planning. Following multidisciplinary Heart Team discussion, complete single-stage surgical resection was successfully performed. Histopathological examination confirmed IVL. At three-month follow-up, the patient remained asymptomatic with no evidence of recurrence.

Conclusion: This case illustrates the complementary role of multimodality imaging in IVL and highlights the incremental value of IVUS for defining tumor-vessel relationships and supporting preoperative decision-making.

## Introduction

Intravenous leiomyomatosis (IVL) is a rare benign smooth muscle neoplasm with an unusual biological behavior, characterized by continuous intraluminal growth through the venous system despite the absence of malignant histological features. Although most tumors remain confined to the pelvis, progressive extension through the inferior vena cava (IVC) into the right cardiac chambers may occur and is associated with potentially fatal cardiovascular complications.[1-7]

Because clinical manifestations are often nonspecific or absent, diagnosis relies heavily on imaging. Echocardiography, contrast-enhanced computed tomography (CT), and cardiac magnetic resonance (CMR) provide complementary information regarding tumor extent and tissue characterization, enabling accurate surgical planning.[7-9] However, information regarding the relationship between the tumor and the venous wall may remain limited when using conventional imaging alone.

Intravascular ultrasound (IVUS) has rarely been described in the evaluation of IVL but may offer direct intraluminal visualization of tumor morphology and vessel wall involvement.[9] We report a patient with extensive IVL extending from the right gonadal vein to the right atrium, highlighting the incremental value of IVUS within a multimodality imaging strategy for preoperative assessment and surgical planning.

## Case presentation

A 37-year-old woman with hypothyroidism and a previous history of symptomatic uterine leiomyomatosis treated with subtotal hysterectomy and bilateral salpingectomy was referred after an incidental finding on routine transthoracic echocardiography. She had no history of cardiovascular disease, venous thromboembolism, or malignancy and remained completely asymptomatic, reporting no dyspnea, chest pain, palpitations, or syncope. Physical examination revealed no signs of right-sided heart failure. The principal clinical events are summarised in Table 1.

Transthoracic echocardiography demonstrated a highly mobile, elongated mass arising from the IVC and extending into the right atrium, without significant obstruction of tricuspid inflow or evidence of right ventricular dysfunction (Figure 1). To further characterize the lesion, CMR identified a well-defined intracavitary mass with intermediate T1- and T2-weighted signal intensity and mild heterogeneous gadolinium enhancement, findings more suggestive of a neoplastic process than thrombus.

Contrast-enhanced thoracoabdominal CT demonstrated continuous tumor extension from the right gonadal vein through the IVC into the right atrium, measuring approximately 72 mm in length (Figure 2). Additional involvement of the right ovarian vein and iliac venous system was identified, without

## Keywords

Leiomyomatosis; Interventional Ultrasonography; Inferior Vena Cava

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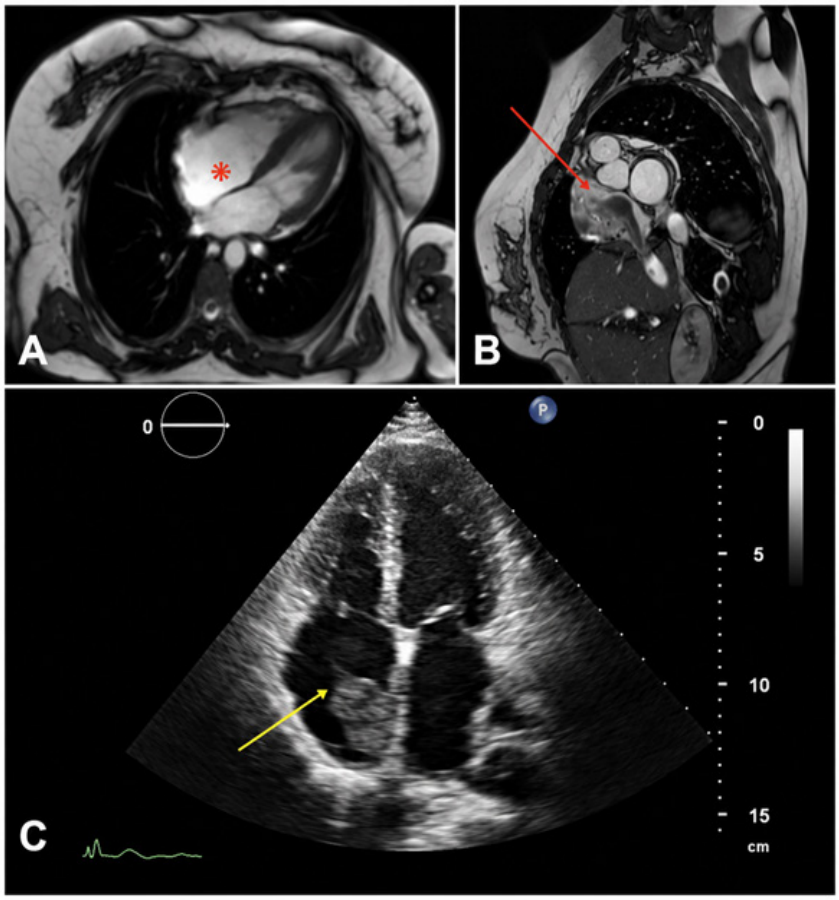



**Figure 1 –** *Initial multimodality imaging evaluation. (A) Axial cardiac magnetic resonance image showing an intracavitary right atrial mass (asterisk). (B) Coronal cardiac magnetic resonance image showing the mass extending towards the right atrium (arrow). (C) Transthoracic echocardiography, apical four-chamber view, demonstrating the right atrial mass (arrow). CMR: cardiac magnetic resonance.*

complete venous occlusion. Several small bilateral pulmonary nodules were also detected but showed no imaging features suggestive of metastatic disease.

To better define the relationship between the tumor and the venous wall, IVUS of the IVC was performed. IVUS demonstrated a homogeneous intraluminal lesion occupying approximately half of the caval lumen, with preservation of venous wall architecture and no evidence of mural invasion or associated thrombus (Figure 2). These findings provided complementary anatomical information beyond conventional imaging and supported the feasibility of complete surgical extraction.

Because primary venous leiomyosarcoma remained an important differential diagnosis, CT-guided biopsy and positron emission tomography/computed tomography (PET/CT) were performed. Histology demonstrated benign smooth muscle proliferation, while PET/CT showed only mild metabolic activity confined to the intravascular lesion, without evidence of distant disease, supporting the diagnosis of IVL.

The case was reviewed by a multidisciplinary Heart Team including cardiovascular surgeons, vascular surgeons, gynecologic oncologists, interventional cardiologists, radiologists, pathologists, and anesthesiologists. Complete single-stage surgical resection was deemed feasible and was successfully performed through combined abdominal and right mini-thoracotomy approaches under cardiopulmonary bypass (Figure 3). Histopathological examination confirmed IVL, demonstrating benign spindle-cell proliferation without significant atypia or tumor necrosis, along with positive immunohistochemical staining for smooth muscle markers and estrogen receptors.

The postoperative course was uneventful, and the patient was discharged on the sixth postoperative day. At three-month follow-up, she remained asymptomatic. Follow-up CT and transthoracic echocardiography confirmed complete tumor removal, preserved caval patency, and no evidence of local recurrence (Figure 4).

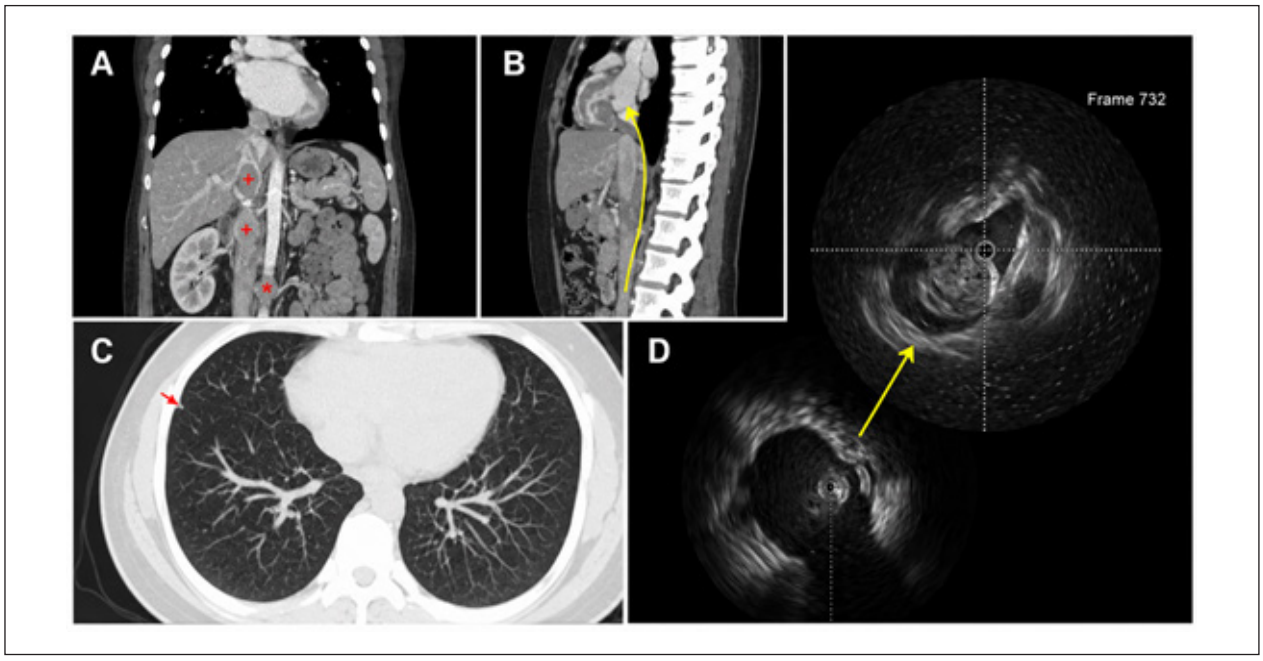



**Figure 2 –** *Computed tomography and intravascular ultrasound findings. (A) Coronal contrast-enhanced CT showing a filling defect in the inferior vena cava and involvement of the right gonadal vein. (B) Sagittal contrast-enhanced CT demonstrating longitudinal extension of the intravascular mass towards the right atrium (arrow). (C) Axial chest CT showing a subpleural pulmonary micronodule (arrow). (D) IVUS showing an intraluminal soft-tissue mass partially occupying the inferior vena cava lumen (arrow). CT: computed tomography; IVUS: intravascular ultrasound.*

## Discussion

IVL is a rare benign smooth muscle neoplasm with an unusual biological behavior, characterized by continuous intravascular growth despite the absence of malignant histological features. Although histologically benign, tumor extension through the IVC into the right cardiac chambers may result in life-threatening complications, including right ventricular inflow obstruction, pulmonary embolism, syncope, heart failure, and sudden death.[3,6,7] Early recognition is therefore essential, particularly in women with a current or previous history of uterine leiomyomas who present with an intracardiac mass.

This case highlights the pivotal role of multimodality imaging in establishing the diagnosis and guiding treatment. Transthoracic echocardiography identified the intracardiac component, while contrast-enhanced CT accurately delineated the cranio-caudal extent of the tumor throughout the venous system. CMR further supported a neoplastic rather than thrombotic origin through tissue characterization, and PET/CT helped exclude an aggressive malignant process. The integration of these complementary techniques enabled precise anatomical mapping before surgery and facilitated multidisciplinary decision-making.[7-10]

The principal novelty of this report is the incorporation of IVUS into the preoperative assessment. Reports describing the use of IVUS in IVL remain exceptionally scarce.[9] In our patient, IVUS provided direct intraluminal visualization of the lesion, confirming preservation of caval wall architecture, absence of mural invasion, and lack of associated thrombus. These findings complemented conventional imaging by more precisely defining the tumor–vessel interface, increasing diagnostic confidence, and supporting complete tumor extraction without extensive caval wall resection. Although IVUS should not replace cross-sectional imaging, our experience suggests that it may serve as a valuable adjunct in selected patients when the relationship between the tumor and the venous wall remains uncertain after CT or CMR.[9]

Complete surgical excision remains the treatment of choice and is the strongest determinant of long-term outcomes. Recent surgical series have shown that, in experienced centers, a single-stage multidisciplinary approach combining gynecologic, vascular, and cardiac surgery is both feasible and safe, minimizing the risk of residual disease and reducing the need for staged procedures.[10,11] Consistent with these reports, our patient underwent successful complete resection with an uncomplicated postoperative course and no evidence of recurrence during early follow-up.

## Conclusion

This case underscores three practical messages. First, IVL should be included in the differential diagnosis of right atrial masses in women with a history of uterine leiomyomas. Second, multimodality imaging is fundamental for defining disease extent and planning surgery. Finally, IVUS may provide unique intraluminal information that complements conventional imaging and may enhance preoperative decision-making in selected cases with intracardiac extension.

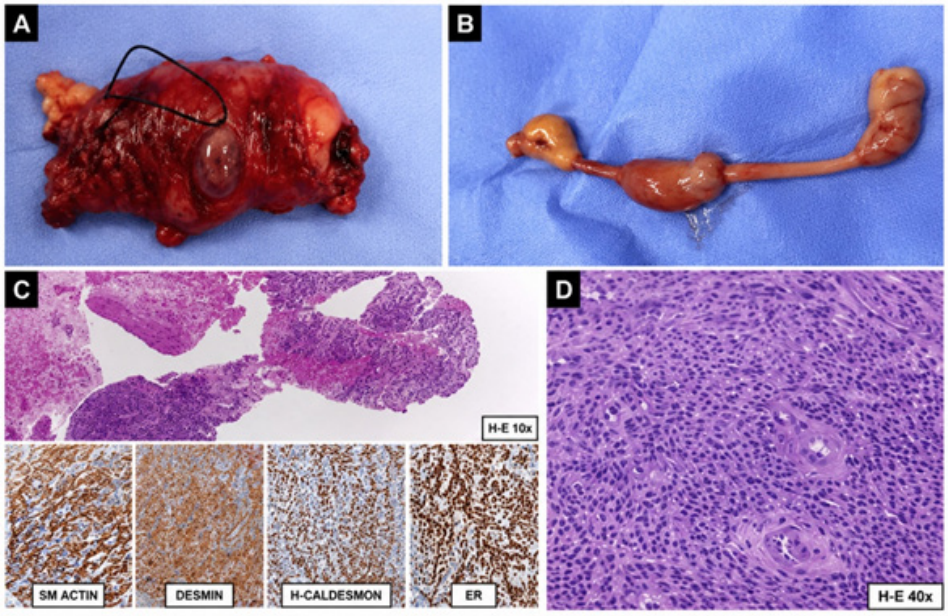



**Figure 3 –** *Surgical and pathological findings. (A) Surgical specimen obtained from the gonadal vessel and right ovary. (B) Elongated intravascular leiomyomatous specimen extending from the right gonadal vessel towards the right atrium. (C) Immunohistochemical staining, including oestrogen receptor, h-caldesmon, Ki-67, desmin, CD10 and smooth muscle actin, as labelled in the panels. (D) Haematoxylin and eosin sections at low and high magnification showing a spindle-cell mesenchymal neoplasm. Interpret the individual marker results according to the original pathology report.*

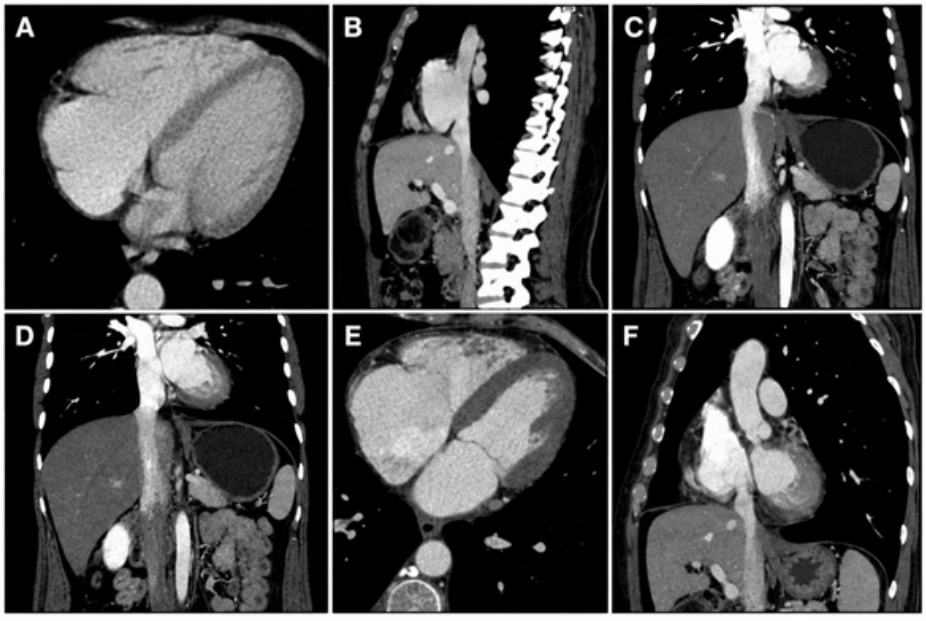



**Figure 4 –** *Postoperative computed tomography. (A) Axial image at cardiac level showing no residual right atrial mass. (B) Sagittal contrast-enhanced reconstruction showing the inferior vena cava without the previously visualised intraluminal mass. (C, D) Coronal contrast-enhanced reconstructions showing the postoperative caval anatomy and patent venous pathway. (E) Axial cardiac image confirming the absence of a residual right atrial mass. (F) Sagittal cardiac reconstruction demonstrating resolution of the previous cavoatrial extension. CT: computed tomography; IVC: inferior vena cava.*

**Table 1 – Clinical timeline**

| Time | Clinical event |
|---|---|
| Previous years | Symptomatic uterine leiomyomatosis treated by subtotal hysterectomy and bilateral salpingectomy |
| Presentation | Incidental right atrial mass detected on routine transthoracic echocardiography |
| Diagnostic work-up | CT, CMR, PET/CT and IVUS confirmed continuous intravascular tumor extension |
| Heart Team | Multidisciplinary discussion recommended complete single-stage resection |
| Surgery | Successful complete tumor excision |
| Histopathology | IVL confirmed |
| 3-month follow-up | No recurrence and complete clinical recovery |

### Learning points

IVL should be suspected in women with a history of uterine leiomyomas who present with a right atrial mass or continuous tumor extension through the IVC.

Multimodality imaging is essential for accurately defining tumor extent and planning complete surgical resection.

IVUS provides complementary intraluminal information regarding tumor–vessel wall relationships and may optimize surgical planning in selected patients.

### Ethics statement

Written informed consent for publication was obtained from the patient. According to institutional policy, formal Ethics Committee approval was not required because this manuscript describes a single case report without experimental intervention or identifiable patient information.

## Author Contributions

Conception and design of the research: Ocaranza-Sánchez R, Bigurra-Nachón EF, Montiel-Regino IG, Fernandez-Candelario BE, Barragan-Garate LR, Lozano-Fuantos F, Pérez-Alva JC; acquisition of data: Ocaranza-Sánchez R, Bigurra-Nachón EF, Betancourt-Villarruel JC, De la Fuente-Macip C; analysis and interpretation of the data: Ocaranza-Sánchez R, Barragan-Garate LR; statistical analysis: Ocaranza-Sánchez R, Montiel-Regino IG, Lozano-Fuantos F, Pérez-Alva JC; writing of the manuscript: Ocaranza-Sánchez R, Bigurra-Nachón EF, Fernandez-Candelario BE, Betancourt-Villarruel JC, Barragan-Garate LR, Lozano-Fuantos F, De la Fuente-Macip C; critical revision of the manuscript for intellectual content: Ocaranza-Sánchez R, Montiel-Regino IG, Fernandez-Candelario BE, De la Fuente-Macip C.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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### Study Association

This study is not associated with any thesis or dissertation work.

### Ethics Approval and Consent to Participate

All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.

### Use of Artificial Intelligence

During preparation of this manuscript, the authors used ChatGPT (OpenAI) to assist with language editing, translation and editorial formatting. The authors reviewed and edited the content and take full responsibility for its accuracy and integrity.

### Availability of Research Data

The underlying content of the research text is contained within the manuscript.

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