

Inferior Vena Cava Mesenchymal Tumor with Intracardiac Extension Mimicking an Atrial Myxoma in a Young Adult: A Case Report

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Abstract: Mesenchymal tumors originating from the inferior vena cava with cranial extension into the right atrium are rare and may closely mimic primary cardiac tumors, particularly atrial myxoma, leading to diagnostic and surgical pitfalls. A 29-year-old previously healthy man was referred for cardiac surgery after a large right atrial mass was detected by transthoracic echocardiography and interpreted as an atrial myxoma. No preoperative cross-sectional imaging was performed. Intraoperatively, the lesion was identified as cranial extension of a tumor arising from the inferior vena cava, prompting partial intracardiac resection. Subsequent magnetic resonance imaging demonstrated a large infiltrative right retroperitoneal mass with extensive inferior vena cava tumor thrombosis. The patient underwent definitive staged multidisciplinary surgery including retroperitoneal tumor resection, inferior vena cava tumor thrombectomy, and caval reconstruction with a vascular graft. Histopathology demonstrated a smooth muscle neoplasm with cellular atypia, diffuse smooth muscle marker expression, Ki-67 labeling index of approximately 15%, and p53 positivity, without tumor necrosis or mitotic activity, supporting classification as a tumor of uncertain malignant potential. This case highlights the critical importance of systematic inferior vena cava assessment in patients presenting with right atrial masses, as misdiagnosis may lead to inadequate surgical planning and increased perioperative risk. It also illustrates the diagnostic complexity of vascular smooth muscle tumors with atypical features and underscores the need for complete surgical resection, long-term anticoagulation with rivaroxaban after caval reconstruction, and structured oncologic surveillance in tumors of uncertain malignant potential.

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1. Introduction

Mesenchymal tumors involving the inferior vena cava (IVC) are rare entities and pose significant diagnostic and therapeutic challenges [3,5,6]. When these tumors extend cranially into the right atrium, they may closely resemble primary cardiac tumors, particularly atrial myxomas [2,4]. Misinterpretation of the tumor origin can lead to inadequate surgical planning and increased perioperative risk. We report a rare case of an IVC-origin mesenchymal tumor with intracardiac extension initially diagnosed as atrial myxoma, managed through a multidisciplinary surgical approach.

2. Case Report

2.1 Patient Information

A 29-year-old male patient with no relevant past medical history was admitted for elective cardiac surgery after detection of an intracardiac mass on transthoracic echocardiography (Figure 1), initially interpreted as a primary atrial myxoma [2,4]. At that time, no preoperative cross-sectional imaging was performed, given the presumed diagnosis of a primary cardiac tumor.

Figure 1. Transthoracic echocardiography showing a large intracardiac mass occupying the right atrium, initially interpreted as an atrial myxoma.



2.2 Clinical Findings

During the initial cardiac surgery, the intracardiac lesion was identified as cranial extension of a tumor originating from the inferior vena cava rather than a primary cardiac tumor. Partial resection of the intracardiac component was performed up to the level just proximal to the renal veins. Cardiopulmonary bypass time was 62 minutes, including 8 minutes of total circulatory arrest.

2.3 Timeline

In May 2025, an intracardiac mass was detected on transthoracic echocardiography and was initially diagnosed as an atrial myxoma. On May 9, 2025, the patient underwent partial resection of the intracardiac mass during cardiac surgery, and histopathological analysis revealed a spindle epithelioid mesenchymal neoplasm with atypia. Subsequently, on May 12, 2025, abdominal magnetic resonance imaging demonstrated a large right pericaval retroperitoneal mass associated with extensive tumor thrombosis of the inferior vena cava. On December 5, 2025, definitive oncologic surgery was performed, including resection of the retroperitoneal tumor, tumor thrombectomy of the inferior vena cava, and vascular reconstruction. The postoperative course was favorable, with no major complications.

2.4 Diagnostic assessment

Abdominal and pelvic magnetic resonance imaging demonstrated a large infiltrative right pericaval retroperitoneal mass measuring approximately 18.9 × 10.3 × 10.2 cm, with heterogeneous contrast enhancement and diffusion restriction, associated with extensive tumor thrombosis involving the infrarenal, juxtarenal, and intrahepatic segments of the inferior vena cava [3,5,6]. Thoracic imaging revealed postoperative changes and a small pericardial effusion, without mediastinal lymphadenopathy or distant metastases.

Histopathological examination of the intracardiac and retroperitoneal tumor specimens revealed a spindle and epithelioid mesenchymal neoplasm with cellular atypia, expansile growth pattern, and absence of mitotic figures in 50 high-power fields, without coagulative tumor necrosis. Immunohistochemical analysis demonstrated diffuse positivity for smooth muscle markers, including desmin, h-caldesmon, calponin, and HHF35, confirming smooth muscle differentiation. The tumor was negative for epithelial (AE1/AE3), endothelial (CD34), neural (S100), melanocytic (HMB45, Melan-A), and skeletal muscle (myogenin) markers. The Ki-67 labeling index was approximately 15%, and p53 expression was positive. These histopathological and immunophenotypic findings are summarized in Table 1 and support the diagnosis of a smooth muscle neoplasm of uncertain malignant potential according to current World Health Organization classification criteria [7,8].

Table 1. Histopathological and immunohistochemical profile of the inferior vena cava smooth muscle tumor.

Parameter	Result	Interpretation
Tumor morphology	Spindle and epithelioid cells	Mesenchymal neoplasm
Growth pattern	Expansile growth with areas of infiltration	Locally aggressive
Cellular atypia	Present	Supports atypical smooth muscle tumor
Mitotic activity	Absent (0 / 50 HPF)	Against high-grade sarcoma criteria
Tumor necrosis	Absent	Does not fulfill criteria for leiomyosarcoma
Desmin	Diffuse positive	Smooth muscle differentiation
H-caldesmon	Diffuse positive	Smooth muscle differentiation
Calponin	Diffuse positive	Smooth muscle differentiation
HHF35	Positive	Smooth muscle differentiation
AE1/AE3	Negative	Excludes epithelial tumor
CD34	Negative	Excludes endothelial tumor
S100	Negative	Excludes neural tumor
HMB45	Negative	Excludes PEComa
Melan-A	Negative	Excludes melanocytic tumor
Myogenin	Negative	Excludes skeletal muscle tumor
Ki-67 labeling index	Approximately 15%	Intermediate proliferative index
Aberrant p53 expression	Positive	Genomic instability

2.5 Therapeutic intervention

The patient underwent staged surgical management. Initial cardiac surgery addressed the intracardiac component of the tumor. Definitive surgery included extensive retroperitoneal tumor resection, IVC tumor thrombectomy, IVC reconstruction with graft, lymphadenectomy, preservation of renal structures, intracardiac tumor biopsy, and pericardial drainage, in accordance with recommended strategies for complex caval tumors [3,5]. Broad-spectrum antibiotics, optimized analgesia, and venous thromboembolism

prophylaxis were provided postoperatively. The staged surgical strategy was adopted due to the unexpected intraoperative finding of inferior vena cava tumor extension during cardiac surgery and the need for careful multidisciplinary planning to safely address the extensive retroperitoneal and vascular involvement. Although an en bloc resection was intended, histologic margin assessment was limited by specimen fragmentation inherent to caval thrombectomy. Therefore, microscopic R0 status could not be definitively confirmed, reinforcing the need for close radiologic surveillance.

2.6 Follow-up and outcomes

The patient had an uneventful immediate postoperative recovery and was discharged on postoperative day 9 in good clinical condition. Long-term anticoagulation was initiated with rivaroxaban 20 mg once daily. Direct oral anticoagulants have been increasingly used after caval reconstruction, with patency and safety comparable to vitamin K antagonists, combined with mechanical thromboprophylaxis, in accordance with published recommendations for inferior vena cava reconstruction with prosthetic grafts. At 2 months of follow-up, the patient remains asymptomatic, with preserved functional capacity and no clinical signs of venous outflow obstruction. Contrast-enhanced computed tomography demonstrated a patent inferior vena cava graft, without evidence of thrombosis or local recurrence. No distant metastases were detected. The patient continues under a structured oncologic surveillance program, with imaging scheduled every 6 months during the first two years.

3. Discussion and Conclusion

Tumors originating from the inferior vena cava with cranial extension into the right atrium represent one of the most challenging entities in vascular and cardiac oncology, both diagnostically and therapeutically. Because their intracardiac component may mimic primary cardiac neoplasms, particularly atrial myxomas, misdiagnosis at presentation is a well-recognized pitfall with potentially serious surgical implications [1-3]. This case illustrates not only the rarity of this condition but also the importance of comprehensive preoperative imaging and multidisciplinary planning in patients presenting with right atrial masses.

In contemporary clinical practice, a right atrial mass that is large, non-pedunculated, or not clearly attached to the interatrial septum should prompt systematic evaluation of the inferior vena cava by cross-sectional imaging, preferably contrast-enhanced computed tomography or magnetic resonance imaging [3,4]. Failure to assess the inferior vena cava may result in underestimation of tumor extent and inappropriate surgical strategy. In the present case, the initial diagnosis of atrial myxoma based on transthoracic echocardiography led to omission of cross-sectional imaging, resulting in an incomplete understanding of the tumor's origin and extent prior to cardiac surgery. This exposed the patient to an unplanned circulatory arrest while a large caval tumor remained in situ. Although no adverse events occurred, this diagnostic limitation significantly impacted surgical planning. Importantly, this case prompted a revision of our institutional diagnostic protocol for right atrial masses, mandating routine cross-sectional evaluation of the inferior vena cava.

The unexpected intraoperative discovery of tumor extension from the inferior vena cava necessitated a staged surgical strategy. In cases where the full extent of disease is known preoperatively, a single-stage combined cardiothoracic and vascular resection is often preferred, as it minimizes repeated anesthesia, tumor manipulation, and delays in definitive oncologic treatment [3,5,6]. However, in the absence of preoperative abdominal staging and given the intraoperative diagnostic surprise, initial intracardiac debulking followed by a carefully planned second-stage oncologic resection was considered the safest and most appropriate strategy.

From a pathological standpoint, classification of this tumor was particularly challenging. The neoplasm showed smooth muscle differentiation, with diffuse positivity for desmin, h-caldesmon, calponin, and HHF35, confirming a leiomyogenic lineage [7]. The presence of cellular atypia, a moderately elevated Ki-67 labeling index (approximately 15%), and p53 positivity raised concern for malignant potential. In published series of inferior vena cava smooth muscle tumors and leiomyosarcomas, elevated Ki-67 and p53 expression have been associated with higher recurrence rates and worse outcomes, emphasizing the biological overlap between atypical leiomyogenic tumors and low-grade leiomyosarcoma [5,6]. However, the key histologic criteria required for a diagnosis of leiomyosarcoma—namely, coagulative tumor necrosis and mitotic activity—were absent, with no mitoses identified in 50 high-power fields and no necrosis observed. According to the World Health Organization classification of soft tissue tumors, these findings support a diagnosis of smooth muscle neoplasm of uncertain malignant potential rather than definitive leiomyosarcoma [7].

Nevertheless, the tumor's large size (18.9 cm), infiltrative growth pattern, and proliferative index indicate biologically aggressive behavior that overlaps with low-grade leiomyosarcoma. In vascular smooth muscle tumors, the term "uncertain malignant potential" reflects the recognized dissociation between histologic grade and clinical behavior, whereby tumors lacking classic malignant criteria may still recur or metastasize [7,8]. Therefore, despite the absence of overt histologic malignancy, this patient must be considered at significant oncologic risk.

Complete surgical resection with negative margins remains the most important prognostic factor in inferior vena cava tumors [3,5,6]. In this case, the retroperitoneal tumor and caval tumor thrombus were resected en bloc with reconstruction of the inferior vena cava using a vascular graft, aiming to achieve oncologic clearance and restore venous outflow. Reconstruction of the inferior vena cava introduces important long-term considerations, particularly the risk of graft thrombosis. Patients with caval grafts often require prolonged or lifelong anticoagulation to maintain graft patency and prevent thromboembolic complications [3,5]. In the present case, the patient was discharged on rivaroxaban 20 mg once daily, adjusted according to current literature and institutional practice for venous graft thromboprophylaxis, in combination with mechanical measures including graduated compression stockings and early ambulation. This strategy aimed to minimize the risk of graft thrombosis while balancing bleeding risk in the setting of extensive oncologic and vascular surgery.

Given the tumor's uncertain malignant potential and its vascular origin, structured oncologic surveillance is mandatory. Long-term follow-up should include contrast-enhanced computed tomography or magnetic resonance imaging of the chest, abdomen, and pelvis every 3 to 6 months during the first two years, followed by annual imaging thereafter, to detect local recurrence, pulmonary metastases, and assess patency of the reconstructed inferior vena cava [5,6]. As no validated serum biomarkers exist for leiomyogenic tumors, radiological and clinical follow-up remains the cornerstone of postoperative surveillance.

In conclusion, this case underscores the importance of maintaining a high index of suspicion for inferior vena cava tumors in patients with right atrial masses, the necessity of comprehensive preoperative imaging, and the value of multidisciplinary surgical management. It also highlights the complexity of pathological classification in large vascular smooth muscle tumors and the need for vigilant long-term surveillance in tumors of uncertain malignant potential. This case prompted a revision of our institutional diagnostic protocol for right atrial masses, mandating cross-sectional imaging of the inferior vena cava to avoid similar diagnostic pitfalls in future patients.

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