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Case Report

# Right Atrial Myxoma Masquerading as Superior Vena Cava Syndrome: A Case Report and Review of the Literature

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**Abstract:**

**Background:** Right atrial myxoma is a rare but clinically significant primary cardiac tumor that can mimic diverse cardiopulmonary conditions, including superior vena cava (SVC) syndrome. Its atypical presentation, particularly when combined with tricuspid valve obstruction, poses a diagnostic and therapeutic challenge.

**Case Summary:** We report the case of a 65-year-old diabetic woman who presented with a one-year history of progressive exertional dyspnea (NYHA class III–IV), alongside the clinical hallmarks of SVC syndrome: jugular venous distension, cape-like edema, bilateral supraclavicular fullness, and prominent thoracic collateral venous circulation. Multimodal imaging — including contrast-enhanced thoracic CT and transthoracic echocardiography — identified a large, homogenous, mobile mass (67 × 37 mm on CT; 60 × 32 mm on echocardiography) originating from the interatrial septum and prolapsing across the tricuspid valve, generating a significant transvalvular gradient of 13 mmHg. The patient underwent emergency surgical resection with complete tumor excision, tricuspid annuloplasty (DeVega technique), and thrombectomy. Histopathological examination confirmed the diagnosis of cardiac myxoma. The postoperative course was uneventful.

**Discussion and conclusion:** This case illustrates the exceptional presentation of a right atrial myxoma revealed by SVC syndrome, a rare and diagnostically misleading condition often suggestive of malignancy. It highlights the critical role of integrated imaging in differentiating benign from malignant intracardiac masses and guiding prompt decision-making. The association with severe tricuspid obstruction and intracardiac thrombus further underscores its clinical complexity. Early surgical management was decisive, preventing potentially fatal hemodynamic and embolic complications.

**Key words:** right atrial myxoma; superior vena cava syndrome; cardiac tumor; tricuspid obstruction; surgical excision

## 1.Introduction

Cardiac myxomas are the most common primary cardiac tumors, accounting for approximately 30–50% of all primary cardiac neoplasms, with an estimated incidence of 0.5 to 1 case per million individuals annually [1, 2]. They are biologically benign and histologically originate from multipotent mesenchymal cells, typically forming gelatinous, pedunculated masses attached to the endocardium via a fibrovascular stalk. Despite their benign histology, myxomas have been aptly described as "functionally malignant" given their propensity to cause hemodynamic obstruction, systemic embolization, and sudden death [3].

The overwhelming majority of cardiac myxomas arise in the left atrium — over 75–85% of cases in large series — where they classically obstruct the mitral valve and simulate rheumatic mitral stenosis [4, 5]. Right atrial myxoma (RAM) constitutes only 12–18% of all myxoma cases, and its clinical presentation is considerably more variable and less well-characterized [6]. Rather than mimicking mitral disease, RAM tends to obstruct the tricuspid valve or the right ventricular outflow tract, leading to features of right heart failure including peripheral edema, ascites, and hepatic congestion. However, in rare and particularly dramatic instances, a right atrial mass may grow large enough — or assume a position that compresses the superior vena cava (SVC) — to generate a full clinical picture of SVC syndrome [7, 8].

Superior vena cava syndrome is a well-recognized clinical entity defined by obstruction of blood flow through the SVC, resulting in venous hypertension of the upper body. Its classic manifestations — facial and upper extremity swelling, dilated cervical and thoracic veins, dyspnea, and plethora — are most often attributed to mediastinal malignancies, intravascular thrombosis, or fibrosing mediastinitis [9]. The recognition of an intracardiac benign tumor as the underlying culprit is rare and may delay diagnosis, with potentially life-threatening consequences.

We herein report a case of a giant right atrial myxoma presenting as SVC syndrome with concomitant tricuspid obstruction in a 65-year-old diabetic woman, successfully treated by emergency surgical excision. We review the pertinent literature and discuss the epidemiology, clinical features, diagnostic approach, and surgical management of this uncommon but compelling presentation.

## 2. Case Report

### 2.1 Clinical Presentation

A 65-year-old woman with a background of type 2 diabetes mellitus and no other notable medical history was referred to our cardiology department with a one-year history of progressive exertional dyspnea. Over the preceding month, her functional capacity had deteriorated sharply, from NYHA class III to class IV, severely limiting her activities of daily living. She also reported significant general deterioration. No history of fever, joint pain, prior cardiac disease, or familial tumor syndromes was elicited.

On examination, the patient was hemodynamically stable with an oxygen saturation of 96% on room air. The clinical picture was dominated by the striking features of SVC syndrome: prominent jugular venous distension, bilateral supraclavicular fullness, cape-like non-pitting edema of the neck and upper chest, and a well-developed network of dilated thoracic collateral veins. Cardiac auscultation was remarkable. There was no peripheral cyanosis or digital clubbing. The abdomen was soft without organomegaly. No peripheral arterial embolic stigmata were detected.

### 2.2 Investigations

The 12-lead electrocardiogram demonstrated generalized low voltage, suggestive of pericardial effusion or right heart compression, with no evidence of arrhythmia or conduction defect.

Contrast-enhanced thoracic computed tomography (CT) revealed a voluminous hypodense intracardiac mass occupying the right ventricle (RV) and right atrium (RA), measuring 67 × 37 mm. Critically, the lesion showed no contrast enhancement after intravenous injection of iodinated contrast material — a feature that, is not typically characteristic of myxomatous tissue suggesting more likely a thrombus. The scan additionally identified bilateral fissural pleural effusions and a moderate pericardial effusion. No direct or indirect signs of pulmonary embolism were detected, and the pulmonary parenchyma appeared otherwise normal.

Transthoracic echocardiography (TTE) was the pivotal diagnostic investigation. It demonstrated a large, homogeneous, echogenic, and highly mobile mass measuring 60 × 32 mm, originating from the interatrial septum and prolapsing through the tricuspid orifice into the right ventricle during diastole (Figure 1). A significant tricuspid obstruction was documented with a mean transvalvular gradient of 13 mmHg. Secondary right-sided remodeling was evident, with functional dilation of the tricuspid annulus and the basal right ventricle and right atrium. A moderate pericardial effusion was also confirmed. Notably, left cardiac chambers appeared unaffected.

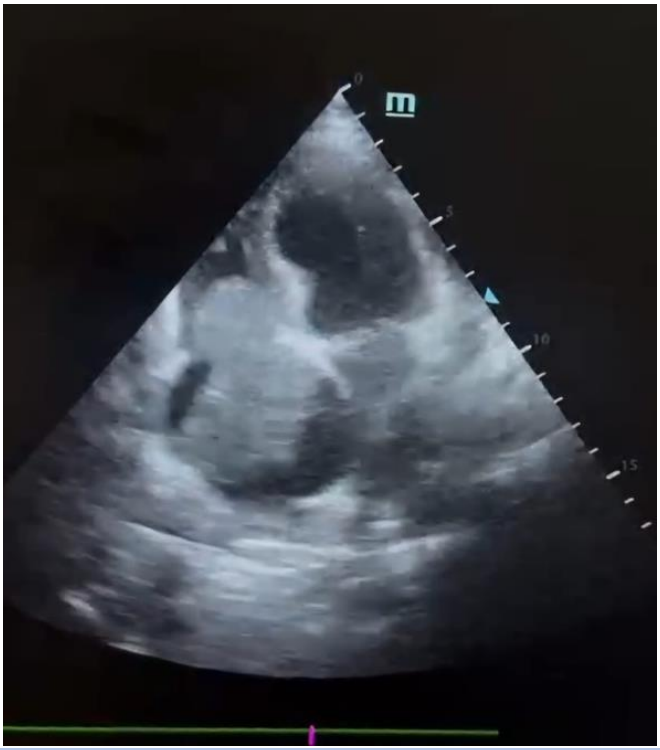

**Figure 1:** four chamber view showing a giant myxoma presenting as an obstructive mass protruding through the tricuspid valve

Abdominal ultrasound was unremarkable with no evidence of a primary abdominal tumor, which, combined with the echocardiographic morphology and attachment pattern, strongly supported a primary cardiac origin of the mass. The complete blood count and other hematologic and biochemical parameters were within normal limits — notably, no anemia, leukocytosis, thrombocytopenia, or elevated inflammatory markers were documented, distinguishing this case from the classic "constitutional triad" of myxoma (fever, weight loss, elevated ESR) that is more frequently associated with left atrial forms.

### 2.3 Surgical management and histopathology

Given the severity of the hemodynamic compromise and the risk of tumor fragmentation and pulmonary embolization, the patient was taken to the operating room without delay. Surgical exploration was performed under cardiopulmonary bypass via median sternotomy. Intraoperative findings confirmed a well-circumscribed tumor mass arising from the septal wall of the right atrium, accompanied by a voluminous associated thrombus — a not uncommon finding given the turbulent flow dynamics generated by the mass. Importantly, the tricuspid valve leaflets showed no rheumatic or infiltrative changes, confirming that the observed valvular dysfunction was entirely due to the mass effect of the prolapsing tumor.

Complete resection of the mass was achieved with wide surgical margins to minimize the risk of recurrence. Thrombectomy was performed to clear the associated thrombus. The tricuspid annular dilatation was corrected with a DeVega annuloplasty. The postoperative course was uneventful, with rapid resolution of the SVC syndrome signs and progressive improvement in dyspnea.

Histopathological examination of the resected specimen confirmed the diagnosis of cardiac myxoma, demonstrating the characteristic loose myxoid stroma with scattered stellate and polygonal cells embedded within an abundant mucopolysaccharide matrix — the pathological hallmark of this entity.

## 3. Discussion

### 3.1 Epidemiology and the right atrial paradox

Cardiac myxomas affect women more frequently than men, with a female predominance of approximately 64–65% across large series [4, 5]. Most patients are diagnosed in the fifth decade of life, although the condition has been reported across all age groups. The sporadic form, which comprises the vast majority of cases, typically presents as a solitary lesion in the left atrium, attached to the fossa ovalis region. Right atrial involvement, as illustrated in our case, is found in approximately 12–18% of patients, and constitutes a distinct clinical subset with its own characteristic features and pitfalls [6, 10].

A systematic review covering 619 patients with right atrial myxoma reported that the mean patient age at diagnosis was 45.7 years — notably younger than in our case — with a slight female predominance [10]. The most common clinical manifestations were cardiac symptoms (77%), followed by systemic (34.8%) and neurologic (21.1%) presentations. The lower rate of embolic events in right-sided compared to left-sided myxomas (3.57% vs. 30.09%) has been consistently documented across studies [6], likely because the pulmonary circulation is more tolerant of small tumor emboli than the systemic arterial tree.

### 3.2 SVC syndrome as a presentation of right atrial myxoma: A rare diagnostic trap

SVC syndrome arising from a right atrial myxoma is an uncommon and potentially treacherous presentation. The SVC obstruction in these cases results from one or more distinct mechanisms: direct mechanical compression of the SVC orifice by a bulky atrial mass, tumor invasion into the SVC itself, or — as illustrated by a fascinating case described by Zhang et al., — a myxoma whose pedicle arises directly from the anterior wall of the SVC rather than from the right atrial endocardium [9]. In our patient, the mass size (67 mm maximal diameter on CT) and its septal origin likely created sufficient bulk to impair SVC inflow at the right atrial junction.

The case reported by Longatto et al., from Brazil [7] bears close parallels to ours: a large right intra-atrial lesion producing SVC syndrome, successfully managed with emergency surgical resection. Similarly, the case published in Frontiers in Cardiovascular Medicine by Issa et al., [9] highlights that SVC syndrome from a right atrial mass is diagnostically treacherous because malignant etiologies — lymphoma, sarcoma, lung cancer, and metastatic disease — are far more prevalent causes of SVC obstruction and may be incorrectly suspected first, particularly when CT demonstrates a large non-enhancing intracardiac mass with pleural and pericardial effusions, as in our patient.

The non-enhancement of our patient's mass on contrast CT was a pivotal clue. Cardiac myxomas are typically hypovascular, they typically show heteregeneous post contrast enhacement on delayed phases, depending on necrosis, calcification, and thrombosis. They sometimes show only a slight increase in attenuation values after contrast injection, or do not enhance after which may be insufficient to distinguish them from thrombi— unlike angiosarcomas, which show avid and heterogeneous enhancement. The absence of extracardiac extension, the lack of mediastinal adenopathy, the negative abdominal ultrasound, and the normal blood count collectively rendered a primary cardiac benign tumor the most parsimonious explanation — a diagnostic reasoning framework essential to avoid unnecessary biopsy attempts or empirical oncological treatment.

### 3.3 The role of multimodal imaging

Echocardiography remains the cornerstone of cardiac tumor diagnosis. A large meta-analysis confirmed its diagnostic sensitivity of 98.1% for cardiac myxoma [4]. The classical TTE features — a mobile, echogenic mass attached to the interatrial septum — were clearly present in our case and allowed precise characterization of the tumor's origin, size, mobility, and hemodynamic impact. Transesophageal echocardiography (TEE), while not performed in this case, offers superior sensitivity (approaching 100%) and spatial resolution, particularly for small or atypically located tumors, and should be considered when TTE is inconclusive or when surgical planning requires anatomical precision [3, 11].

Contrast-enhanced CT provided complementary information, particularly regarding extracardiac extent, vascular relationships, and the absence of pulmonary embolism — a critical consideration in a patient with a large right-sided cardiac mass and clinical SVC obstruction. CT also confirmed the absence of pulmonary parenchymal metastases or mediastinal involvement, supporting the primary intracardiac mass etiology of the SVC. Cardiac MRI, the reference standard for tissue characterization of cardiac masses [3], was not performed given the urgency of the clinical

presentation and the high degree of diagnostic confidence provided by combined CT and TTE.

### 3.4 Surgical strategy and the imperative of urgency

The contemporary consensus is unambiguous: once a cardiac myxoma is identified, surgical resection should be undertaken promptly, as the risks of watchful waiting — tumor fragmentation, catastrophic embolization, acute hemodynamic decompensation, or sudden death — far outweigh the risks of elective cardiac surgery [1, 4, 18]. The excellent surgical outcomes documented in large institutional series corroborate this approach: operative mortality in contemporary cohorts is approximately 1.27%, with survival rates comparable to the general population and a recurrence rate below 1 per 1,000 person-years [4].

In our patient, the decision to operate without delay was reinforced by the advanced hemodynamic compromise, the degree of tricuspid obstruction (mean gradient 13 mmHg, indicative of severe stenosis), and the presence of an associated thrombus — which carried an independent risk of pulmonary embolization. The concomitant DeVega annuloplasty was a necessary solution to the secondary tricuspid annular dilation, which would otherwise have perpetuated right heart dysfunction despite tumor removal.

### 3.5 Histopathological Confirmation and the Importance of Systematic Analysis

Histopathological examination remains the definitive diagnostic tool for intracardiac masses, even when the clinical, echocardiographic, and CT findings are strongly suggestive of myxoma. Pathological confirmation is essential to exclude malignant mimics — notably angiosarcoma and primary cardiac lymphoma — which may share similar imaging characteristics and require entirely different therapeutic strategies.

In our case, the characteristic loose myxoid stroma with scattered stellate and polygonal cells provided unequivocal confirmation [3, 11]. Of note, the resected specimen also included an associated organized thrombus, which likely contributed to the clinical SVC picture by further reducing the effective RA lumen.

### 3.6 Comparison with Published Similar Cases

Table 1 summarizes selected published cases of right atrial myxoma presenting with SVC syndrome or significant SVC-related obstruction. Across the literature, the affected patients range in age from the third to the seventh decade, tumor sizes on imaging typically exceed 5 cm in at least one dimension, and the clinical presentation consistently involves the classic signs of SVC obstruction alongside signs of right heart failure. Surgical resection is universally the definitive treatment, and outcomes are generally favorable when intervention is timely [7, 8, 9].

| Reference | Age/Sex | Tumor Size | Attachment Site | Key Features | Outcome |
|---|---|---|---|---|---|
| Longatto et al., 2018 [7] | Not specified | >50 mm | Right atrium | SVC syndrome, giant RA mass, emergency surgery | Successful excision |
| Zhang et al., 2013 [9] | 59 y / F | Large (SVC origin) | Anterior wall of SVC | Tricuspid obstruction, right heart failure, SVC origin | Complete excision, full recovery |
| Teixidó et al., 2007 [8] | Not specified | Large | SVC extending to RA | Myxoma from SVC extending to right pulmonary artery | Surgical resection |
| Present case, 2026 | 65 y / F | 67 × 37 mm (CT) 60 × 32 mm (echo) | Interatrial septum | SVC syndrome, NYHA IV, tricuspid obstruction (gradient 13 mmHg), pericardial effusion, thrombus | Complete excision + DeVega annuloplasty + thrombectomy; uneventful recovery |

*RA: right atrium; SVC: superior vena cava; F: female; CT: computed tomography.*

**Table 1:** Published Cases of Right Atrial Myxoma with Superior Vena Cava Involvement

## 4. Conclusion

The present case illustrates the extraordinary diagnostic and therapeutic challenge posed by a right atrial myxoma presenting as SVC syndrome — a clinical scenario that is easy to misattribute to malignancy or mediastinal pathology. Several lessons emerge from this case and from the broader literature:

First, right atrial myxoma must be included in the differential diagnosis of any patient presenting with SVC syndrome, particularly in the absence of a primary extra thoracic malignancy or prior venous instrumentation. The combination of a non-enhancing intracardiac mass on CT with the morphological features of myxoma on echocardiography provides a high degree of diagnostic certainty.

Second, and most importantly, surgical resection — once the diagnosis is established — should be performed without delay. The exceptional postoperative outcomes documented in this and similar cases confirm that timely intervention transforms a potentially fatal condition into a largely curable one.

## Declarations

**Patient consent:** informed consent was obtained from the patient for publication of this case report.

**Conflicts of interest:** The authors declare no conflicts of interest.

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