# Cardiac Angiosarcoma

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## INTRODUCTION

The detection of cardiac masses can pose a diagnostic challenge. Cardiac masses encompass a broad spectrum of entities that can be categorized into nonneoplastic masses, benign neoplasms, primary malignant neoplasms, metastatic tumors, and pseudomasses/normal variants. Thrombus is the most common cardiac mass overall, whereas metastasis is the most common cardiac neoplasm.[1,2] We present a rare case of a patient with upper venous congestion due to a rapidly growing cardiac angiosarcoma, demonstrating the use of multimodality imaging in the diagnostic pathway and highlighting the potential of simultaneous orthogonal transesophageal echocardiography (TEE) to guide complicated cardiac biopsies.

## CASE PRESENTATION

A 68-year-old patient was transferred for further diagnosis and treatment after a computed tomography (CT) scan that raised suspicion of a large thrombus in the superior vena cava (SVC). For the past several weeks, the patient had experienced swelling of the eyelids, increasing exertional dyspnea, fever, and weight loss (6 kg in 3 weeks); in addition, infection markers were elevated. Cardiovascular examination revealed no cardiac murmurs or lower extremity edema, the heart rate was 82 beats per minute, blood pressure was 120/60 mm Hg, and peripheral oxygen saturation was 96%.

The mass could barely be visualized on transthoracic echocardiography (TTE) and thus could have been easily missed. Because TTE results were inconclusive, we therefore performed a TEE, which revealed an extensive, heterogeneous, and irregularly shaped soft tissue mass attached to the wall of the SVC and the right atrium (Figure 1, Videos 1–4). Systolic function of the right and left ventricle was normal. There was complete occlusion of the SVC (Figure 1B, D), accounting for the eyelid edema via SVC congestion. The mass also encased the atrial pacemaker lead implanted 18 months prior (marked with black arrow) (Figure 1B, D; Figure 2A, B; Videos 2 and 4). Pacemaker interrogation showed normal function without abnormalities in sensing or impedance. Color flow and continuous-wave Doppler showed increased flow and aliasing, which suggested obstruction of tricuspid inflow (Figure 2A–C, Videos 5–6). TEE also showed a large patent foramen ovale (Figure 1A, *arrow*), which was held open by the altered blood flow through the inferior vena cava into the right atrium. As the imaging modalities raised the possibility of a superimposed thrombus, indicating an increased risk for

## VIDEO HIGHLIGHTS

**Video 1:** Two-dimensional TEE, mid-esophageal long-axis (110°) bicaval view, demonstrates a large, heterogenous, multilobular, partially mobile mass within the right atrium and a leftward deviated interatrial septal position with an apparent anatomically patent foramen ovale. The pacemaker wire and associated acoustic shadowing artifact are also seen.

**Video 2:** Two-dimensional TEE, mid-esophageal long-axis (120°) bicaval view, more fully demonstrates the origin of the large, heterogenous, multilobular, partially mobile mass completely filling and protruding from the dilated SVC. The entrapped pacemaker device lead is seen as a bright echo within the center of the mass.

**Video 3:** Two-dimensional TEE, mid-esophageal short-axis (60°) view, more fully demonstrates the portion of the large, heterogenous, multilobular, partially mobile mass within the right atrium. The mass directly encases the pacemaker lead and extends to the tricuspid valve. Also visualized are the leftward deviation of the interatrial septum, the relationship to the right ventricular outflow tract and the pulmonic valve, and a normal trileaflet aortic valve.

**Video 4:** Two-dimensional TEE, mid-esophageal simultaneous orthogonal long-axis (*left*, 110°; *right*, 200°) view obtained at the time of the biopsy, demonstrates the steerable delivery sheath entering the right atrium via the inferior vena cava and positioned directly adjacent to the large mass for deployment of the bioptome.

**Video 5:** Two-dimensional TEE, mid-esophageal 4-chamber (0°) right heart focused view, demonstrates the normal biventricular size and global systolic function, the large, heterogenous, multilobular, partially mobile mass within the right atrium, and its anatomic relationship to the tricuspid valve and the pacing lead.

**Video 6:** Two-dimensional TEE, mid-esophageal 4-chamber (0°) right heart focused view with color flow Doppler, demonstrates the narrow passage of blood flow around the large, heterogenous, multilobular, partially mobile mass within the right atrium, with flow acceleration suggesting mild inflow obstruction.

**View the video content online at www.cvcasejournal.com.**

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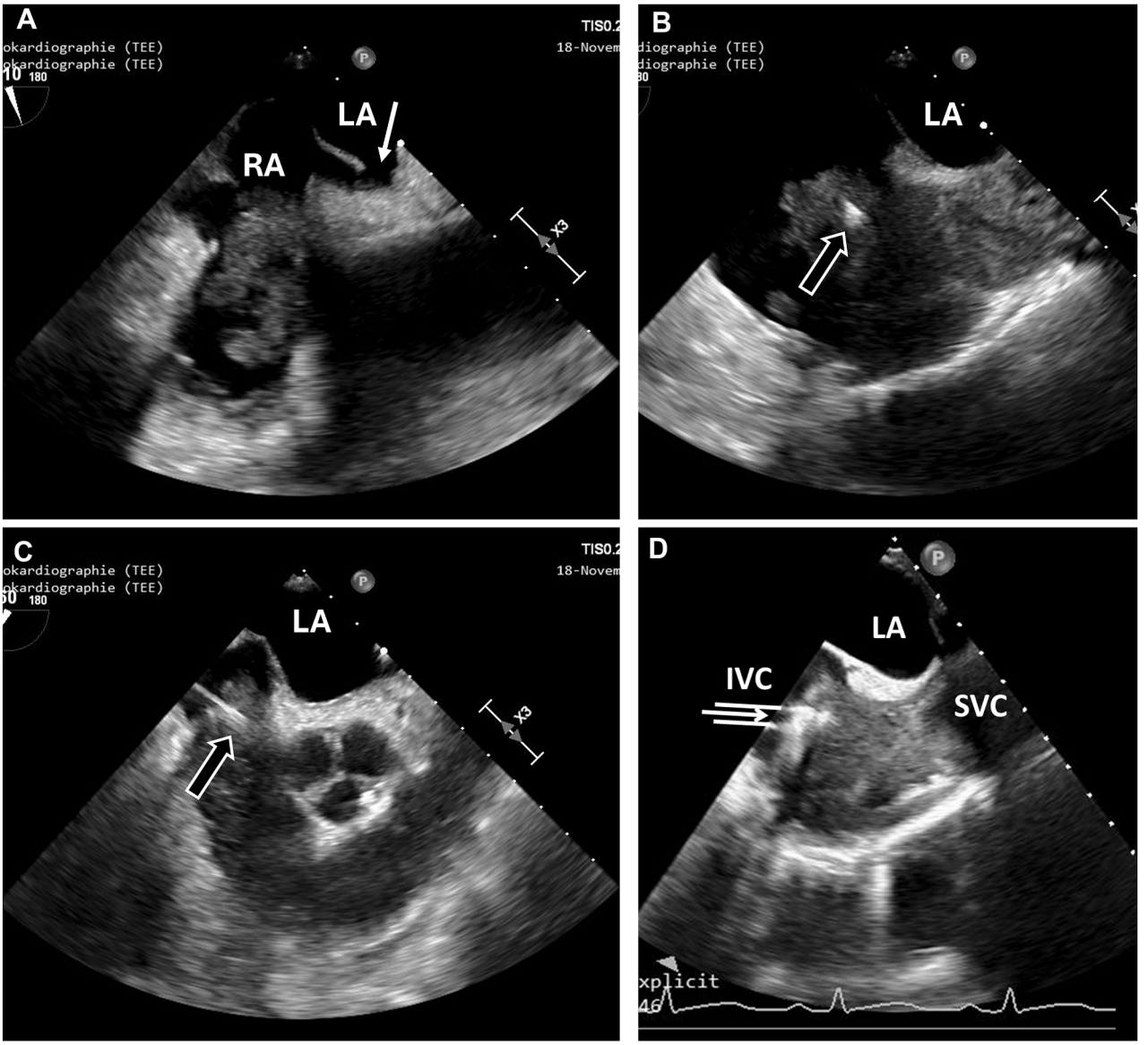



**Figure 1** Two-dimensional TEE, mid-esophageal bicaval long-axis (**A**, 110°; **B**, 120°), and short-axis (**C**, 60°) views obtained before biopsy, demonstrates a large, heterogenous, multilobular, partially mobile mass within the RA, completely filling and protruding from the dilated SVC with extension to the tricuspid valve. The image also shows the relationship to the right ventricular outflow tract and the pulmonic valve, a normal trileaflet aortic valve (in diastole), an entrapped pacemaker device lead (*black arrow*), and a leftward deviated interatrial septal position with an apparent anatomically patent foramen ovale (*white arrow*). Two-dimensional TEE, mid-esophageal long-axis (**D**, 120°) view obtained at the time of the biopsy, demonstrates the steerable delivery sheath (*horizontal arrow within parallel white lines*) entering the RA via the IVC and positioned directly adjacent to the large mass for deployment of the bioptome. *IVC*, Inferior vena cava; *LA*, left atrium; *RA*, right atrium.

paradoxical embolisms, anticoagulation and intensified monitoring were initiated. The primary strengths of echocardiography include its high spatial and temporal resolution, which provide detailed imaging of cardiac masses and cardiac function. However, additional imaging modalities are necessary to achieve tissue characterization and to provide the exact relationship between the mass and the surrounding tissue. Therefore, we also performed a positron emission tomography (PET)/CT scan and an additional high-resolution CT scan. The PET scan of the patient showed increased glucose uptake of the mass (Figure 3A, B). The CT scan demonstrated the obstruction of the SVC and the infiltration of the vessel wall.

Due to the lack of other access routes, a TEE-guided biopsy of the mass was performed via the femoral vein and inferior vena cava (8.5F steerable sheath) without complications (Figure 1D, Video 4). Live simultaneous orthogonal TEE imaging helped to steer the catheter and to avoid damage to the thin atrial wall and the pacemaker lead. The mass was histologically identified as an angiosarcoma. Treatment with paclitaxel was started. The initial dose was

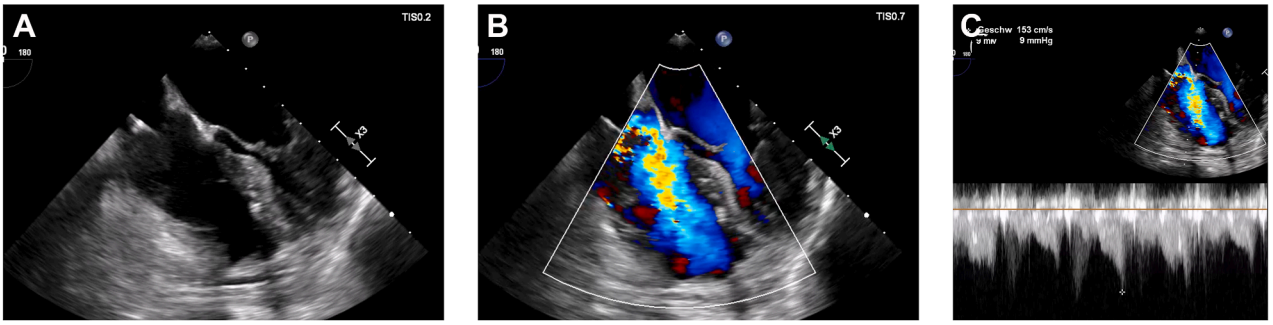



Figure 2 Two-dimensional TEE, mid-esophageal 4-chamber (0°) right heart focused view **(A)** without and **(B)** with color flow Doppler, including **(C)** continuous-wave spectral Doppler display, demonstrates the large mass within the right atrium, its anatomic relationship to the tricuspid valve and the pacing lead, the narrow passage of blood flow around the mass with color aliasing and flow acceleration confirmed with continuous-wave Doppler, suggesting mild inflow obstruction (maximal pressure gradient 9 mm Hg).

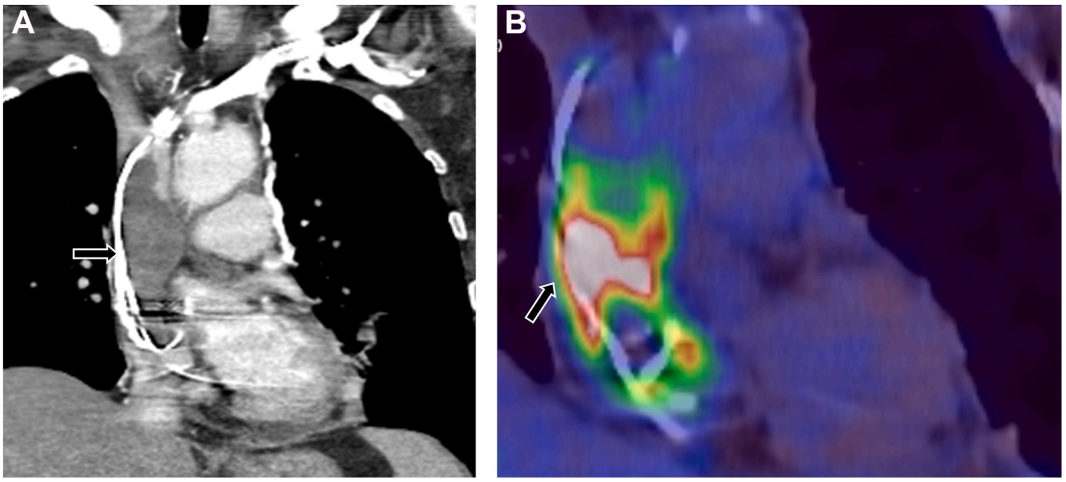



Figure 3 **(A)** Chest CT scan with contrast and **(B)** PET/CT scan, sagittal displays, demonstrate the large mass within the SVC and RA that is metabolically active (fluorodeoxyglucose uptake; *solid white* within the narrow *red*, *yellow*, and *green* borders) was later confirmed to be an angiosarcoma; the pacemaker lead is also noted (*black arrow*).

administered in hospital and was well tolerated; future treatment was organized as outpatient therapy. Four months after therapy initiation, the patient's health status further deteriorated. Diagnostic workup revealed new hemorrhagic brain metastasis and new pulmonary metastasis. The cerebral metastasis was treated symptomatically with dexamethasone, resulting in clinical improvement. Due to the disease progression despite chemotherapy, the therapy concept was changed to best supportive care.

## DISCUSSION

Primary malignant cardiac tumors are extremely rare, and incidence is not easily estimated (approximately 0.0001%–0.0003%).[3,4] The most common symptom is dyspnea (59%–88%),[3,5] which is likely caused by obstructive growth but can also include pericardial effusion, cardiac tamponade, and valvular dysfunction.[3,6] The main symptom in our patient was upper venous congestion. Diagnosis included multimodality imaging, including TTE, TEE, CT, PET/CT, and magnetic resonance imaging, followed by a tissue biopsy.[3] The present case strikingly demonstrates a rapidly growing lesion, highlighting how echocardiography, in combination with other imaging modalities and invasive procedures, is essential to guide the diagnostic pathway and inform therapeutic decisions.

Total or partial obstruction of large veins often leads to collateral pathways that can be further explored with contrast-enhanced echocardiography.[7] Surgical removal is often challenging because of the infiltrative growth[3] and was technically not possible in our patient. Omair *et al*.[8] reported a similar case of angiosarcoma that caused obstruction of the right outflow tract. The mass could be partly surgically removed and histologically examined. As this was not feasible in our case, we performed a TEE-guided biopsy of the tumor to determine the definitive histopathology of the mass. The procedure was complicated for 2 reasons: first, the right atrial wall is very thin and can easily be punctured by a misguided bioptome, and second, the encompassed pacemaker lead also posed a possible obstacle, which could be damaged by the bioptome. The unique strength of real-

time simultaneous orthogonal TEE imaging is to provide high temporal and spatial resolution that helped to safely guide the procedure without complications. This approach can be recommended for cardiac biopsies with complicated anatomy. Conservative treatment options include chemotherapy. The regimen depends on various factors, such as tumor stage, patient's overall health, and individualized treatment plans. In our case, the tumor board decision was taxanes (i.e., paclitaxel), and therapy was started. This case highlights a rare case of a rapidly growing angiosarcoma and the value of initial echocardiography and subsequent multimodality imaging.

## CONCLUSION

Cardiac angiosarcomas are a very rare primary malignant cardiac tumor. Our case shows the importance of multimodality imaging in the diagnosis of cardiac masses. Also, it showcases the use of TEE for guiding tumor mass biopsy.

## ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with the following guidelines (detailed in box below).

## CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

## FUNDING

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## DISCLOSURE STATEMENT

The authors reported no actual or potential conflicts of interest relative to this document.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2026.09.004.

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