KANSAS JOURNAL of MEDICINE

Case Report

Metastatic Cardiac Angiosarcoma Presenting as Superior Vena Cava Syndrome and Cardiac Tamponade

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Received Nov. 23, 2024; Accepted for publication Apr. 25, 2025; Published online Jun. 16, 2025
Kans J Med 2025 May-Jun; 18:61-63. https://doi.org/10.17161/kjm.vol18.23143

INTRODUCTION

Primary cardiac angiosarcomas are rare, aggressive vascular endothelial tumors associated with a poor prognosis among soft tissue sarcomas.[1] Their clinical presentation often is variable and nonspecific, commonly including chest pain, shortness of breath, and palpitations, symptoms that result from hemodynamic changes due to structural invasion of the heart.[2,3] The diagnostic workup involves a combination of noninvasive imaging, such as transthoracic echocardiogram (TTE), computed tomography (CT), and magnetic resonance imaging (MRI), along with invasive biopsy and immunohistochemical analysis for confirmation.

There is limited literature establishing a robust standard of care for cardiac angiosarcoma. Current treatment approaches include surgery, radiotherapy, and chemotherapy.[1,4] However, recent advances in care, particularly those guided by immunogenetics, show promise, with success often dependent on early diagnosis.[4]

We describe a challenging case of newly diagnosed cardiac angiosarcoma in a 50-year-old male who presented with superior vena cava (SVC) syndrome, complicated by cardiac tamponade.

CASE REPORT

A 50-year-old male with a history of hypertension initially presented with throat pain, right-sided sublingual swelling, episodic secretions, and voice changes lasting three weeks. CT imaging of the neck revealed retropharyngeal effusion and significant edema of the upper oropharyngeal airway, though the laryngeal airway remained intact. Given the patient's chills and new leukocytosis (16,430 WBCs/μL), he was started on broad-spectrum intravenous (IV) antibiotics and experienced symptomatic improvement. Flexible fiberoptic laryngoscopy showed esophageal collapse without edema, and cultures were negative. The patient was discharged on oral antibiotics with a decreased suspicion for Ludwig's angina.

Two days post-discharge, the patient returned with recurrent throat pain and sublingual swelling, now accompanied by new facial plethora. CT of the chest revealed a filling defect within the SVC and upper right atrium, prominent mediastinal venous collaterals, moderate circumferential pericardial effusion (up to 1.6 cm anteriorly), and a 1.3 cm right thyroid nodule. He was started on IV heparin due to concern for SVC syndrome from a possible thrombus or embolus. Positron emission tomography (PET)/CT identified a hypermetabolic right atrial mass (5 x 4.6 cm, Standardized Uptake Value [SUV] max 11.6) infiltrating the atrial appendage, and focal activity in the right thyroid nodule (SUV max 10.6). Given a newly elevated thyroid stimulating hormone (TSH) level (10.5 mIU/L) and a family history of head and neck cancer, the thyroid nodule was biopsied and found to be benign. The patient opted for discharge with close outpatient follow-up and was transitioned to oral apixaban after resolution of facial plethora and throat symptoms.

Two weeks later, he returned with facial swelling and new-onset dizziness. He was found to be in atrial flutter with a resting heart rate of 100 beats per minute. Repeat imaging showed an enlarging right atrial mass (5.6 x 5.3 x 4.9 cm) invading the right mediastinum and SVC, with thrombus extending to the level of the azygos vein. Mediastinal edema was seen extending into the SVC-cavoatrial junction (Figure 1). CT of the chest also revealed a new, large pericardial effusion. The patient exhibited clinical signs of cardiac tamponade (Beck's triad: hypotension, muffled heart sounds, and jugular venous distension), requiring urgent pericardiocentesis with drainage of 650 mL of dark blood and catheter placement. Limited TTE showed reduced systolic function, which improved following the procedure.

Biopsy of the right atrial mass and pericardial fluid analysis were positive for erythroblast transformation-specific related gene (ERG) and cluster of differentiation (CD31) on immunohistochemistry. The fluid was negative for MOC31, Ber-EP4, WT1, and Calretinin. These findings were concerning for metastatic angiosarcoma. The case was presented at a multidisciplinary sarcoma tumor board. Due to extensive tumor infiltration into the right atrial wall (Figure 2), the patient was not a surgical candidate. He was started on neoadjuvant chemotherapy with weekly paclitaxel.

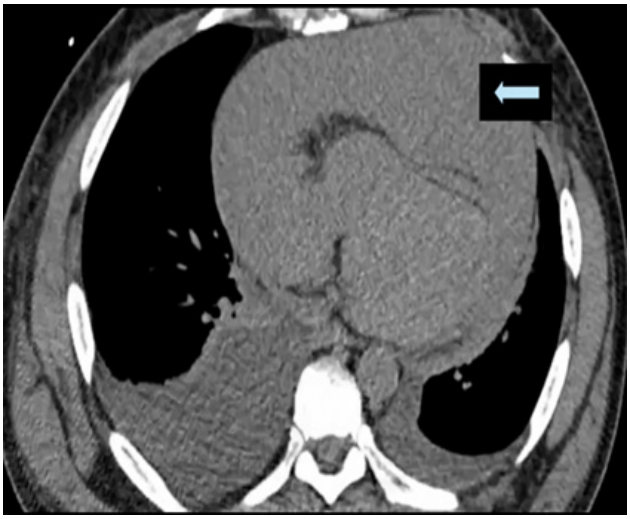

Figure 1. Axial CT demonstrating new large pericardial effusion with tamponade.

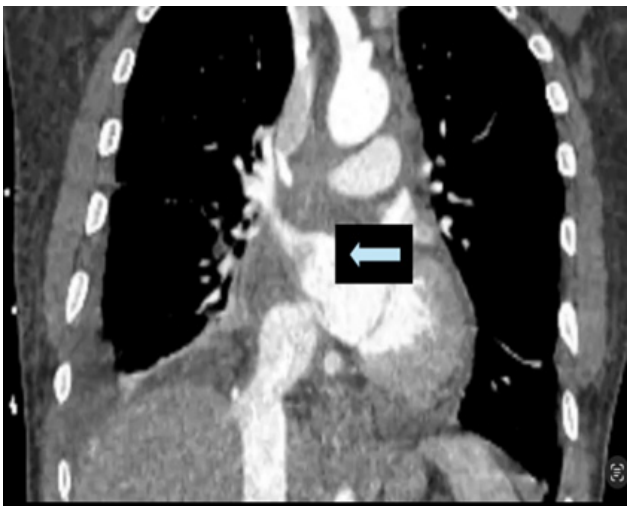

Figure 2. Coronal CT demonstrating right atrial mass measuring 5.6 x 5.3 x 4.9 cm, invading the superior vena cava with thrombosis and right atrial wall.

DISCUSSION

Cardiac angiosarcomas account for less than 2% of all sarcomas.[5] The case presented in this article highlights the extensive challenges involved in the timely and accurate diagnosis of cardiac angiosarcoma. Although these tumors are highly aggressive, their initial presentation often is variable and nonspecific, complicating diagnosis.

This patient initially presented with throat pain, voice changes, and swelling in the floor of the mouth, raising concern for Ludwig's angina, a condition that requires urgent evaluation and was ultimately ruled out.[6] The presentation of Ludwig's angina in the context of an underlying sarcoma is rare. Vijapur et al.[7] reported a case of a 21-year-old with submandibular swelling concerning for Ludwig's angina; subsequent imaging revealed mandibular osteomyelitis and led to the diagnosis of osteosarcoma. However, to date, there are no similar reports linking Ludwig's angina–like presentation with cardiac angiosarcoma. This underscores the complexity of clinical decision-making, especially when initial imaging suggests a potential airway emergency in the absence of thrombosis.

Concern for SVC syndrome arose following new CT imaging that revealed a filling defect within the SVC and right atrium, along with prominent venous collaterals.[8] While over 50% of SVC syndrome cases are associated with malignancy, they are most commonly due to non-small cell lung cancer, small cell lung cancer, or lymphoma.[9-14] Few cases in the literature describe SVC syndrome caused by angiosarcoma. Abratt et al.[15] described a 7 cm mediastinal mass from a primary angiosarcoma involving the SVC, and Salgueiro et al.[16] reported SVC thrombosis secondary to primary angiosarcoma involving the brachiocephalic veins. SVC syndrome also has been observed in other sarcomas, such as sarcomatoid renal cell carcinoma and osteosarcoma.[7,17,18]

The clinical suspicion for SVC syndrome in this case led to early CT angiography, which helped identify thrombosis. The presence of venous collaterals can be an important imaging clue. Subsequent PET/CT imaging aided in characterizing metabolic activity and helped distinguish benign from malignant lesions.[8,15,16]

Cardiac tamponade is another rare presentation of cardiac angiosarcoma.[20-22] In this case, the patient initially had a moderate (1.6 cm) circumferential pericardial effusion, which progressed to tamponade and required pericardiocentesis. The aggressive nature of cardiac angiosarcoma may contribute to rapid fluid accumulation. While there are case reports of direct oral anticoagulants causing hemorrhagic tamponade, no studies to date have explored this specifically in the setting of cardiac angiosarcoma, although it remains a plausible contributing factor.[23-25]

Due to its rarity, there are no standardized treatment protocols for cardiac angiosarcoma. Surgical resection or debulking often is considered, though the tumor's aggressive and infiltrative nature may necessitate complex reconstruction of intrathoracic structures to achieve negative margins.[26,27] In this case, atrial flutter further complicated clinical management. Patients without surgical resection typically have a median survival of only three to six months.[28,29]

Some cases have reported the use of neoadjuvant chemotherapy in unresectable cardiac angiosarcoma.[30,31] A multicenter phase II trial of paclitaxel in patients with metastatic or unresectable angiosarcoma demonstrated a non-progression rate of 62.5% and a median survival of 16 months.[31] However, additional research is needed to determine the most effective treatment strategies for this rare and challenging malignancy.

CONCLUSIONS

This case highlights the complex diagnostic challenges associated with cardiac angiosarcoma. The patient initially presented with symptoms suggestive of SVC syndrome, which were due to an underlying, undiagnosed cardiac angiosarcoma. The diagnosis was ultimately made during a subsequent presentation with cardiac tamponade. This underscores the importance of maintaining a high index of suspicion for cardiac angiosarcoma in patients presenting with SVC syndrome, as early recognition is crucial for initiating appropriate treatment.

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Keywords: superior vena cava syndrome, cardiac tamponade, sarcoma, clinical oncology