Cambridge City Publishers

Case Reports in Clinical Cardiology Journal

Case Report | Vol 6 Iss 3

A Complex Pericardial Effusion and Diagnostic Dilemma: A Case of Primary Pericardial Angiosarcoma

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Received: April 09, 2026; **Accepted:** April 27, 2026; **Published:** May 05, 2026

Abstract

Pericardial angiosarcoma has a poor prognosis and remains challenging to treat. There is no standard of treatment due to its rarity, but successes have been reported in the literature. Early diagnosis is essential to improve overall mortality and often requires multimodal imaging. Perhaps more recognition can lead to more established treatment guidelines, allowing clinicians in the future to act accordingly and improve outcomes.

Keywords: Angiosarcoma; Pericardial Effusion; cMRI

Introduction

Angiosarcomas are tumors of endothelial cell origin. They are largely found on the skin and soft tissue. Primary cardiac angiosarcomas comprise most cardiac malignancies and are exceedingly rare but are aggressive, often carrying a poor prognosis [1,2]. Surgical resection remains the gold standard for treatment and combination with adjuvant therapy has also been effective and has been previously reported in the literature [3]. We describe a patient who underwent pericardiocentesis for symptomatic pericardial effusion with cardiac tamponade and who required multiple readmissions due to refractory symptoms and recurrence despite guideline-derived medical therapy and acute interventions. The patient was found to have a conventional type of angiosarcoma through pericardium biopsy. Angiosarcomas, being rare, have not been extensively reported in the literature, and this report aims to educate on the diagnostic and therapeutic challenges associated with this disease process.

Case Presentation

Our case describes a 48-year-old female with no significant past medical history who presented with complaints of syncope. Initial echocardiogram showed a large pericardial effusion with tamponade physiology. Pericardiocentesis was performed with sanguinous output. Initial workup was only positive for CCP and dsDNA antibodies, with negative TB PCR and cytology. She was thus treated with hydroxychloroquine and ibuprofen for presumed rheumatoid arthritis pericarditis.

She had recurrent effusive pericarditis with readmission in one month. CT imaging demonstrated a hyperdense complex pericardial effusion as well as multiple bilateral pulmonary nodules (Figure A). A cardiac window was pursued, but no pericardial fluid was identified. A cardiac MRI (cMRI) was then obtained, which showed a large, irregularly contoured mediastinal mass extending from the aortic arch to the pericardium with a superior aspect of the mass encasing the ascending thoracic aorta, aortic arch, aorta root, and main pulmonary artery (Figure B,C and D). The patient underwent a Chamberlain Procedure, but the pathology from the evacuated clot was unrevealing. PET-CT redemonstrated a complex pericardial effusion without a discrete mass and non-avid pulmonary nodules. She further underwent sternotomy and pericardial evacuation and was discharged after a lengthy hospital admission.

She presented again with recurrent sanguinous effusion and underwent a palliative pericardiocentesis, which was complicated by hypotension, bradycardia, and cardiac arrest. Ultimately, the patient passed from hemorrhagic tamponade.

The pericardium was found to be diffusely infiltrated by a malignant spindle and epithelioid tumor that consisted of anastomosing thin-walled vascular channels filled with erythrocytes and lined by moderately pleomorphic round to spindle-shaped cells with distinct nucleoli. Mitotic figures were frequently seen. Tumor cells were positive for CD31, CD34, and ERG and negative for CK, AE1/AE3, calretinin, WT-1, D2-40 and HHV8. Pathology reports were positive for angiosarcoma, the conventional type (Figure E,F).

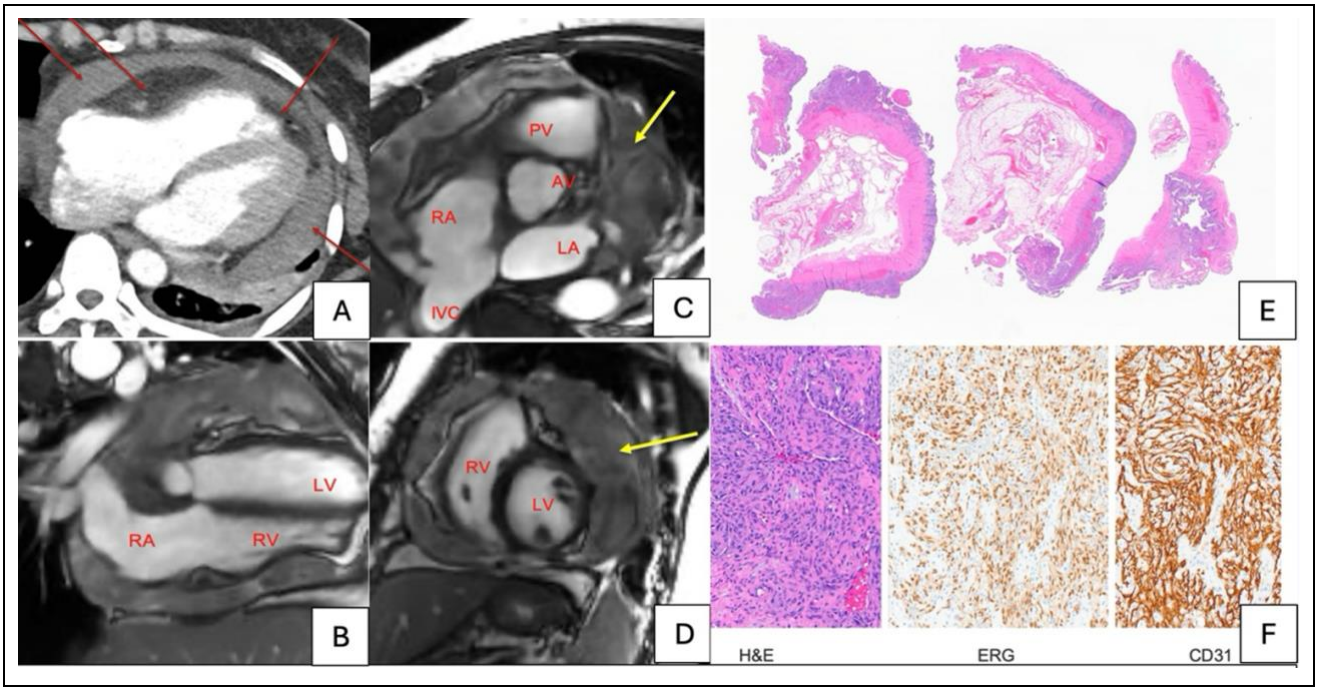

Figure A: CT chest / abdomen showed a large hyperdense opacity within the pericardium, likely representing a complex pericardial effusion with average HU up to 53.

Figure B,C and D: Cardiac MRI with contrast showed a large, irregularly contoured mediastinal mass with heterogenous contrast enhancement, encasing the ascending thoracic aorta, aortic root, and main pulmonary artery.

Figure E and F: Pericardium tissue, diffusely involved by a malignant spindle and epithelioid tumor, tumor cells stained positive for ERG and CD31.

Discussion

Cardiac angiosarcomas, although uncommon, account for over 25% of all primary cardiac malignancies. They are considered the most aggressive and carry a dismal prognosis [1]. It predominantly affects the right side of the heart, and its early infiltration and metastasis make treatment challenging. No standard of treatment currently exists due to the rare nature of disease, although some reports of radiotherapy, surgical resection, and adjuvant chemotherapy have seen successes [1,3,4]. Most cases are found in patients less than 65 years of age, and metastasis is common by the time of diagnosis, making full surgical resection impractical.

Some cases of success have been reported in the literature. The literature reported a case report patient who followed a similar course presentation as our patient with a pericardial effusion. She was initially discharged but presented again with chest pain and a CT and cMRI showed a mass in the right atrium with a biopsy confirming the diagnosis of cardiac angiosarcoma. She underwent chemotherapy and radiotherapy to shrink the tumor and remained asymptomatic apart from mild dysphagia after complete surgical resection eight months post-surgery [1].

Patients in the early stages of angiosarcoma often present with nonspecific complaints, which often lead to a delayed diagnosis. Patients may present later with worsening symptoms of dyspnea due to fluid buildup and infiltration of the tumor into the lungs. Metastasis leads to systemic symptoms. Prompt diagnosis is essential to improve prognosis due to the aggressive nature of the disease but often requires multimodal imaging.

Currently, imaging studies such as cMRI and immunochemistry evaluations are used to diagnose angiosarcomas, and histological features include anastomosing vascular channels and solid spindle cell areas. Immunochemistry reveals markers such as CD31 and CD34 as seen in our patient, and factor VIII-related proteins [4]. However, as in the case of our patient, initial tissue sampling techniques, including a pericardial window and Chamberlain Procedure, were unrevealing, necessitating pericardial washout to confirm angiosarcoma, demonstrating the challenge of diagnosis.

Complete resection of the tumor before metastasis provides the best prognosis. However, this is difficult due to the rapid spread of the disease into surrounding structures. Radiotherapy before surgical resection and adjuvant chemotherapy post-surgical resection have been used in reported literature as mentioned above but the tumor often responds poorly to chemotherapy and radiation.

Conclusion

• Early recognition and diagnosis, often requiring multimodal imaging for angiosarcoma is critical to improve outcomes

• Despite multidisciplinary care, prognosis is often poor

• Future work is necessary to determine standard approach when clinicians are confronted with this disease process.

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