

Case Report

Primary Cardiac Angiosarcoma With Pulmonary Metastasis in A Young Female: A Case Report

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ABSTRACT

Cardiac angiosarcoma is a rare and highly aggressive malignancy with nonspecific clinical manifestations, often leading to delayed diagnosis. In this article, we report a 38-year-old woman who presented with diarrhea, dry cough, and a history of blunt chest trauma 3 weeks prior to admission. Initial echocardiography revealed massive pericardial effusion complicated by cardiac tamponade. Further diagnostic evaluation, including cardiac computed tomographic angiography, demonstrated right atrial wall rupture. Intraoperative sampling of the pulmonary nodules and the ruptured atrial wall was performed, and histopathologic examination confirmed metastatic cardiac angiosarcoma. Despite surgical intervention, and given the advanced stage of the tumor, the oncology team recommended palliative care. The patient developed progressive right-sided heart failure and died after a 53-day stay in the intensive care unit. This case highlights the diagnostic challenges of cardiac angiosarcoma and underscores the importance of early recognition and stage-appropriate management to improve clinical outcomes. (*Iranian Heart Journal* 2026; 27(4): 80-86)

KEYWORDS: angiosarcoma; malignant cardiac tumor; right atrial mass; pulmonary metastasis

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CASE PRESENTATION

A 38-year-old woman was admitted to our center with massive pericardial effusion and bilateral pleural effusions, predominantly on the right side. She had a history of blunt chest trauma 3 weeks prior to admission. Shortly after the trauma, caused by her 6-year-old child jumping on her chest, which caused severe acute chest pain, she developed premature fatigue, chronic diarrhea (3 weeks), tachypnea, and peripheral edema. On initial

evaluation, tachycardia, tachypnea, mild ascites, and mild lower-extremity edema were observed. Echocardiography revealed massive pericardial effusion with a swinging heart, diastolic collapse of the right atrium (RA) and right ventricle, and normal size and function of the left and right ventricles (Figure 1). Additionally, 500 mL bloody pericardial fluid was drained through needle pericardiocentesis. Because of persistent dyspnea and worsening bilateral pleural effusions, thoracentesis was performed. It

showed an exudative effusion with unremarkable cytology. Considering the diagnostic workup and the context of the COVID-19 pandemic, suspected infections were evaluated and excluded by a negative chest computed tomographic (CT) scan and SARS-CoV-2 polymerase chain reaction (PCR) result. Rheumatologic and immunologic test results were unremarkable, and tumor markers-including carcinoembryonic antigen (CEA), carbohydrate antigen (CA) 19-9, CA 15-3, and CA 125-as well as erythrocyte sedimentation rate, were within normal

limits. Only a mildly elevated C-reactive protein level was noted. Evaluation for COVID-19-related myopericarditis was performed using cardiac magnetic resonance 1 month later, which revealed mild left ventricular systolic dysfunction (left ventricular ejection fraction = 50%) without signs of myocarditis, myocardial edema, or fibrosis. Moreover, right atrial wall thrombosis was incidentally detected, most likely representing a sealed right atrial free-wall rupture or contusion. At the time, a conservative management approach was chosen by clinicians.

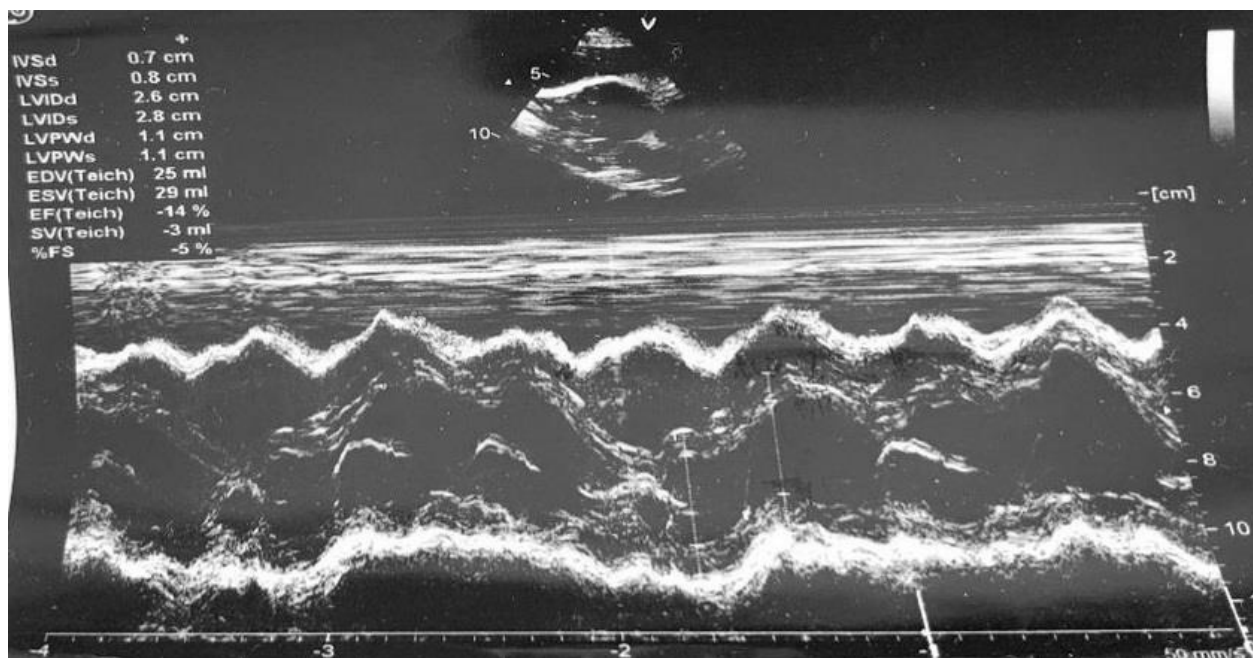


Figure 1. Echocardiography showing a massive pericardial effusion with a swinging heart

The patient remained largely asymptomatic for 2 months, although occasional cough and tachycardia (heart rate = 100–120 beats/min) were noted, while follow-up chest radiographs were unremarkable. After this period, her symptoms progressed, including worsening cough, dyspnea, and general malaise. A CT scan performed 1 month later revealed bilateral ground-glass opacities, findings commonly associated with miliary tuberculosis or advanced COVID-19 infection (Figure 2). Repeat SARS-CoV-2

PCR testing was performed and yielded a negative result. Several days prior to her second admission, she experienced acute worsening of dyspnea and generalized weakness. Another chest CT scan revealed multiple pulmonary nodules in both lungs, demonstrating a more aggressive pattern compared with the previous study, suggestive of neoplastic involvement (Figure 3). Further, the patient underwent echocardiography and contrast-enhanced multidetector computed tomographic (MDCT) imaging.

Echocardiography revealed severe pericardial effusion containing echogenic fibrinous strands and increased pericardial thickness; contrast-agitated saline was injected, but no bubble passage was observed. Contrast-

enhanced MDCT demonstrated an infiltrative tumor arising from the right atrium with a defect in the right atrial wall and extravasation of contrast, along with a large pericardial effusion (Figure 4).

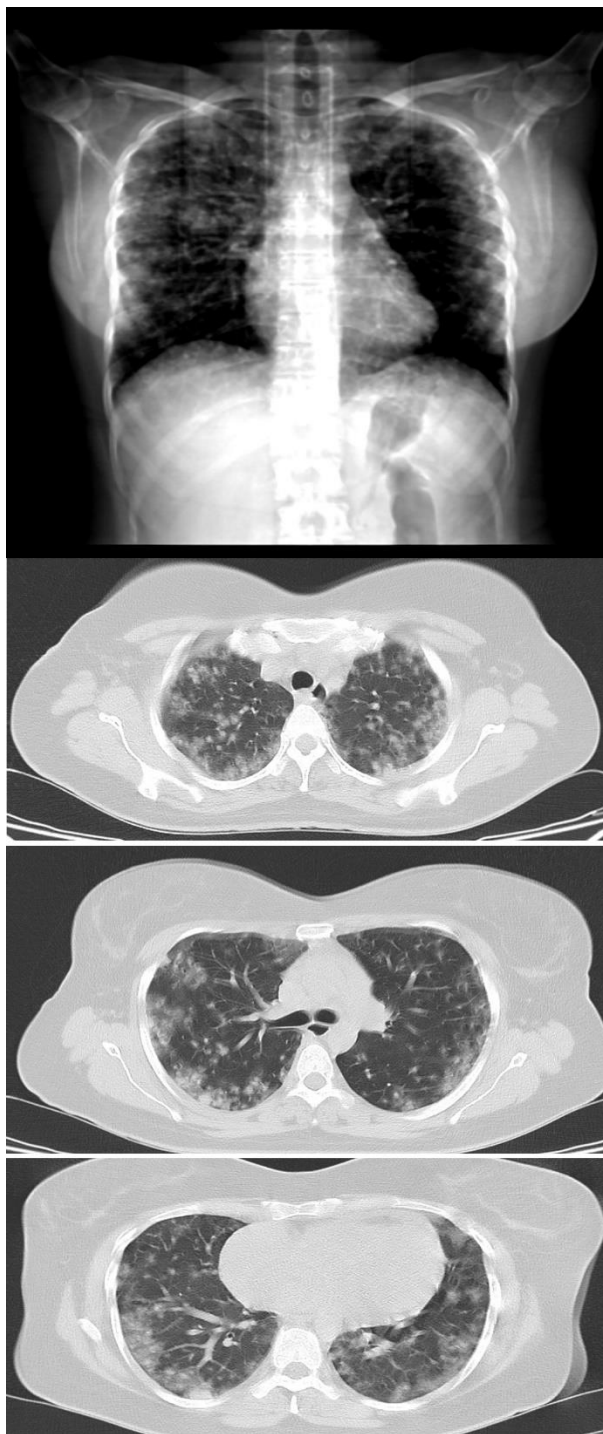


Figure 2. Chest radiograph and high-resolution computed tomographic (HRCT) scan showing bilateral ground-glass opacities

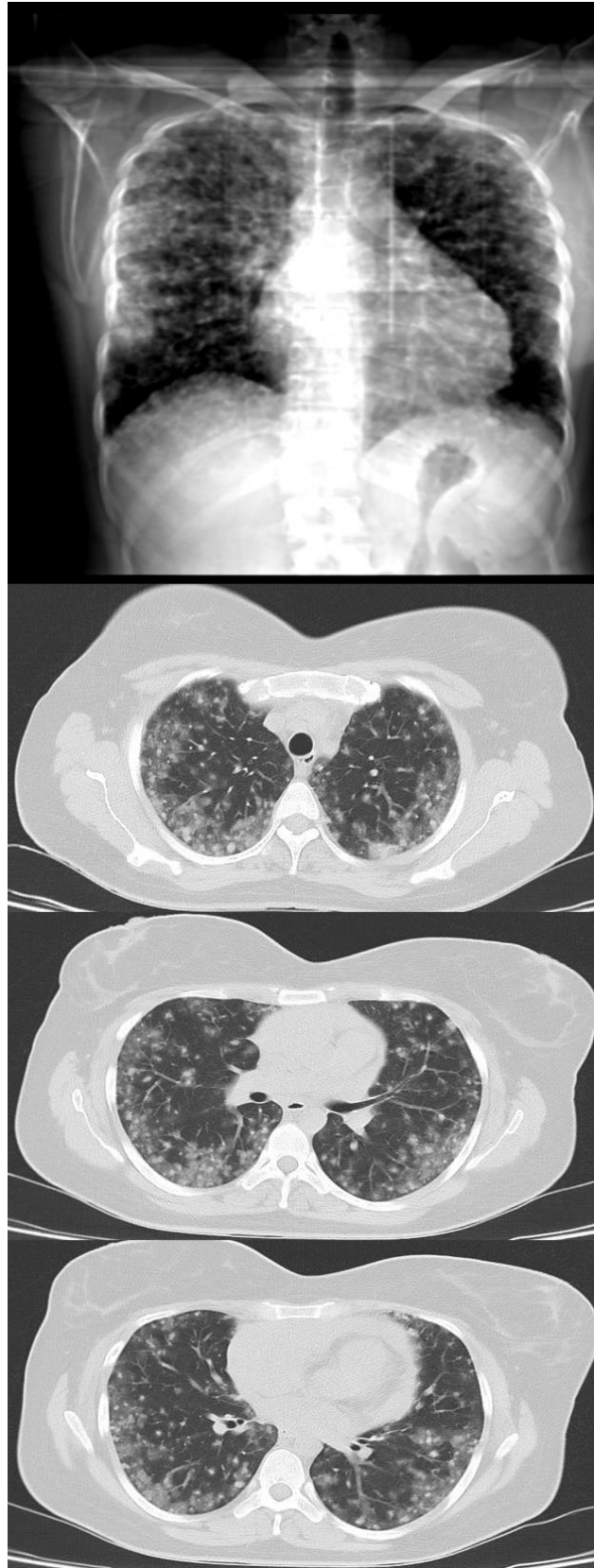


Figure 3. Chest radiograph and chest high-resolution computed tomographic (HRCT) scan showing a bilateral nodular pattern and pericardial effusion

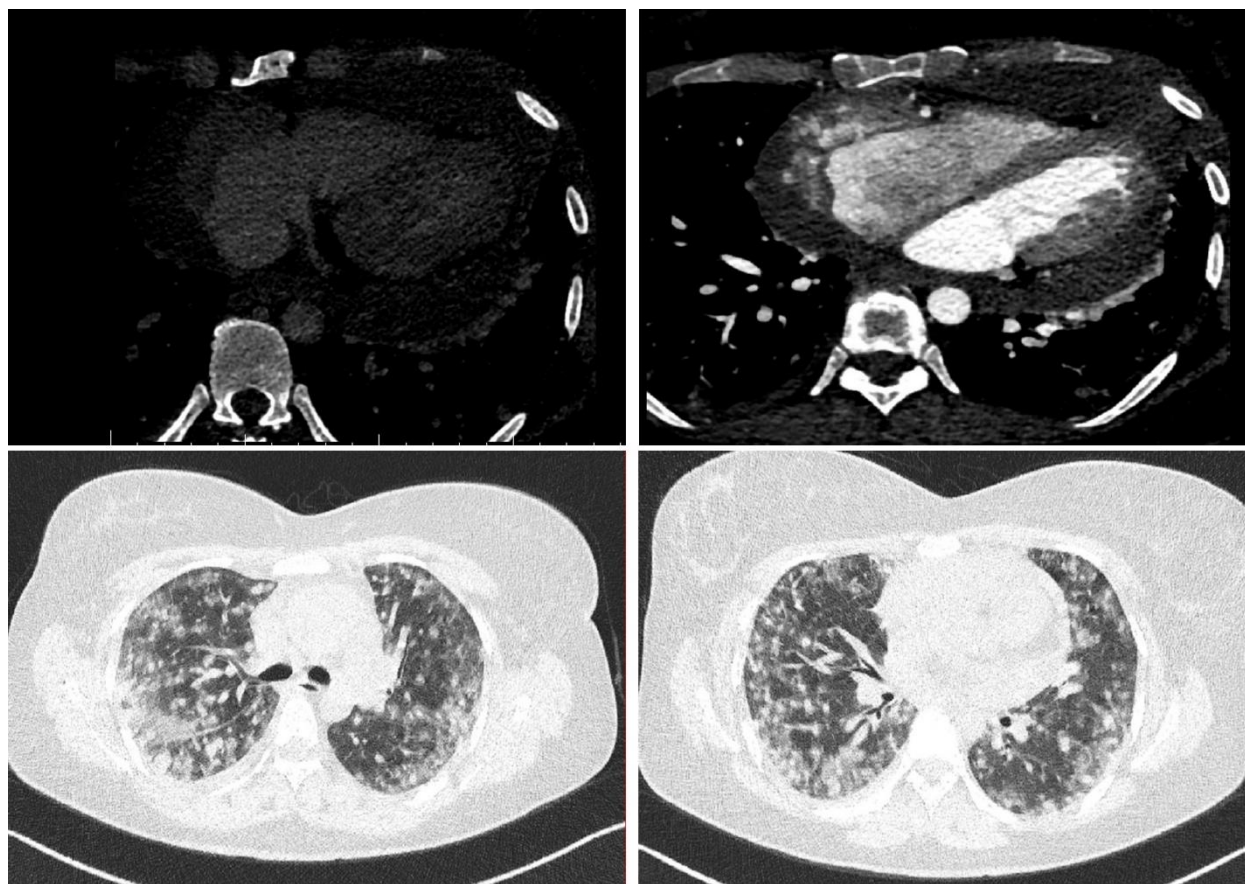


Figure 4. Contrast-enhanced spiral cardiac multidetector computed tomographic (MDCT) scan showing an infiltrative tumor arising from the right atrium, a right atrial wall defect with sealed intravenous (IV) contrast, a large pericardial effusion, and multiple pulmonary nodules

Ultimately, with a diagnosis of cardiac tamponade secondary to right atrial rupture, the patient underwent open-heart surgery following expert consensus. During a midline sternotomy, the pericardium appeared markedly inflamed, with active bleeding from the visceral pericardium. A large perforation measuring 30 × 30 mm was identified in the right atrial free wall, with adhesions between the pericardium and the site of the healed perforation. The surgeon also noted diffuse small, soft, yellow nodular lesions scattered throughout the parenchyma of all lung lobes. Biopsies were obtained from the lung nodules, lymph nodes, pericardium, and the tissue surrounding the right atrial defect. The patient was successfully weaned off cardiopulmonary bypass. Histopathologic

and immunohistochemical analysis of the lung nodules confirmed a poorly differentiated neoplasm suggestive of angiosarcoma, with tumor cells positive for CD34 and CD31 and negative for PanCK, CAM5.2, SALL4, calretinin, melan-A, S100, desmin, Bcl-2, and H-caldesmon. Histopathologic examination of the pericardium and lymph nodes revealed 2 reactive lymph nodes and fibrovascular tissue containing organized thrombi and focal chronic inflammation. However, despite surgical intervention, and given the advanced stage of the tumor, the oncology team recommended palliative care. Afterwards, the patient developed progressive right-sided heart failure and died after a 53-day stay in the intensive care unit (ICU).

DISCUSSION and CONCLUSIONS

Primary cardiac angiosarcoma is an extremely rare cardiac tumor (0.001%–0.03%), most commonly diagnosed between the third and fifth decades of life. Primary angiosarcoma mainly occurs with unclear etiologies, but secondary angiosarcoma could be attributed to radiotherapy, lymphedema, environmental carcinogens, or certain familial syndromes.^{1,2} Although it can originate from any part of the heart, it typically develops in the right atrium (90%) with invasion of adjacent structures.³ Early diagnosis of cardiac angiosarcoma is still deceptive since it almost remains asymptomatic until it has grown to a specific size or location or metastases occur.^{4,5} The clinical course is regrettably rapidly progressive to death shortly after diagnosis. Presenting signs and symptoms of cardiac angiosarcoma are nonspecific and may include right-sided heart failure, symptoms of pericardial involvement, pulmonary emboli, and arrhythmia.^{3,6-8} In countries where tuberculosis is endemic, clinical presentation of the disease may be misdiagnosed.⁵ Although the diagnosis of the tumor is suspected on imaging-including transthoracic echocardiography, CT scanning, and CMR-definitive diagnosis of angiosarcoma is based on the histopathologic and immunohistochemical findings. Characteristic pathologic findings of angiosarcoma are solid nodules consisting of spindle-shaped tumor cells. Immunohistochemistry might reveal positively stained blood vessel-related markers such as CD31 and CD34.^{9,10}

Blunt cardiac injury is more common than we expect. Clinical features of cardiac injury secondary to nonpenetrating trauma are various and can be lethal if left undiagnosed. Cardiac rupture is the most catastrophic complication of blunt trauma, with a high mortality rate.¹¹ The structures most frequently involved in cardiac rupture after nonpenetrating trauma are the right chambers (right atrium more than right

ventricle).¹² Injuries to tricuspid chordae and papillary muscles occur during blunt trauma and can lead to tricuspid regurgitation.¹³

In our case, the initial diagnosis due to rupture of the right atrium and pericardial effusion as well as pulmonary involvement raised the possibility of right atrial angiosarcoma. On the other hand, the radiologic findings of pulmonary involvement with pulmonary symptoms in the COVID-19 pandemic era misled the diagnosis; in the meantime, a rupture in the right atrium during trauma is an innocent association with cardiac angiosarcoma.

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Conflict of Interest: None

Ethical Statement

The authors attest that the study complies with the guidelines of the institution's human studies committees and the United States Food and Drug Administration. Informed consent for the publication of research was obtained from the patient's family.

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