

Primary cardiac angiosarcoma with right atrial wall rupture

A case report

Jinah Kim, MD^a, Bo Da Nam, MD^a, Jung Hwa Hwang, MD, PhD^{a,*}, Soo Bin Park, MD^a, Min-Ho Lee, MD^b, Dong Won Kim, MD, PhD^c, Young Woo Park, MD, PhD^d, Min-Young Lee, MD^e

Abstract

Rationale: Cardiac angiosarcoma is the most common malignant tumor of the heart and a rare disease with rapid disease progression and poor prognosis. Cardiac wall rupture is an extremely rare complication.

Patient concerns: A 32-year-old woman presented with an acute onset of epigastric pain and chest discomfort at first time when she visited an emergency room.

Diagnoses: A cardiac mass was identified on echocardiography and subsequently performed chest computed tomography and cardiac magnetic resonance imaging revealed the cardiac tumor at right atrium with right atrial wall rupture and hematogenous lung metastasis. Histopathologic diagnosis of metastatic angiosarcoma was done by open lung biopsy.

Interventions: The patient was treated with palliative chemotherapy for the primary cardiac tumor and hematogenous lung metastasis.

Outcomes: The follow-up imaging studies revealed treatment response of the primary cardiac tumor and hematogenous lung metastasis.

Lessons: Clinical and radiologic evaluation of the cardiac angiosarcoma was well performed in our case with various diagnostic imaging modalities including echocardiography, chest computed tomography, cardiac magnetic resonance imaging, and fluorodeoxyglucose-positron emission tomography/computed tomography. This case report well demonstrates typical imaging findings of a rare cardiac tumor and emphasizes importance of early investigation for accurate diagnosis and proper management of the cardiac tumor.

Abbreviations: CT = computed tomography, ER = emergency room, FDG = fluorodeoxyglucose, IVC = inferior vena cava, MRI = magnetic resonance imaging, PET-CT = positron emission tomography-computed tomography, RA = right atrium, TEE = transesophageal echocardiography.

Keywords: angiosarcoma, heart, magnetic resonance imaging, rupture

1. Introduction

Primary cardiac malignancies are not common and account for 25% of cardiac tumors. Sarcomas are the most common primary

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^a Department of Radiology, ^b Department of Cardiology, ^c Department of Pathology, ^d Department of Cardiothoracic Surgery, ^e Department of Oncology and Hematology, Soonchunhyang University Seoul Hospital, Soonchunhyang University Seoul Hospital, Soonchunhyang University College of Medicine, Seoul, Republic of Korea.

* Correspondence: Jung Hwa Hwang, Department of Radiology, Soonchunhyang University Seoul Hospital, 59, Daesagwan-ro, Yongsan-gu, Seoul 04401, Republic of Korea (e-mail: jhhwang@schmc.ac.kr).

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cardiac malignancies and angiosarcoma is the most common histologic subtype (30% of all primary cardiac malignant tumors).^[1] Cardiac angiosarcoma is a very aggressive tumor with rapid progression and poor prognosis. Metastases are often accompanied at initial diagnosis.^[2] Cardiac wall rupture is a very fatal complication of the tumor, but extremely rare. Only a few cases have been reported in the literature.^[3,4] Early diagnosis is challenging because of non-specific symptoms and rarity of the disease. However, advances in imaging modality improved early detection of cardiac tumor and can be beneficial for proper management.

We report a case of a primary cardiac angiosarcoma in a 32-year-old woman who presented with cardiac tamponade and subsequent right atrial wall rupture. The patient revealed progression of primary cardiac tumor and hematogenous lung metastasis during the follow-up with delayed evaluation and diagnosis.

2. Case report

A 32-year-old woman visited an emergency room (ER) of our institution and she presented with epigastric pain and chest discomfort. On initial laboratory findings, red blood cell and hemoglobin levels were decreased. There was leukocytosis, neutrophilia, mild elevation of C-reactive protein. Initial electrocardiography at ER revealed tachycardia with no other

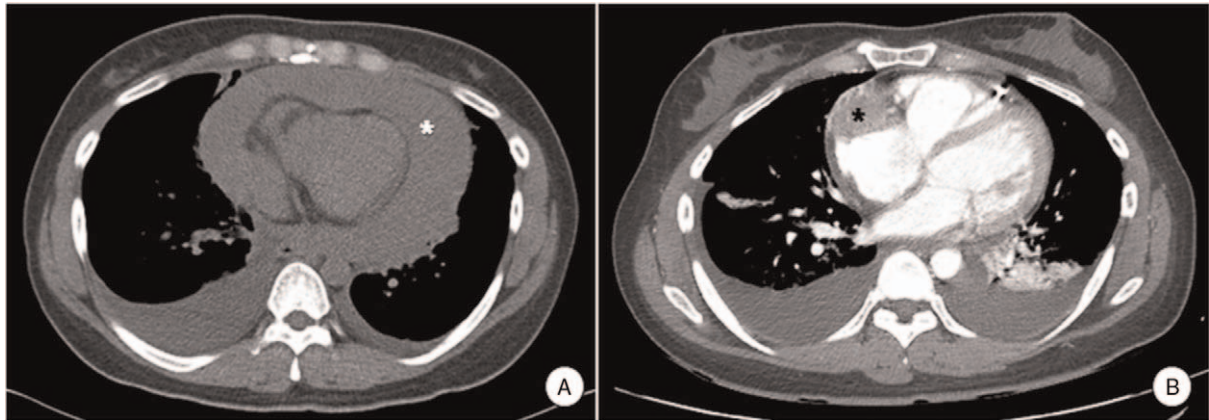


Figure 1. An initial pre-enhanced abdominal CT scan and a contrast-enhanced chest CT scan in a 32-year-old woman. (A) On initial abdominal CT scan, large amount of pericardial effusion is noted with pre-enhanced high-attenuation (40 Hounsfield unit) (white asterisk) and there are bilateral pleural effusions. (B) On contrast-enhanced chest CT scan after three days, about 3.4 cm sized a low density mass is noted in the right atrium (black asterisk). The amount of pericardial effusion is decreased with insertion of a drainage catheter.

remarkable finding. Initial chest radiography showed globular enlargement of the cardiac shadow and bilateral pleural effusions. An abdominal computed tomography (CT) scan was done for evaluation of epigastric pain and which revealed large

amount of hemopericardium (40 Hounsfield unit on pre-enhanced scan) and moderate amount of bilateral pleural effusions (Fig. 1A). Enlargement of inferior vena cava (IVC), hepatic vein, and renal vein were noted with contrast reflux in

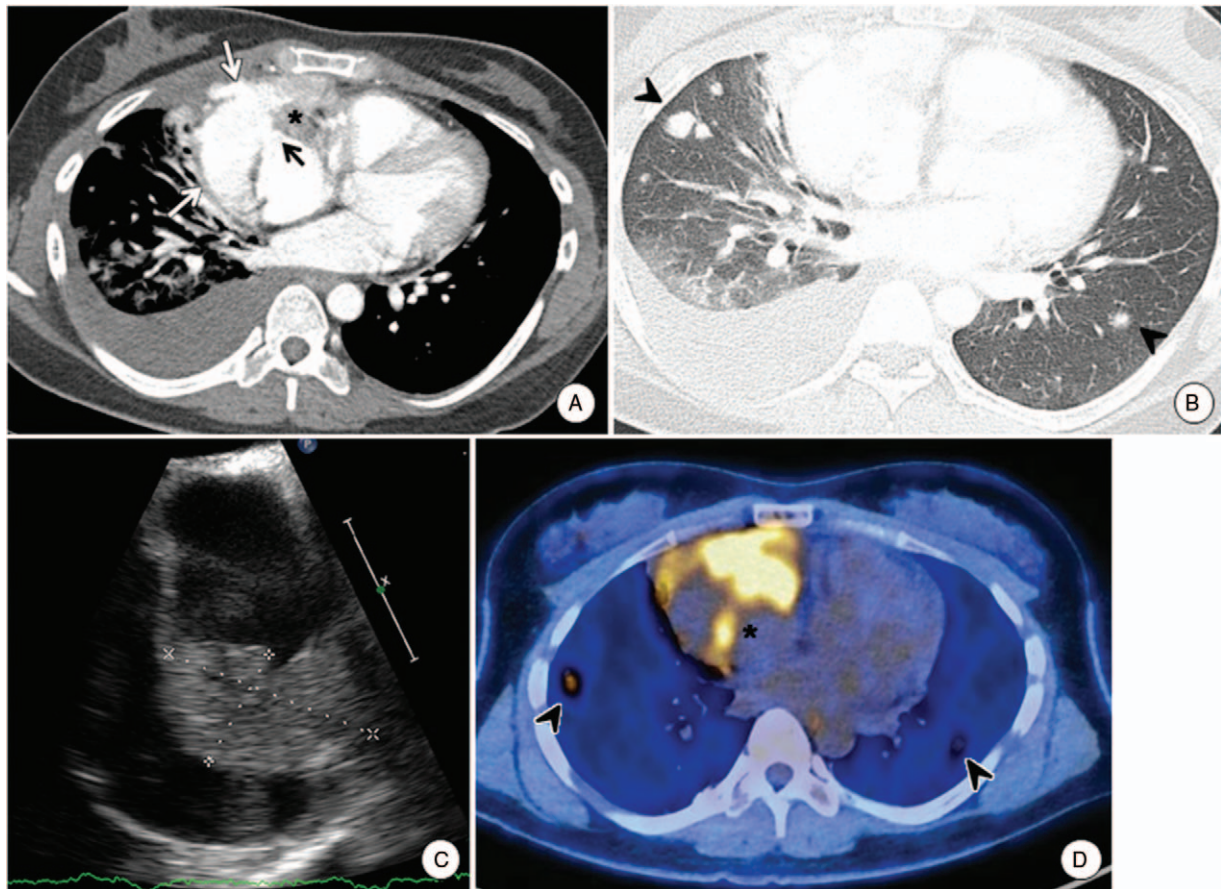


Figure 2. A follow-up contrast-enhanced chest CT scan, a TEE and a FDG-PET/CT scan in a 32-year-old woman. (A) Five months later, a follow-up chest CT scan shows about 4 cm sized a low density mass in the right atrium with invasion of atrioventricular groove and right ventricle (asterisk). There is about 8 cm sized an extraluminal contrast-filled sac on the right paracardiac area (white arrows), which is associated with focal perforation of the right atrial wall (black arrow) and moderate amount of right pleural effusion. (B) A lung window image shows multiple metastatic nodules (arrowheads) with increased size and number since initial exam. (C) On a TEE, about 4 cm sized a lobulated echogenic mass is identified in the right atrium. (D) On a FDG-PET/CT scan, hypermetabolism (maximum standardized uptake values = 15.9) is seen in the cardiac mass of the right atrium (asterisk) and also in the metastatic pulmonary nodules (arrowheads).

IVC and periportal edema, suggesting of cardiac tamponade. Echocardiographic findings were compatible with cardiac tamponade with large amount of pericardial effusion and right ventricular collapse. Pericardiocentesis was done with insertion of a drainage catheter and about 600 cc of bloody fluid was drained. Cytologic evaluation of the pericardial fluid revealed no evidence of malignancy. After three days, a contrast-enhanced chest CT scan was performed. The chest CT scan showed enhancing pericardial thickening and decreased amount of pericardial effusion with catheter drainage. The amount of bilateral pleural effusions was slightly increased during the follow-up period. There was a poorly defined low density focal lesion in the right atrium (RA) (Fig. 1B). A few millimeter-sized well-defined lung nodules were identified and which were presumed possible hematogenous metastasis (not shown). Differential diagnoses for the lesion in the RA included a thrombus, incomplete contrast blending, and a cardiac tumor, and then further diagnostic work-up was recommended. However, the patient refused further evaluation.

Five months later, the patient revisited ER with acute chest pain and her vital signs were stable. A follow-up chest radiography showed large amount of right pleural effusion and newly appeared bulging contour on right cardiac margin. On follow-up chest CT scan, there was persistently noted and enlarged about 4 cm sized an irregular low density mass in the RA with invasion to atrioventricular groove and right ventricle. Focal perforation of the RA wall was identified and which communicated with an about 8 cm sized encapsulated extraluminal contrast-filled sac on the right paracardiac area (Fig. 2A). Small amount of pericardial effusion was also seen. Lung window images showed increased number and size of multiple nodules with peripheral ground-glass

halo, representing progression of hemorrhagic metastasis (Fig. 2B). Radiologic diagnosis of a primary cardiac tumor, particularly angiosarcoma, was suggested. A trans-esophageal echocardiography (TEE) revealed about 4 cm sized a mass in the RA (Fig. 2C). A fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scan was performed for systemic evaluation and which showed hypermetabolism (maximum standardized uptake values=15.9) in the cardiac mass and metastatic lung nodules (Fig. 2D). A gadolinium-enhanced cardiac magnetic resonance imaging (MRI) was undergone. About 4 cm sized a lobulated mass was seen at the right lateral wall of RA near the atrioventricular groove and which showed high signal intensity on T2-weighted image with heterogeneous delayed enhancement (Fig. 3A–B). And there was revealed focal rupture with presumed a pseudoaneurysm on the right lateral wall of RA (Fig. 3C–D).

After review of risks for open surgery with biopsy of cardiac mass, the physician planned a lung biopsy. A surgical biopsy with wedge resection of the lung nodules in left upper lobe was performed by video-assisted thoracoscopic surgery. The gross surgical specimen revealed multiple hemorrhagic nodules (Fig. 4A) and microscopically, the tumor was composed of atypical endothelial cells with poorly formed vascular channels (Fig. 4B–C). On immunohistochemistry, the tumor cells revealed positive for CD34 and CD31 (Fig. 4D). Histopathologic diagnosis of metastatic angiosarcoma was confirmed. The patient underwent chemotherapy with paclitaxel and follow-up evaluation with chest CT and cardiac MRI was performed after two months. On chest CT and cardiac MRI scans, the size of cardiac mass was slightly decreased from 4 to 3.5 cm and metastatic lung nodules were much decreased in size and number (not shown).

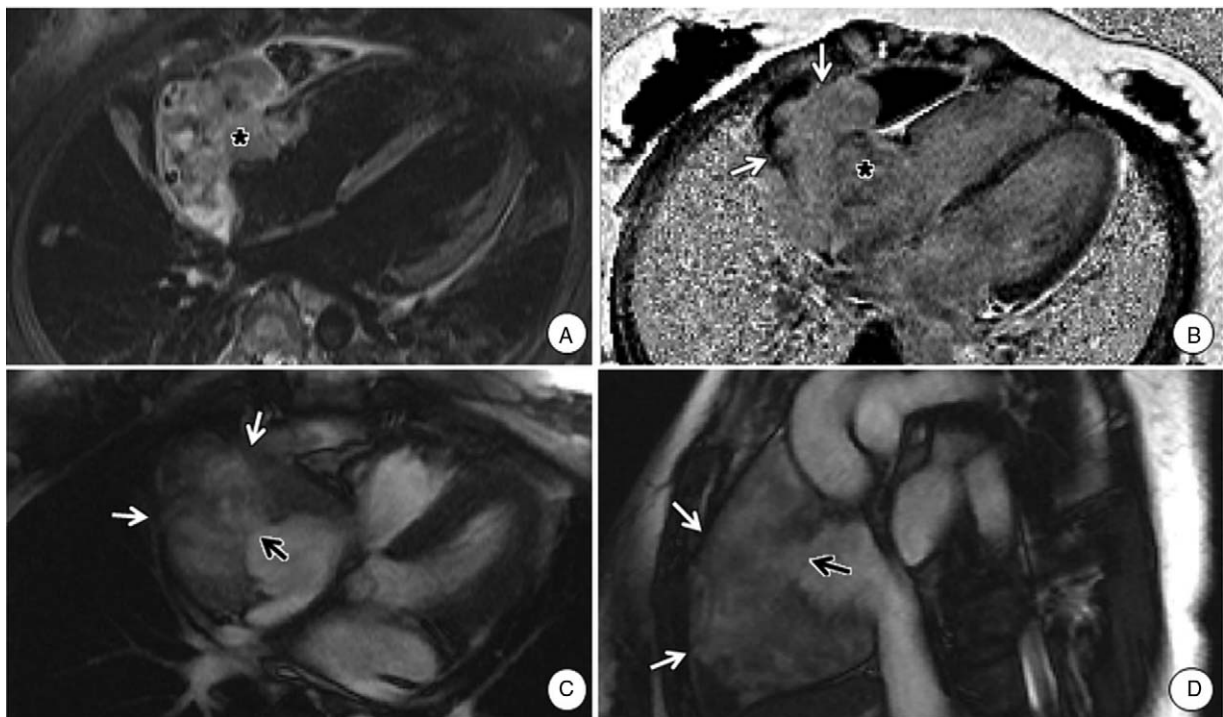


Figure 3. A gadolinium-enhanced cardiac MRI in a 32-year-old woman. (A) On black blood four chamber view, about 4 cm sized a lobulated and heterogeneous high SI mass (asterisk) is seen at the lateral wall of RA near the atrioventricular groove. (B) On ten minutes delayed enhanced image with phase-sensitive inversion recovery, the mass shows heterogeneous enhancement (asterisk) and an extraluminal sac, presumed a pseudoaneurysm, on the right paracardiac area is seen (white arrows). (C–D) On four chamber and short axis CINE images, there is seen a focal defect on the right lateral wall of RA (black arrow) with presumed a pseudoaneurysm (white arrows).

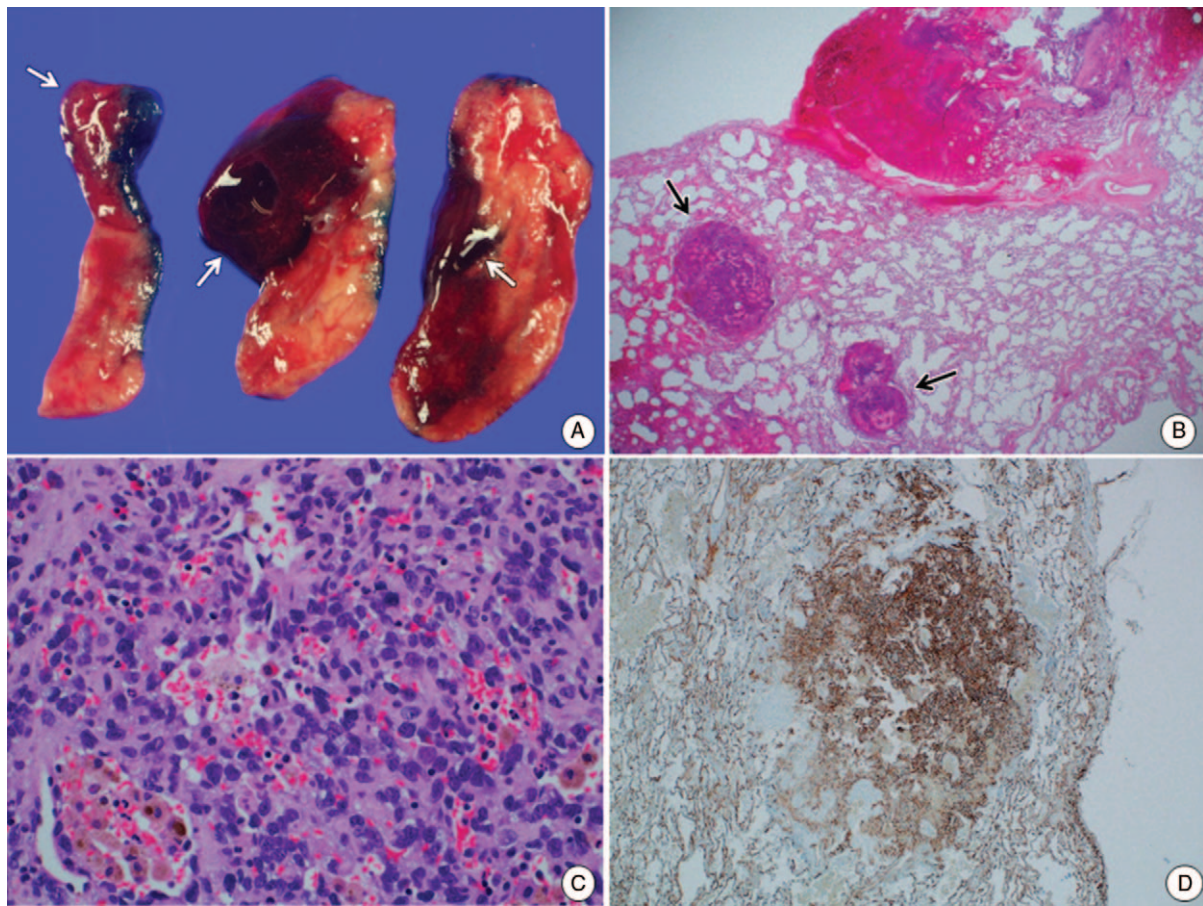


Figure 4. Pathologic findings of the specimen obtained from wedge resection of the lung. (A–B) On gross specimen and microscopy (x12.5, H and E), there are multiple hemorrhage nodules in resected lung (arrows). (C) The tumor is composed of atypical endothelial cells with poorly formed vascular channels and shows a few extravascular erythrocytes and mitoses (x400, H and E). (D) On immunohistochemistry, the tumor cells show positive staining for CD34 (x100).

3. Discussion

A primary cardiac angiosarcoma is an endothelial cell tumor showing infiltrative growth within the surrounding myocardial wall. It usually affects males three times more often than females and tends to occur in people aged 30 to 50 years, but any age can be affected.^[2] A cardiac angiosarcoma most commonly develops in the RA with invasion of adjacent structures and frequent pericardial involvement.^[1]

A patient with a cardiac angiosarcoma is usually asymptomatic. The symptoms are non-specific and which include dyspnea, chest pain, general weakness, and weight loss. Clinical findings depend on the extent of tumor infiltration and metastasis. The most common complications are pericardial effusion and cardiac tamponade and more than half of the patients reveal pericardial effusion.^[5] Pericardial fluid is bloody and often not containing malignant cell, even if the tumor invaded the pericardium.^[2] Then the cytologic result of pericardiocentesis is not sufficient to exclude malignancy. Our patient also had large amount of pericardial effusion and cardiac tamponade at initial presentation and the pericardial fluid revealed negative for malignancy on cytologic review. Necrosis of the myocardial wall can cause cardiac rupture. However, cardiac rupture related to angiosarcoma is extremely rare and only a few cases have been reported in the literature.^[3,4] It is a very fatal complication that can lead to sudden cardiac death. The patient of our case showed rupture of

the right cardiac wall caused by direct tumor invasion at her second hospital visit and she survived fortunately.

Early diagnosis of primary cardiac angiosarcoma is difficult due to disease rarity and non-specific clinical symptom. Delayed diagnosis can lead to a devastating effect on the patient's prognosis. Therefore, radiologists with various imaging modalities should play an important role in the early diagnosis, especially when a cardiac tumor is clinically suspected. Echocardiography is the main diagnostic tool for cardiac tumors. TEE has high sensitivity (up to 97%) in detecting cardiac masses.^[2] Transthoracic echocardiography is useful for evaluation of the location, size, shape, and mobility of a cardiac tumor.^[1] A contrast-enhanced CT can provide additional information about a cardiac tumor. On CT, a cardiac angiosarcoma is seen as a heterogeneous enhancing mass with infiltration of myocardium and pericardium, which is frequently associated with pericardial effusion and/or thickening.^[6] If there is hematogenous lung metastasis, common CT findings are multiple solid nodules or thin-walled cysts with peripheral ground-glass attenuated halo due to peritumoral hemorrhage.^[7] However, the CT assessment of a cardiac tumor is somewhat limited. Because of high-attenuated contrast pool in the cardiac cavity, an enhancing cardiac mass can be misinterpreted as a thrombus or can be neglected as a pseudo mass due to incomplete contrast blending. Frequent motion artifact related to cardiac

pulsation can be another diagnostic pitfall. Then, if there were suspected a cardiac mass on CT scan, further diagnostic work-ups have to be performed. Cardiac MRI is more preferable for evaluation of a cardiac tumor with better soft tissue characterization than CT scan. It can help distinguishing intracardiac thrombi from tumors.^[2] A thrombus is mostly found in the left atrium and can show variable signal intensity depending on the age of a thrombus.^[8] In a cardiac angiosarcoma, the tumor shows heterogeneous signal intensity on T1- and T2-weighted images due to necrosis and hemorrhage. Arterial enhancement of a tumor is shown at first-pass perfusion imaging and it suggests rich vascularity. Also heterogeneous enhancement is seen at late gadolinium enhancement imaging with peripheral enhancement and central hypointensity due to necrosis.^[8] Cardiac MR also provides detailed information about a cardiac mass including tissue composition and its relationship to adjacent structures.

In our case, the patient presented typical clinical manifestation of angiosarcoma with hemopericardium and cardiac tamponade. The patient revealed subsequent RA wall rupture caused by direct tumor invasion and which is a very rare and fatal complication. A cardiac angiosarcoma and associated complications are well evaluated in our case with various imaging modalities including echocardiography, CT, MRI, and also FDG-PET/CT scan. Based on the imaging findings, proper diagnosis and therapeutic plans can be made. We emphasize that a primary cardiac malignancy should be included in one of the important differential diagnoses when a young patient presented with an unknown cardiac tamponade, even if the cytologic evaluation of the pericardial fluid reveals negative result. Meticulous further diagnostic work-up using various imaging modalities is imperative.

Author contributions

Conceptualization: Jinah Kim, Bo Da Nam, Jung Hwa Hwang.

Investigation: Jinah Kim, Bo Da Nam, Jung Hwa Hwang, Soo Bin Park, Min-Ho Lee, Dong Won Kim, Young Woo Park, Min-Young Lee.

Supervision: Bo Da Nam, Jung Hwa Hwang.

Writing – original draft: Jinah Kim.

Writing – review & editing: Jinah Kim, Bo Da Nam, Jung Hwa Hwang.

References

- [1] Patel SD, Peterson A, Bartczak A, et al. Primary cardiac angiosarcoma – a review. *Med Sci Monit* 2014;20:103–9.
- [2] Riles E, Gupta S, Wang DD, et al. Primary cardiac angiosarcoma: a diagnostic challenge in a young man with recurrent pericardial effusions. *Exp Clin Cardiol* 2012;17:39–42.
- [3] Jain G, Mukhopadhyay S, Kurien S, et al. Ruptured cardiac angiosarcoma with pulmonary metastases: a rare disease with a common (mis) diagnosis!. *Indian Heart J* 2012;64:603–6.
- [4] Tang K, Shang QL, Zhou QC, et al. Primary cardiac angiosarcoma with spontaneous ruptures of the right atrium and right coronary artery. *Echocardiography* 2013;30:E156–160.
- [5] Look Hong NJ, Pandalai PK, Hornick JL, et al. Cardiac angiosarcoma management and outcomes: 20-year single-institution experience. *Ann Surg Oncol* 2012;19:2707–15.
- [6] Araoz PA, Eklund HE, Welch TJ, et al. CT and MR imaging of primary cardiac malignancies. *Radiographics* 1999;19:1421–34.
- [7] Tateishi U, Hasegawa T, Kusumoto M, et al. Metastatic angiosarcoma of the lung: spectrum of CT findings. *Am J Roentgenol* 2003;180:1671–4.
- [8] Motwani M, Kidambi A, Herzog BA, et al. MR imaging of cardiac tumors and masses: a review of methods and clinical applications. *Radiology* 2013;268:26–43.