

[CASE REPORT]

Primary Cardiac Angiosarcoma Accompanying Cardiac Tamponade

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Abstract:

A *de novo* cardiac malignant tumor is rare and sometimes challenging to diagnose. We encountered a 67-year-old man without any medical history complaining of dyspnea on effort. On admission, his hemodynamics were deteriorated due to cardiac tamponade, which was improved by percutaneous drainage of 1,200 mL pericardial effusion, showing 11.0 g/dL of hemoglobin. We suspected primary cardiac malignancy following multidisciplinary tests, and a cardiac biopsy via sternotomy demonstrated the definitive diagnosis of primary malignant tumor (angiosarcoma) infiltrating the right atrial myocardium. We initiated weekly paclitaxel therapy. Further studies are warranted to establish the optimal diagnostic and therapeutic strategy for *de novo* cardiac malignancy.

Key words: heart failure, pericardial effusion, hemodynamics, cardiogenic shock

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Introduction

Primary cardiac tumor is a rare disease (1), with 80% of cases being benign. The most well-known benign cardiac tumor is myxoma, which is often observed in the left atrium and sometimes affects hemodynamics (2). Prompt resection is required, but the post-operative course is quite good, with an operative mortality rate under 5%.

In contrast, 20% of primary cardiac tumors are malignant (3). Of them, sarcomas are one of the predominant lesions. There is no established therapeutic strategy for such tumors, and the median survival is only 6 to 12 months even with complete resection (4).

We encountered a patient presenting with cardiac tamponade due to pericardial effusion that was eventually diagnosed as a rare primary cardiac malignant tumor (angiosarcoma) infiltrating the right atrial myocardium.

Case Report

On admission

A 67-year-old man without any obvious medical history was admitted to our institute complaining of palpitation and dyspnea on effort for 2 weeks. He had received no medications. His body height was 162 cm, and his body weight was 66 kg. His blood pressure was 130/68 mmHg, heart rate was 85 bpm, and oxygen saturation was 99% on nasal 3 L/min oxygen support.

Chest X-ray showed cardiomegaly with a cardiothoracic ratio of 0.65 and bilateral pulmonary congestion (Fig. 1A). An electrocardiogram showed a heart rate of 82 bpm with sinus rhythm and no ST-changes (Fig. 1B). The plasma B-type natriuretic peptide level was 56 pg/mL. Transthoracic echocardiography showed a moderate amount of pericardial effusion (estimated volume was 1,200 mL) without right heart collapse (Fig. 1C, D). Chest computed tomography showed bilateral mild pleural effusion and circumference pericardial effusion with an increased density (45 Hounsfield

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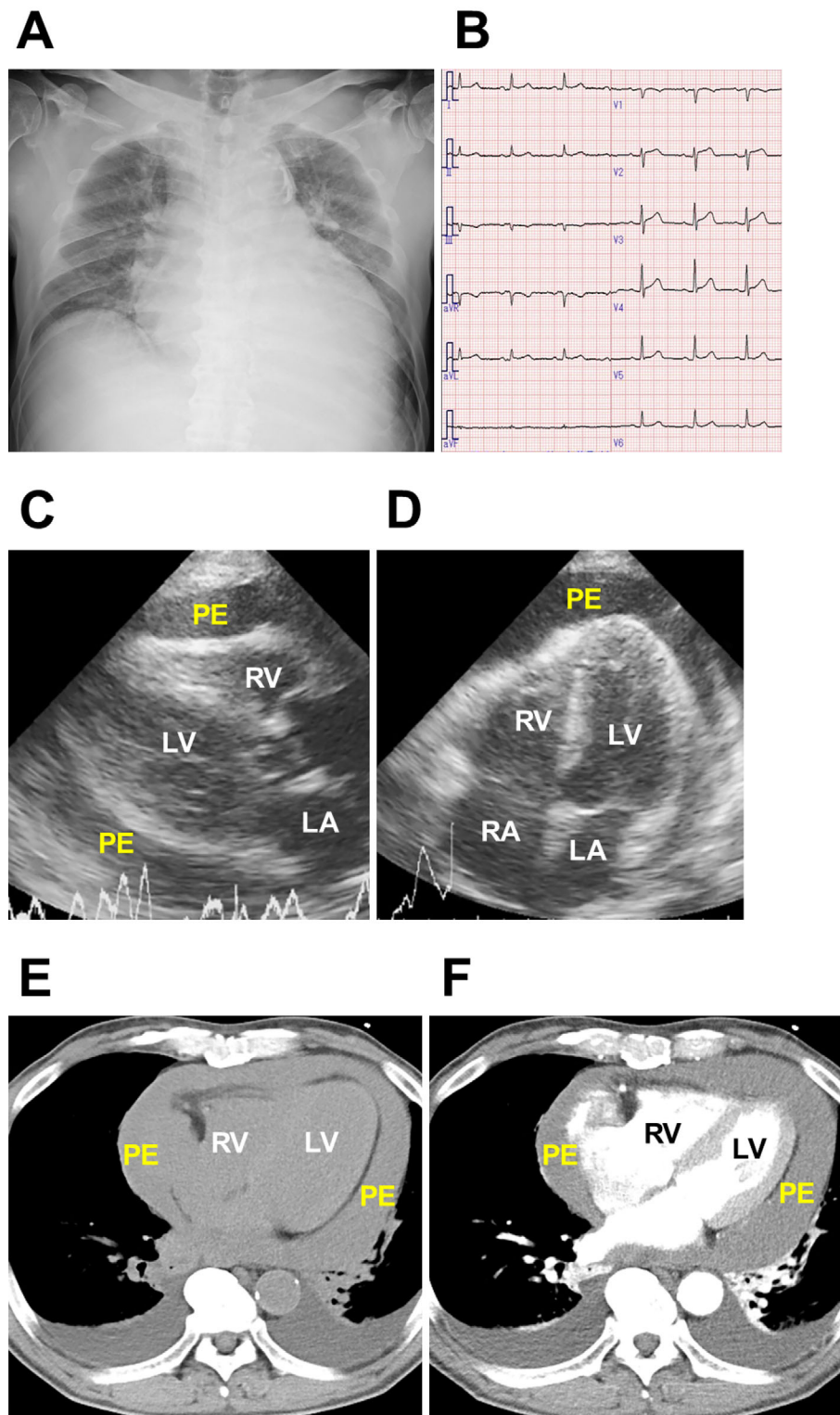


Figure 1. Chest X-ray (A), electrocardiogram (B), transthoracic echocardiography showing circumference pericardial effusion, obtained in the long-axis view (C) and four-chamber view (D), and chest computed tomography showing circumference pericardial effusion without perforation of myocardium, obtained as a plane procedure (E) and as an enhanced procedure (F). PE: pericardial effusion, LV: left ventricle, LA: left atrium, RV: right ventricle, RA: right atrium

unit), indicating bloody fluid (Fig. 1E). Enhanced computed tomography denied active perforation of the myocardium (Fig. 1F).

In-hospital course

On day 2, cardiogenic shock accompanying pulmonary congestion developed, triggered by the occurrence of atrial

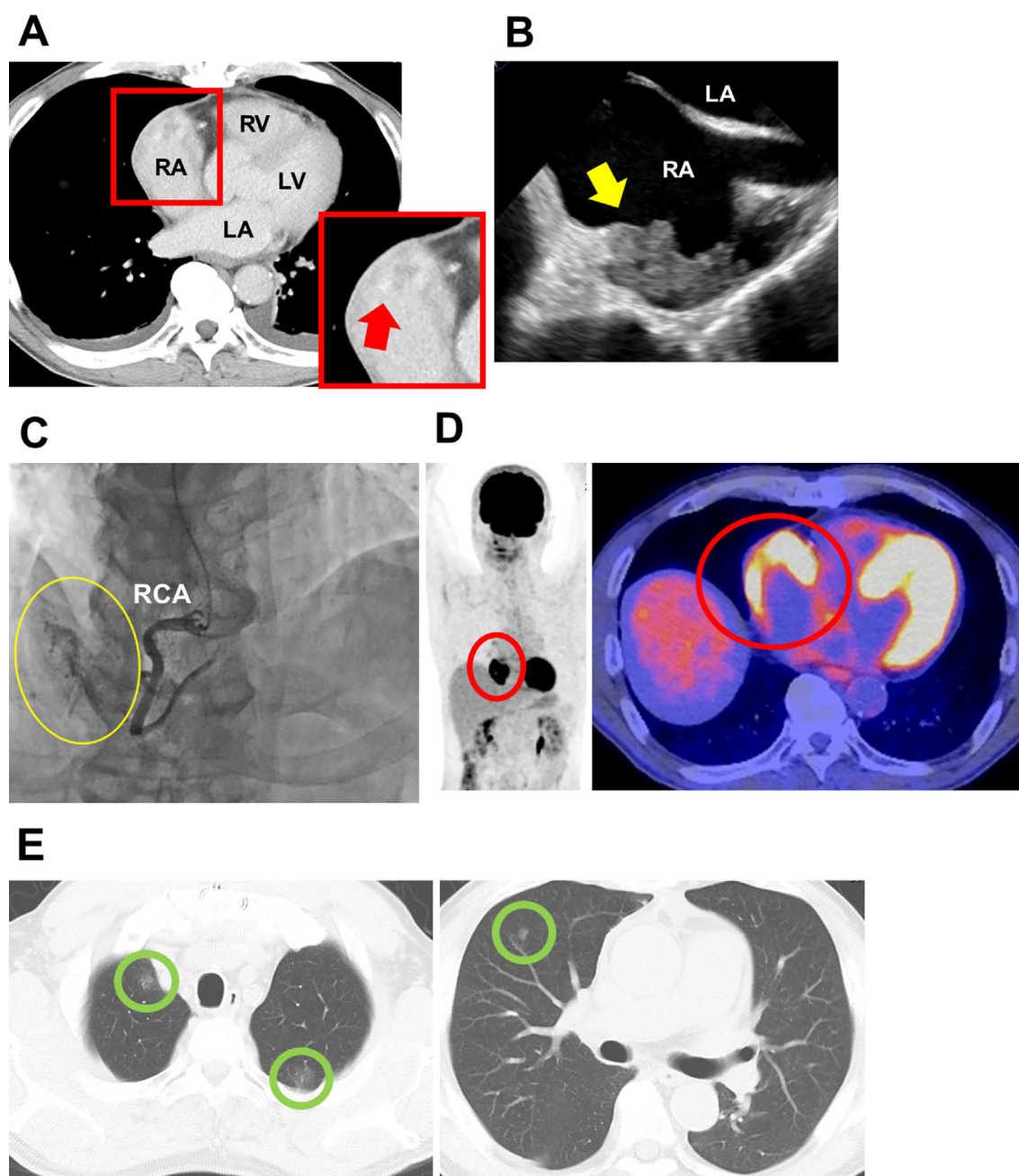


Figure 2. Detailed assessments of the cardiac tumor using cardiac computed tomography (A), trans-esophageal echocardiography (B), coronary angiography (C), positron emission tomography (D), and multi-slice chest computed tomography. Heterogeneous deficiency (red arrow) in the right atrium (A); a heterogeneous mass (yellow arrow) with an irregular border in the right atrium (B); collateral arteries from right coronary artery (yellow circle; C); strong staining at the free wall of the right atrium (red arrow; D); multiple ground-glass appearances in the bilateral lungs (green circles; E).

fibrillation and 120 bpm of tachycardia. We diagnosed him with cardiac tamponade and performed percutaneous drainage of pericardial effusion, resulting in the immediate stabilization of his hemodynamics. One liter of bloody fluid with 11.0 g/dL of hemoglobin was obtained. We suspected primary or metastasis of malignancy.

Results of a detailed assessment of malignancy

Cytology of the obtained pericardial fluid showed class II

findings. Cultures of bacterium and acid-fast bacillus were negative. Cardiac contrast-enhanced computed tomography revealed heterogeneous contrast with non-enhancement lesion in the right atrium (Fig. 2A). Transesophageal echocardiography showed a sessile nodule 53×26 mm in size in the right atrium (Fig. 2B). Coronary angiography showed collateral circulation from the right coronary artery to the suspected lesion in the right atrium (Fig. 2C). Positron emission tomography showed an abnormal accumulation in the

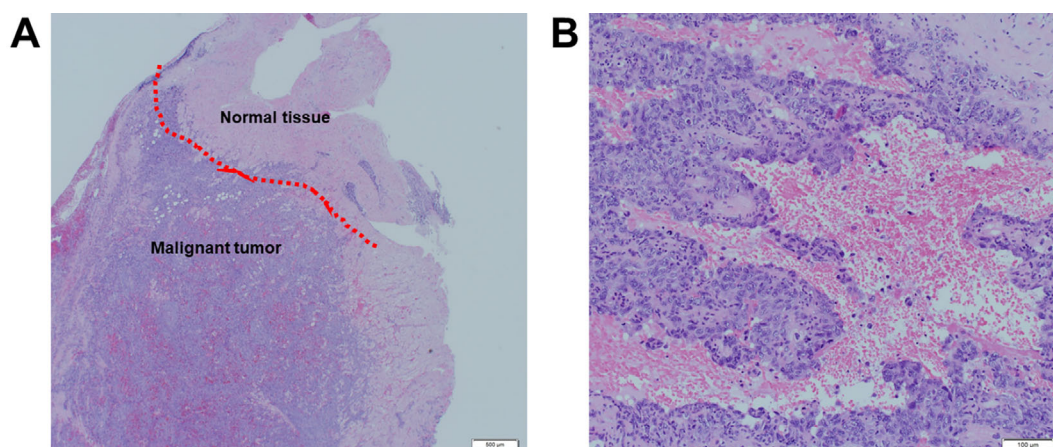


Figure 3. Histopathological image of the cardiac tumor obtained by a surgical biopsy that was diagnosed as cardiac angiosarcoma [$\times 4$ with Hematoxylin and Eosin (H&E) staining (A) and $\times 10$ with H&E staining (B)].

right atrium (Fig. 2D). Multi-slice chest computed tomography identified multiple ground-glass appearances in the bilateral lungs (Fig. 2E).

On day 57, we performed a biopsy of the cardiac tumor via surgical sternotomy and diagnosed the patient with angiosarcoma (CD31, CD34, D2-40, and Factor VIII positive) (Fig. 3). Given that his Eastern cooperative oncology group performance status scale was 1 and he was suspected of having lung metastasis, we initiated weekly paclitaxel therapy on day 70.

Discussion

The diagnosis of cardiac tumor

In general, primary cardiac tumors are easy to distinguish by standard surveillance using trans-thoracic echocardiography. For example, a benign myxoma can be detected as a large nodule typically located in the left atrium (2). However, some malignant tumors are challenging to detect by standard echocardiography, as we experienced, probably because they are infiltrating the myocardium (4). Furthermore, many primary cardiac tumors are asymptomatic until hemodynamic changes develop, as in our patient.

Cardiac computed tomography and transesophageal echocardiography were useful for locating the abnormal mass on the anterior right atrial wall, which was missed on trans-thoracic echocardiography. Coronary angiography and positron emission tomography were also helpful for determining the spread of the tumor.

For the definitive diagnosis, a histopathological assessment is essential. We declined to perform a percutaneous biopsy given the risk of right atrial wall perforation and tumor embolization (5). Instead, we performed a tissue biopsy during open cardiac surgery, a viable alternative. Complete resection is recommended, but we preferred to perform a tissue biopsy via sternotomy given the apparent tumor invasion in the right atrial myocardium and suspected lung metastasis.

sis.

Angiosarcoma

Angiosarcoma is an extremely rare primary cardiac tumor that often accompanies bloody pericardial effusion, as in our patient (4). Achieving a definitive diagnosis while the patient is alive is sometimes challenging due to its rapid and fatal proliferation and widespread myocardial infiltration, as discussed above.

Several perioperative chemotherapy has been proposed, but most of them are case reports or retrospective reviews, and there have been no randomized control trials (6). The median overall survival is 12 months (7). The survival in patients with complete resection is 17 months, whereas that in patients with partial resection is 6 months. Further studies are warranted to establish optimal diagnostic and therapeutic strategies for cardiac malignant tumors, including angiosarcoma.

The authors state that they have no Conflict of Interest (COI).

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