Spontaneous Rupture and Thrombosis of Right Atrium

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Primary cardiac tumors are extremely rare entities. One-fourth of them is malignant, with sarcoma accounting for 75% of malignant cases. While most angiosarcomas originate from head and neck, only a few arise from the heart. We reported an uncommon case of cardiac angiosarcoma presenting as right atrium rupture.

A 78-year-old female presented to our hospital in December 2015 with progressive chest distress for more than one month. She had an early history of meningioma treated with gamma knife. The patient had been previously admitted to our hospital one month ago in November 2015 with chest distress, abdominal distension, and edema in lower extremities. Transthoracic echocardiography at that time revealed moderate pericardial effusion without apparent signs of tamponade or systolic dysfunction (left ventricular ejection fraction: 65%). Computed tomography (CT) of the chest showed pleural and pericardial effusion. The 18F-fluorodeoxyglucose (FDG) positron emission tomography CT revealed pericardium thickening, right atrium enlargement, a strip of high FDG signal in pericardial fat tissue, and multiple enlarged mediastinal lymph nodes with moderate FDG signal [Figure 1a and 1b]. Pericardiocentesis was performed and pericardial and pleural fluid analysis did not show elevation in tumor markers or neutrophil counts. After a few day's pericardial drainage, the patient's symptoms relieved and she was discharged with oral diuretics.

On readmission in December 2015, the patient complained recurrent chest distress and abdominal distension, suggesting relapse of pericardial effusion. Her serum level of cancer antigen (CA)-125 was 85.8 U/ml (normal range: 0–35.0 U/ml), while other tumor markers, for example, carcinoembryonic antigen and CA-153, were within normal range. Echocardiography showed massive pleural and pericardial effusion and multiple low-attenuation thrombi in the right atrium. Enhanced mediastinum CT revealed abnormal density in the anterior right atrium and regional contrast leakage into pericardial space [Figure 1c]. Contrast echocardiography with SonoVue® showed enhancement of the right atrium wall and intracardiac mass, indicating granulomatosis or cardiac angiosarcoma.

The patient underwent pericardiectomy on January 14, 2016. During the operation, the heart showed extensive exudations, and the pericardium was locally adherent to the heart. An invading mass on the right atrium was found, which resulted in local atrium rupture [Figure 1d]. Subpericardial mass and exudation tissue were resected and sent for frozen biopsy, which suggested clotting and fibrinous exudation. The right atrium was repaired and a tube for subxiphoid pericardial window drainage was placed. The patient was referred to Surgical Intensive Care Unit after operation. However, due to deteriorating cardiac function, the patient died two days later.

Postoperative pathology revealed grayish, white, and dark-red tissue, with massive fibrinous exudation and necrosis. Hematoxylin and eosin stain showed papillary tumor tissue comprised of malignant cells with marked pleomorphism [Figure 1e]. Immunohistochemistry (IHC) staining showed CD31 (+), F8-R-Ag (+), Fli-1 (+), Ki-67 (high), CK5/6 (−), calretinin (−), Meso-cell (−), EMA (−), desmin (−), CK (pan) (−), and D2-40 (−), consistent with molecular features of angiosarcoma [Figure 1f and 1g].

Primary cardiac tumors are extremely rare, most of which are benign. Of the malignant cardiac tumors, sarcoma constituted 75% of cases. Diagnosing a cardiac angiosarcoma is typically challenging because of its rarity and because tumors originating from the upper right atrium are always covered by pulmonary tissue or fat pad, rendering them less detectable by transthoracic echocardiography. In our case, the tumor had infiltrated into both right atrium and pericardium, resulting in atrium rupture that had been covered by thrombus and inflammatory fibrous tissues. Such capsule structure

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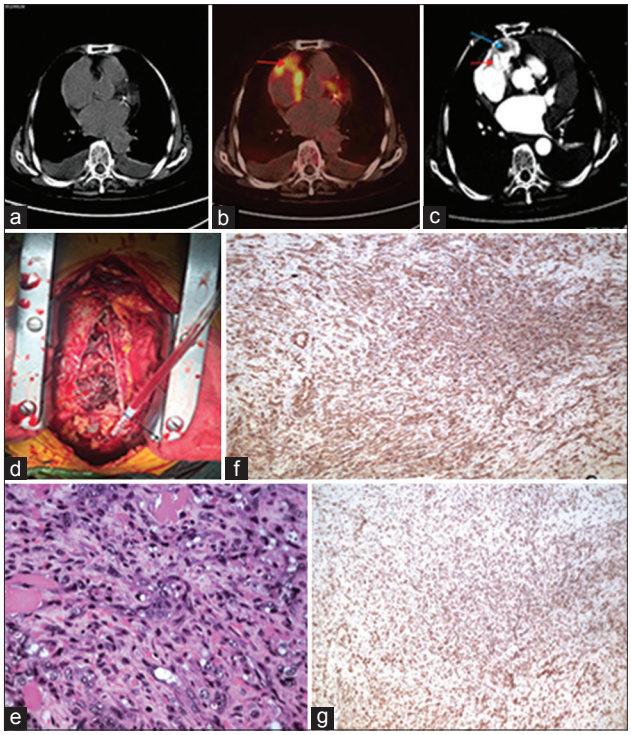

Figure 1: Cardiac angiosarcoma resulting in right atrium rupture. (a and b) PET-CT showed thickened pericardium with small pericardial effusions and enlargement of the right atrium with a strip of high FDG signals surrounding it (arrow). (c) Contrast-enhanced CT revealed contrast exudation from the right atrium into the pericardial cavity (red arrow) and a hollow cyst consisting of thrombus and inflammatory fibrous tissue (blue arrow). (d) Surgical exploration showed an invading mass in the right atrium. (e) Pathology showed papillary tumor tissue consisting of evident atypical cells (H & E staining, original magnification ×400). (f and g) The tumor was positive for CD31 (f) and FLI-1 (g) (IHC staining, original magnification ×200). PET: Positron emission tomography; CT: Computed tomography; FDG: 18F-fluorodeoxyglucose.

masked the presence of tumor, so our original diagnostic impression was atrial diverticulum or an inflammatory mass.

Most patients with primary cardiac tumors remain asymptomatic at early disease stage. Tumor may stay hidden until the occurrence of complications, including pericardial effusion, cardiac tamponade, malignant arrhythmia, and distant metastasis. The patient in our case presented with recurrent pericardial effusions, but we at first did not find evidence of more frequent causes such as tuberculosis, infections, autoimmune diseases, or metastatic tumors. However, enhanced chest CT revealed abnormal density in the right atrium and regional contrast leakage into pericardial space, which could explain pericardial effusion. Spontaneous rupture of the atrium with cardiac tumor is extremely rare and is associated with poor prognosis. Although undergoing surgical correction, patients could still die from cardiogenic shock.[1,2] Our patient remains hemodynamically stable before surgery, thanks to the formation of fibrinous thrombus that had covered the ruptured region. Definitive diagnosis of angiosarcoma is based on pathological examination and IHC. In our case, positive IHC staining of CD31 and FLI1 definitely confirmed the diagnosis of angiosarcoma.[3]

Therapy for cardiac angiosarcoma mostly is predominantly surgical resection, while the efficacy of chemotherapy and radiotherapy are very limited. Prognosis for angiosarcoma is extremely poor, with overall median survival of merely three months without surgical resection.[4] Several exceptions have been reported, with one case reporting a patient lived thirty-three months after sequential treatment with surgical resection, chemotherapy, radiotherapy, and molecular targeting therapy.[5] The patient in our case received surgical resection to alleviate her symptoms of decompensated heart failure and tamponade, as well as to seek a definitive diagnosis. Unfortunately, she did not recover from the surgery and died afterward.

As a rare cause of pericardial effusion, cardiac tumors located at the right atrium are less likely to be detected by echocardiography with standard scanning planes. In our case, only after the contrast-enhanced CT, we discovered the cardiac mass and leakage of contrast medium into the pericardial cavity. The final surgical exploration and pathological examination confirmed the diagnosis of a primary cardiac angiosarcoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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