Right Atrial Pseudoaneurysm Complicating Epithelioid Hemangioendothelioma

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ABSTRACT

Cardiac pseudoaneurysm is uncommon among young adults with trauma. Infection, prior cardiac procedure, or cardiac operations are the most common reported causes. Right atrial pseudoaneurysm (RAPA) is extremely rare. Although often challenging to diagnose, advances in noninvasive imaging have improved the ability to diagnose cardiac pseudoaneurysms. We present a case of RAPA, highlighting the diagnostic accuracy of echocardiography in this rare entity.

Key words: Cardiac pseudoaneurysm, epithelioid hemangioendothelioma, right atrial pseudoaneurysm

INTRODUCTION

Right atrial pseudoaneurysm (RAPA) is a contained rupture of the wall of the right atrium (RA) with to-and-fro blood flow into a cavity contained by pericardium, thrombus, or adhesions. A pseudoaneurysm will typically have a narrow neck, in contrast to a true aneurysm, which usually has a broad base. Cardiac epithelioid hemangioendothelioma (EHE) is an extremely rare vascular tumor.

A primary cardiac occurrence of RAPA and EHE is an extremely rare and deadly combination if not diagnosed and managed.

CASE PRESENTATION

A 44-year-old Arab female with no history of cardiovascular disease presented to the emergency department with acute onset of epigastric and retrosternal chest pain associated with vomiting. There were no other associated symptoms. Clinically, she was conscious, with blood pressure of 91/77 mmHg and pulse rate 110 beats/min. Cardiovascular examination revealed muffled heart sounds with no murmur. Her abdomen was soft with diffuse tenderness, and other systemic examinations were unremarkable. She had a past medical history of four cesarean sections, excision of fibroadenoma of the breast, right salpingectomy for ectopic pregnancy and tonsillectomy.

The 12 lead electrocardiographic (ECG) ruled out cardiac ischemia. High sensitive cardiac troponins were negative. Chest X-ray was remarkable for cardiomegaly. Transthoracic echocardiography (TTE) confirmed large pericardial effusion with echocardiographic signs of cardiac tamponade. She underwent successful pericardiocentesis under echocardiographic guidance and drained about 800 cc of serosanguinous fluid. She was investigated to identify the possible causes of the pericardial effusion, including malignancy, infectious, or connective tissue disease, but all were negative.

A repeat transthoracic echo showed normal study, and she was discharged. After 6 months, she presented with severe central and right-sided chest pain associated with shortness of breath (SOB), orthopnea and hemoptysis of acute onset. The 12 lead ECG ruled out cardiac ischemia. High sensitive cardiac troponins were negative. Transthoracic echocardiography (TTE) confirmed large pericardial effusion with echocardiographic signs of cardiac tamponade. She underwent successful pericardiocentesis under echocardiographic guidance and drained about 800 cc of serosanguinous fluid. She was investigated to identify the possible causes of the pericardial effusion, including malignancy, infectious, or connective tissue disease, but all were negative.

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Figure 1: TTE subcostal view, left: shows moderate encysted anterior pericardial effusion, and right: zoom frame with multiple clots (arrows) in the pericardial space. RV: Right ventricle, RA: Right atrium, LV: Left ventricle, LA: Left atrium

Figure 2: Agitated saline contrast injection: Left panel; starting to fill the right side, and right panel is a few seconds later image showing when contrast is cleared from the right side with some residual in the pericardial space (arrow), diagnostic of a connection between the right side and the pericardial space (pseudo-aneurysm). RV: Right ventricle, RA: Right atrium, LV: Left ventricle and LA: Left atrium

Figure 3: TEE showing right atrial pseudoaneurysm, and color flow saline into the pseudoaneurysm, left panel (arrow) and back into the RA, right panel (arrow). A clot (c) visualized in the pericardial space. RV: Right ventricle, RA: Right atrium, LV: Left ventricle and LA: Left atrium
masses [Figure 1]. Saline contrast study revealed filling of this space from the RA. The contrast persisted for some time after its clearance from the right heart cavity [Figure 2].

The transesophageal echocardiography (TEE) confirmed the same encysted pericardial effusion. The space adjacent to the RA showed effusion with multiple masses. Color flow showed multiple bidirectional flow between the RA and this space through multiple tears [Figure 3]. Agitated contrast saline injection showed well-defined multiple communications with the RA confirming the diagnoses of RAPA, with masses inside RA [Figure 4].

Computed tomography (CT) and magnetic resonance imaging of the chest confirmed the diagnosis of pseudoaneurysm of the RA that communicates freely with the RA. The pseudoaneurysm was in the form of a thin membrane containing blood with several well-formed clots, and it was communicating with the RA through a 3 cm opening. The right atrial defect was directly sutured, and biopsies were taken from the atrial wall, the pericardium, and the pleura. She had an uneventful postoperative course and discharged home after 10 days. The right atrial biopsy showed EHE with atypical cells and mitotic figures. EHE is a rare vascular tumor, classified as an intermediate malignancy that sometimes metastasizes and may recur.[1-3]

Further investigations showed liver and lung metastases, and she received multiple courses of chemotherapy and radiotherapy. One month later, she presented with nonspecific chest pain and SOB. TTE and contrast CT demonstrated the presence of pericardial hematoma with three points of communication between RA and the hematoma sac. The hematoma was extending over the right ventricular outflow tract (RVOT) with nonsignificant compression of the right ventricle.

She underwent re-expansion operation, and several points of communications were found between mass.
the RA and the hematoma from the suture line of the previous operation. Specimens were taken from multiple sites in the mediastinum, including the pericardium, and histopathology showed that all the specimens were involved by the same tumor with the same morphology EHE. However, the disease progressed rapidly, and she passed away after 9 months of the diagnosis.

**DISCUSSION**

Cardiac pseudoaneurysm is defined as a contained rupture of the myocardium with a tenuous pericardium and a fibrous tissue containing the leak. Pseudoaneurysms need to be differentiated from true aneurysms, which are focal full-thickness dilatation of the cardiac wall. The hallmark of true aneurysms is that they are bounded by all three anatomic layers, i.e., endocardium, myocardium, and pericardium. On the other hand, a pseudoaneurysm is bounded only by the pericardium, and they should be managed aggressively as they have higher risks of rupture and hemodynamic compromise. 

The most common site of cardiac pseudo-aneurysm is the left ventricle. Other cardiac areas include the mitral aortic intervalvular fibrosa, native and grafted coronary arteries, and rarely the atria. Myocardial infarction is the most common cause of cardiac pseudoaneurysm, especially pseudoaneurysm of the left ventricle. Other causes include prior valve operation or after percutaneous coronary intervention, radiofrequency ablation procedures, endocarditis, and rarely after blunt or penetrating trauma.

RAPA is very rare, and only a few cases are reported. Probably one of the reasons behind the scarcity of reports describing it is the difficulty in its diagnosis and its fatal prognosis. RAPA may occur rarely after blunt or penetrating chest trauma. It occurs in 40% of patients with chest trauma because the RA is thin walled and anteriorly located. In our case, RAPA occurred most likely secondary to the malignancy.

The patient may present with chest pain, dyspnea, or congestive heart failure. Approximately 10% of the patients are asymptomatic. Clinical examination may reveal a diastolic murmur due to the swirling motion of the blood in the pseudoaneurysm. ECG findings are nonspecific in atrial pseudoaneurysm. Chest X-ray is usually inconclusive as cardiomegaly may be the sole finding.

Diagnosis is possible by echocardiography but it is a highly operator-dependent modality and requires a high level of suspicion. The addition of agitated saline contrast injection improves the sensitivity of echocardiography for right-sided lesions, and it should be considered whenever this condition is suspected. Contrast study for the left side opacification may be utilized for suspected left-sided lesions.

In our case, the use of agitated saline contrast was diagnostic. TEE allows a much better visualization of the cardiac anatomy but has limited availability. In this reported case, contrast study confirmed the diagnosis and defined the communication between the RA and the pericardial space. Furthermore, it demonstrated accurately the multiple masses within the pericardial space. However, TEE should be used cautiously whenever malignancy is suspected in the mediastinal area. CT scan is the diagnostic modality of choice in such conditions as it offers a quick and easily available option for definitive diagnosis.

Operative repair or conservative management may be performed depending on the hemodynamic status of the patient, but eventually, operative repair is required in most patients.

Although the imaging of pseudoaneurysm in the body has been extensively described in literature, the uncommon location of the pseudoaneurysm in the RA makes our case extremely rare.

**CONCLUSION**

RAPAs are extremely rare but clinically significant lesions, which may complicate some malignant lesions. TTE is the initial investigation of choice for the diagnosis, but it requires a high level of suspicion and the use of saline contrast injection to increase its accuracy. CT scan is the modality of choice as it offers a quick and easily available option for definitive diagnosis of this entity. Emergency operative repair may be lifesaving in such cases.

**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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