

An unusual presentation of pericardial cyst: Recurrent syncope in a young patient

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Abstract

Although pericardial cysts are generally benign structures and detected incidentally, they may be associated with life-threatening complications. We present the case of a 24 year-old man with a giant hemorrhagic pericardial cyst diagnosed after evaluation for recurrent syncope which caused compression of the right ventricle. Spontaneous hemorrhage into a pericardial cyst is an extremely rare event, and to our knowledge this is the first case in which a pericardial cyst has been shown to cause recurrent syncope. (Cardiol J 2010; 17, X: xx–xx)

Key words: pericardial cyst, syncope, pericardial mass

Introduction

Syncope is a loss of consciousness due to cerebral hypoperfusion. Reflex syncope (neurally mediated syncope), syncope due to orthostatic hypotension and cardiac (cardiovascular) syncope are the three principal pathophysiological causes of syncope [1]. Reflex syncope is the common cause of syncope and it refers to a heterogeneous group of conditions such as vasovagal and carotid sinus syncope [2]. Syncope due to cardiovascular disease (CVD) is the second commonest cause and arrhythmias are the leading mechanism. Structural CVD such as aortic stenosis, hypertrophic obstructive cardiomyopathy, pulmonary emboli, pericardial effusion and tamponade can also cause syncope. However, to the best of our knowledge, this is the first case to implicate a pericardial cyst as a cause of recurrent syncope.

Case report

A 24 year-old man, without any past medical history, was admitted to our cardiology department because of recurrent syncope. He had experienced six syncope episodes in the previous three years which had occurred both when standing and when supine, without any prodromal symptoms. His medical history was unremarkable with no history of fever, chest pain, cough, weight loss or night sweats.

On physical examination, his blood pressure was 90/75 mm Hg and his heart rate was 86 bpm. There was a third heart sound on cardiac auscultation. His electrocardiography (ECG) showed sinus rhythm, a normal QT interval and non-specific ST-T changes. Postero-anterior and lateral chest X-rays were taken on suspicion of structural heart disease, and they revealed a large pericardiac mass with calcified borders (Fig. 1). Transthoracic echocardi-

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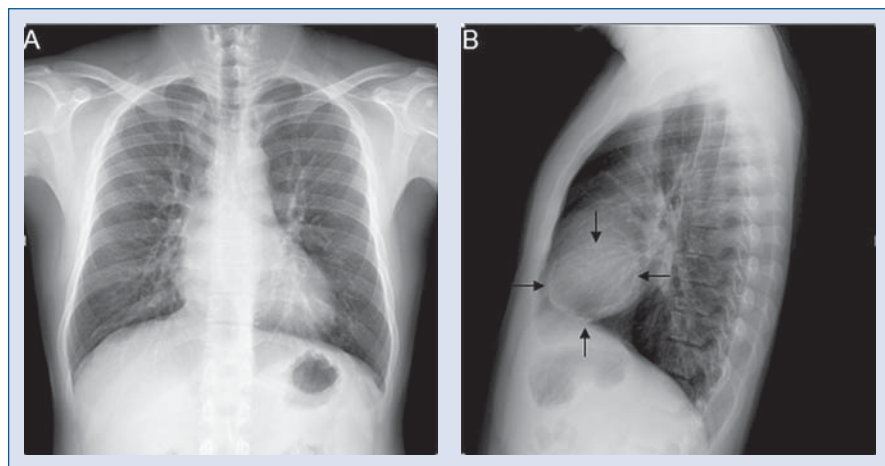


Figure 1. Postero-anterior chest X-ray (A) showed no abnormality. The lateral X-ray (B) showed a calcific pericardial mass (arrows).

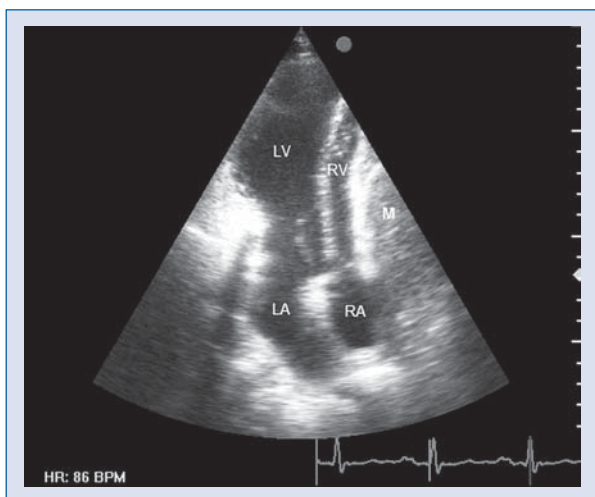


Figure 2. Apical four-chamber window of transthoracic echocardiography showing a mass (M) compressing the right ventricle (RV); RA — right atrium; LA — left atrium; LV — left ventricle.

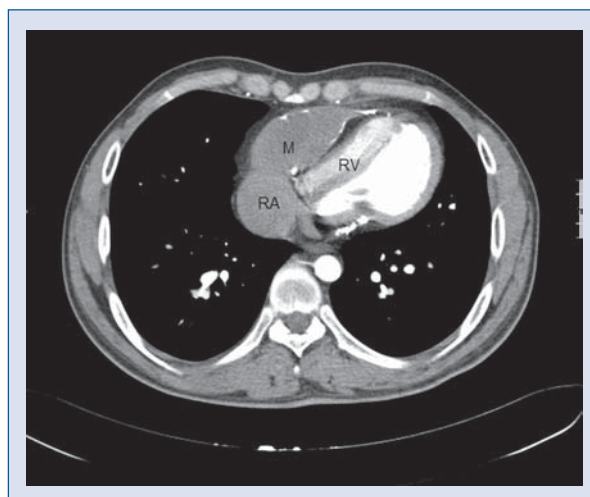


Figure 3. Computed tomography showing a calcific mass (M) compressing the right ventricle (RV); RA — right atrium.

graphy (TTE) showed marked compression of the right ventricle (RV) by an extracardiac mass (Fig. 2). TTE also revealed prominent respiratory inflow variability of the right ventricle, a dilated inferior vena cava and an exaggerated septal bounce, indicating echocardiographic signs of tamponade. A contrast-enhanced computed tomographic scan confirmed the presence of a hypodense, 62 × 60 × 36 mm in diameter, cystic lesion with a calcific border, anterior to the RV, that was causing severe compression of the ventricle (Fig. 3).

He underwent cardiac surgery in the same week due to the echocardiographic signs of tamponade.

After opening the chest cavity through a median sternotomy, a giant pericardial cyst localized posterior to the sternum was found at the right cardiophrenic angle. The outer layer of the cyst was firm because of dense fibrosis and calcification. It was firmly attached to the RV, and therefore it was excised by fragmentation of the wall. The cyst was filled with blood (Fig. 4). There was no communication of the cyst with heart chambers or vascular structures. The cystic structure was excised completely without any complications. The pathological diagnosis was a hyalinized and calcific cystic structure with non-specific chronic inflammation

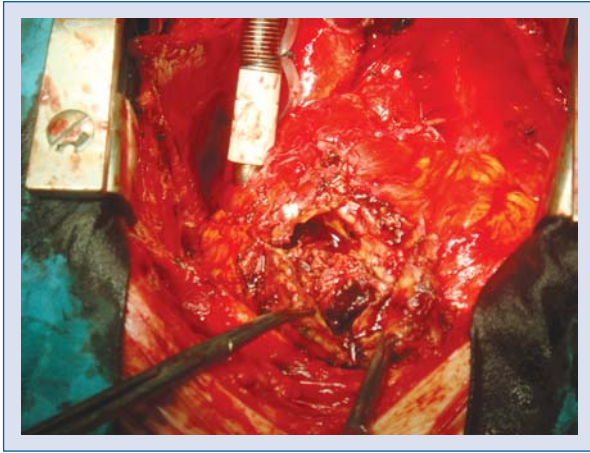


Figure 4. Intra-operative appearance of the cyst and its hemorrhagic content.

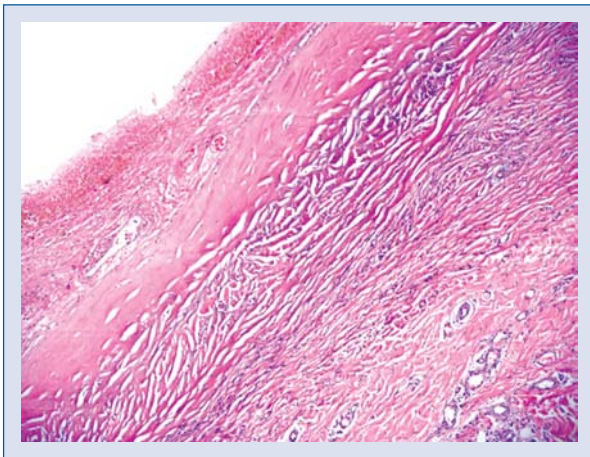


Figure 5. Pathological examination revealed a hyalinized cystic structure with fresh and old hemorrhage (H&E \times 100).

but without any lining epithelium (Fig. 5). The material in the cavity was reported as both fresh and old hemorrhage. The post-operative course was uneventful. One month later, the patient was asymptomatic, with a blood pressure of 127/75 mm Hg.

The patient had no history of trauma, rheumatic fever, tuberculosis or thoracic surgery and the cyst had no communication with any vascular structure. Therefore, a diagnosis of pseudoaneurysm or another cause was unlikely. Pathological examination excluded tuberculosis, hydatid cyst or neoplasms. We believe that a reasonable explanation of that structure is that it was a hemorrhagic and calcific pericardial cyst that had lost its epithelium.

Discussion

Pericardial cysts are benign developmental anomalies seen in approximately 1 in 100,000 people [3]. These structures constitute 6.5% of all primary tumors of the heart and pericardium [4], and they are either congenital or acquired after an infection or injury. They generally have a unilocular cavity with trabeculated inner lining, filled with serous fluid. Histologically, a single layer of mesothelial cells covers the inner surface of these cysts, with the remainder of the wall composed of connective tissue with collagen and elastic fibers.

Pericardial cysts are typically located at the cardiophrenic angles (at the right side in 51–70% of patients and at the left side in 28–38% of patients), and rarely in other mediastinal locations not adjacent to the diaphragm (8–11%) [5].

Although pericardial cysts are known to be benign structures and usually detected incidentally [6], they may be associated with life-threatening complications including rupture, pericardial tamponade, RV outflow tract obstruction, invasion of adjacent structures, such as the RV wall, atrial fibrillation, and even sudden death [7–9]. Hemorrhage into a pericardial cyst is an unusual finding that may occur after chest trauma [10] or even spontaneously in rare cases [9]. Hemorrhage into a pericardial cyst may enlarge an existing pericardial cyst and may lead to isolated compression of the cardiac chambers, such as the RV, as in our case.

Vasovagal syncope is the first diagnosis that comes to mind in a young and apparently healthy individual. However, a history of some syncopal episodes having occurred in the supine position, and the absence of any trigger for syncope, made that diagnosis less likely in our patient. On the other hand, his low blood pressure with a narrow pulse pressure and S3 heart sound, forced us to seek a cardiac etiology.

Conclusions

Even though cardiac syncope due to structural heart diseases is rarely seen in young individuals, a careful history and physical examination may lead to the correct diagnosis and treatment. Although rare, cardiologists should consider pericardial cysts in the differential diagnosis of a patient with syncope of unexplained cause.

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References

1. Moya A, Sutton R, Ammirati F et al. Guidelines for the diagnosis and management of syncope (version 2009): The Task Force for the Diagnosis and Management of Syncope of the European Society of Cardiology (ESC). *Eur Heart J*, 2009; 30: 2631–2671.
2. Strickberger SA, Benson DW, Biaggioni I et al. AHA/ACCF Scientific Statement on the evaluation of syncope. *Circulation*, 2006; 113: 316–327.
3. Roober P, Maisin J, Lacquet A. Congenital pleural pericardial cysts. *Thorax*, 1963; 18: 146–150.
4. Basso C, Valente M, Poletti A et al. Surgical pathology of primary cardiac and pericardial tumors. *Eur J Cardiothorac Surg*, 1997; 12: 730–737.
5. Stoller JK, Shaw C, Matthay RA. Enlarging, atypically located pericardial cyst. Recent experience and literature review. *Chest*, 1986; 89: 402–406.
6. Duwe BV, Sterman DH, Musani AI. Tumors of the mediastinum. *Chest*, 2005; 128: 2893–909.
7. Ng AF, Olak J. Pericardial cyst causing right ventricular outflow tract obstruction. *Ann Thorac Surg*, 1997; 63: 1147–1148.
8. Chopra PS, Duke DJ, Pellet JR et al. Pericardial cyst with partial erosion of the right ventricular wall. *Ann Thorac Surg*, 1991; 51: 840–841.
9. Borges AC, Gellert K, Dietel M et al. Acute right-sided heart failure due to hemorrhage into a pericardial cyst. *Ann Thorac Surg*, 1997; 63: 845–847.
10. Temizkan V, Onan B, Inan K et al. Hemorrhage into a pericardial cyst and associated right ventricular compression after blunt chest trauma. *Ann Thorac Surg*, 2010; 89: 1292–1295.