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Congenital Total Absence of the Pericardium: A Case Report

Case Report

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Abstract: We report the case of a 52-year-old woman who presented with a several-year history of palpitation (exacerbated by emotional stress and physical activity) and recent development of atypical chest pain. An investigation was undertaken to diagnose the patient's problem and to recommend the best possible therapy. Transthoracic echocardiography and a computerized axial tomography scan showed evidence of complete absence of the pericardium, which is a rare congenital heart defect.

Keywords: *Pericardium • Congenital heart disease • Transthoracic echocardiography*

1. Introduction

Congenital absence of the pericardium, which may be partial or complete, is an uncommon anomaly [1]. Right-sided or complete deficiencies are rarer than left-sided defects. Patients with complete pericardial absence may be asymptomatic or present with symptoms. This anomaly can be isolated or occur along with other heart defects. Herein, we report a case of isolated total absence of the pericardium with complaint of palpitation and atypical chest pain.

2. Case presentation

A 52-year-old woman with a complaint of palpitation and chest pain was referred to Tehran Heart Center to undergo echocardiography. She had experienced palpitation for several years. Recently, palpitations became worse as a result of emotional stress and physical activity. In the past three months she began to experience sharp transient chest pain with no radiation or any relationship to physical activity. New York Heart Association functional class was grade I with no experience of dyspnea. No history of cardiac disease or related conditions were found in the patient's past medical of family history.

Physical examination revealed marked displacement of the point of maximum impulse to the left. Electrocardiography showed a normal sinus rhythm, normal axis deviation and an inverted T-wave in inferior leads. Chest radiography showed only displacement of the cardiac silhouette to the left with loss of the right heart border.

Transthoracic echocardiography (TTE) in a subcostal view evaluation showed abdominal and atrial situs solitus, levocoardia and atrioventricular-ventriculoarterial concordance.

In TTE, there were very poor echo views in the left parasternal long axis and apical windows due to extreme left-sided cardiac deviation. Two differential diagnoses were supposed: 1) partial absence of the left pericardium or 2) compression of the heart by the right lung/retraction of the heart by the left lung.

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Figure 1. Apical 4-chamber view in echocardiography depicting extreme left sided cardiac shift. RV indicates right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium.

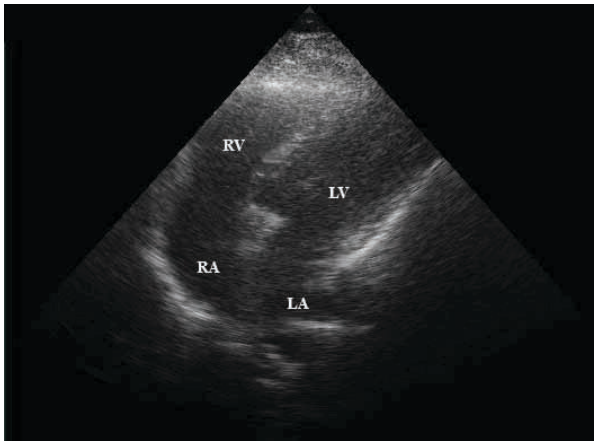


Figure 2. Air herniation between ascending arota (AO) and pulmonary artery (PA) due to pericardium absence (arrow).

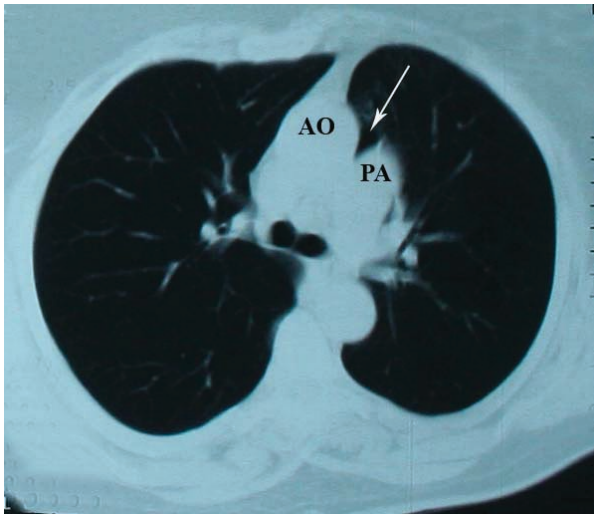
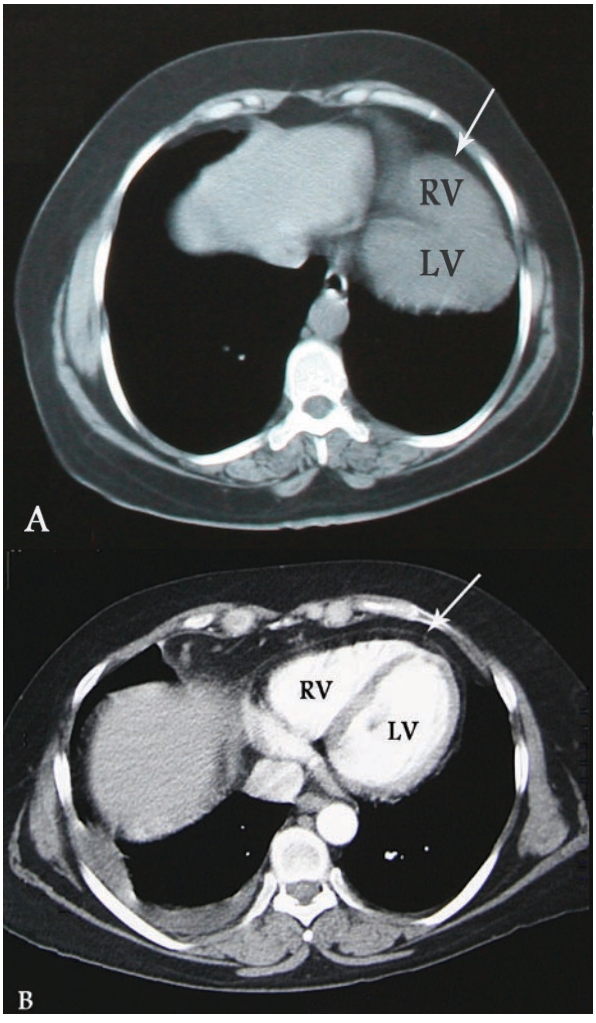


Figure 3. (A) Absence of pericardium around the heart (arrow) and cardiac shift to the left in current case. (B) Normal feature of pericardium in axial view (arrow). RV indicates right ventricle; LV, left ventricle.



Transesophageal echocardiography (TEE) was performed to assess the structure and function of cardiac chambers and valves and to rule out possible presence of congenital heart disease. In TEE, there were normal size cardiac chambers with good ventricular systolic function, and grade 1 diastolic left ventricular dysfunction. No wall motion abnormality and no evidence of significant valvular heart defects or visible shunt flow were seen (Figure 1). Therefore, the patient was referred to undergo a chest CT-scan to confirm the diagnosis.

Four slice spiral CT without cardiac gating was performed. The CT-scan was interpreted to show air herniation between the pulmonary artery and ascending aorta, and the pericardium on the right ventricle or diaphragmatic side of heart was not visualized (total pericardial absence). The heart was deviated to the left side. The lungs appeared normal and there was no evidence of a mediastinal mass or plural effusion (Figure 2, 3).

3. Discussion

Congenital absence of the pericardium is a rare anomaly [1]. Right-sided or bilateral (complete) deficiencies are even rarer than left-sided defects. An isolated defect in the anterior pericardium has been reported only once in literature [2]. Pericardial absence can be of a sporadic or familial type [3].

Congenital absence of the pericardium can occur alone or in association with other congenital anomalies

such as bronchogenic cyst, pulmonary sequestration, hypoplasia of the left lung and left pulmonary artery, dextroisomerism, diaphragmatic hernia, hepatic hemangioendothelioma, ruptured type A aortic dissection, or tricuspid regurgitation [4-8]. There was no coexisting heart anomaly in our case.

Patients with complete pericardial absence may either be asymptomatic or present with chest pain, arrhythmia, dyspnea or a sensation of "shifting heart". Described chest pain differ in severity, type, duration and frequency; however, pain can mimic coronary artery disease in some cases [1]. Probable causes of pain include impaction, ischemia or heart mobility due to the pericardial defect [1,7]. Interestingly, our case presented with palpitation for several years and only atypical chest pain in the recent three months.

Most reported cases of pericardial absence have been incidental findings during thoracic surgery and imaging for unrelated medical conditions, or at postmortem examination [1,9].

Although chest x-ray film and echocardiography can guide one to identify this abnormality, magnetic resonance imaging (MRI) and computed tomography (CT) seem to be the best imaging techniques. Nakashima et al. diagnosed pericardial absence through echocardiography and ECG during several positional changes; CT scan and MRI were, however, inconclusive [10].

In contrast to complete absence of the pericardium, which is benign, partial absence may lead to herniation or strangulation of the heart or its appendages through the defect [1]. Thus, it is important to distinguish the partial from complete type when considering whether prophylactic surgery should be performed.

Complete pericardial absence usually requires no treatment unless the patient is symptomatic. Primary closure, pericardiectomy, left atrial appendectomy, division of adhesions, extension of the defect, or pericardioplasty are the surgical procedures employed for partial defects or symptomatic patients [1,6]. For our patient, we recommended a β-blocker to treat palpitations, with close follow-up to ensure symptom resolution.

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