Case Report

CONGENITAL COMPLETE ABSENCE OF THE LEFT PERICARDIUM DIAGNOSED BY ECHOCARDIOGRAPHY

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Abstract

Congenital absence of the pericardium is a rare condition. The most common form is complete absence of the left pericardium and asymptomatic patients are discovered during cardiac surgery for an unrelated condition and symptomatic patients may experience chest pain or dyspnea. A 37 year-old female presented with dyspnea and during echocardiography with positional changing maneuver. The patient was found to have complete absence of the left pericardium that confirmed with CT scan. Echocardiography with positional changing maneuver can be diagnostic helpful tool for diagnosis of congenital complete absence of the left pericardium.

Keywords: Congenital heart anomaly, Echocardiography, Case report.

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Introduction

Congenital absence of the pericardium is a rare condition that is typically discovered at autopsy or during cardiac surgery [1,2]. The most common form is complete absence of the left pericardium that is a benign condition compatible with a normal life span [3].The second most frequent form is partial absence of the left pericardium with life-threatening consequences such as herniation and rupture.Right sided lesions and bilateral complete absence of the pericardium are extremely rare [3]. In symptomatic patients with complete left absence,stabbing chest pain and/or dyspnea are most common symptoms [4,5]. ECG, CXR, Echocardiography, CT scan and MRI can help in the diagnosis.

Case presentation

A 37 year-old female presented with dyspnea that was non-exertional and unpredictable without chest pain, cough or localization of dyspnea. On examination, vital signs were normal. Jugular venous pressure (JVP) and lung auscultation were normal. On cardiac examination, apical impulse was displaced to the mid-axillary line with a grade II/VI systolic murmur heard best at the left sternal border without radiation. Findings of the abdomen and extremities examinations were unremarkable.

A 12 lead ECG showed normal sinus rhythm with right axis deviation and incomplete RBBB.

A CXR showed displacement of the heart to the left hemithorax without tracheal deviation, the right heart border superimposed on the spines and flattening of the left cardiac silhouette.

Because of dyspnea with murmur, the patient underwent an echocardiography. In standard parasternal long- axis view in transthoracic echocardiography (TTE), unusual echocardiographic window was observed including anterior rotation of the right atrium, yielding the unusual appearance of a 4 chamber view in the standard parasterenal long- axis view and also posterior and lateral rotation of the LV apex observed (Figure 1a) .In standard 4 chamber view, a highly lateral probe position was required for perfect 4 chamber imaging. In this moment the patient was suspected to have congenital absence of the pericardium and thus the echocardiography was carried out in right lateral decubitus. Because of the shift of the heart to the right in right lateral decubitus, into the principal location, unusual view corrected and

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standard view recorded in standard position (Figure 1b) This maneuver confirmed movement of the heart inside the chest from side to side and thus this finding can be helpful diagnostic clue for presence of complete absence of the left pericardium.

Correction of septal motion abnormality and RBBB in ECG in this disease by positional changing maneuver was previously reported [6].

For verifying the diagnosis, the patient underwent CT scan and after ruling out the mediastinal lesions for shifting heart, the diagnosis of the absence of the left pericardium verified. In CT scan, leftward and posterior rotation of the heart and absence of complete left pericardium was observed (Figure 2b).

Interposition of the lung, between aorta and pulmonary trunk also seen, pathognomonic of this disease (Figure 2a) [7].

Because of the mild symptom and complete absence of the left pericardium, the patient underwent conservative management. Nonetheless, all patients with partial absence, because of hazardous complications like herniation and severe symptomatic patients with complete absence must be undergoing corrective surgery[1].

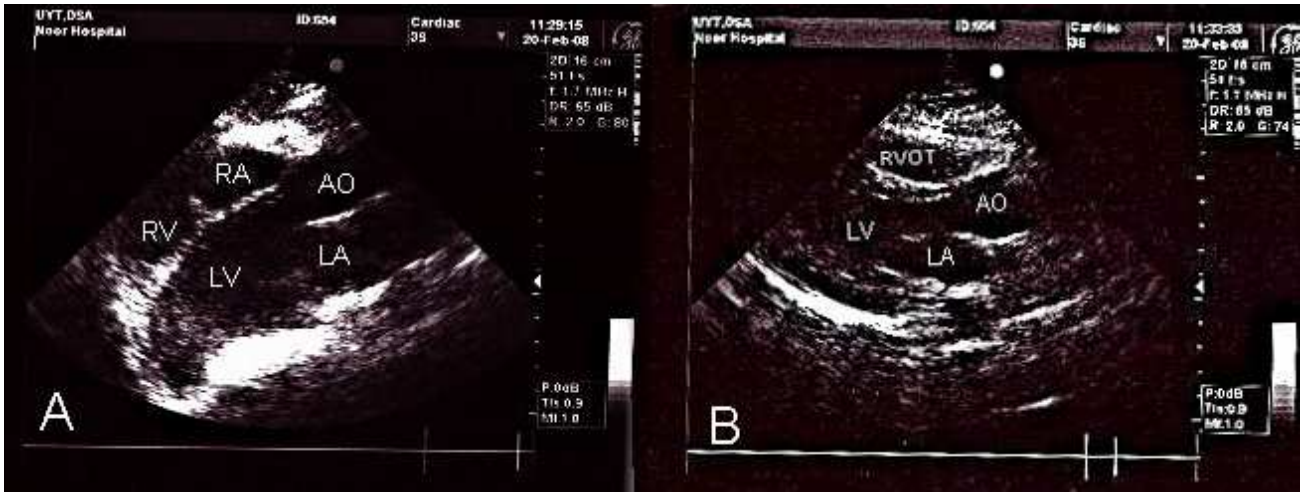

FIGURE 1: (a) Echocardiography in left lateral decubitus in standard parasternal long-axis view.
(b) The same patient after position changing to right lateral decubitus with corrected view.

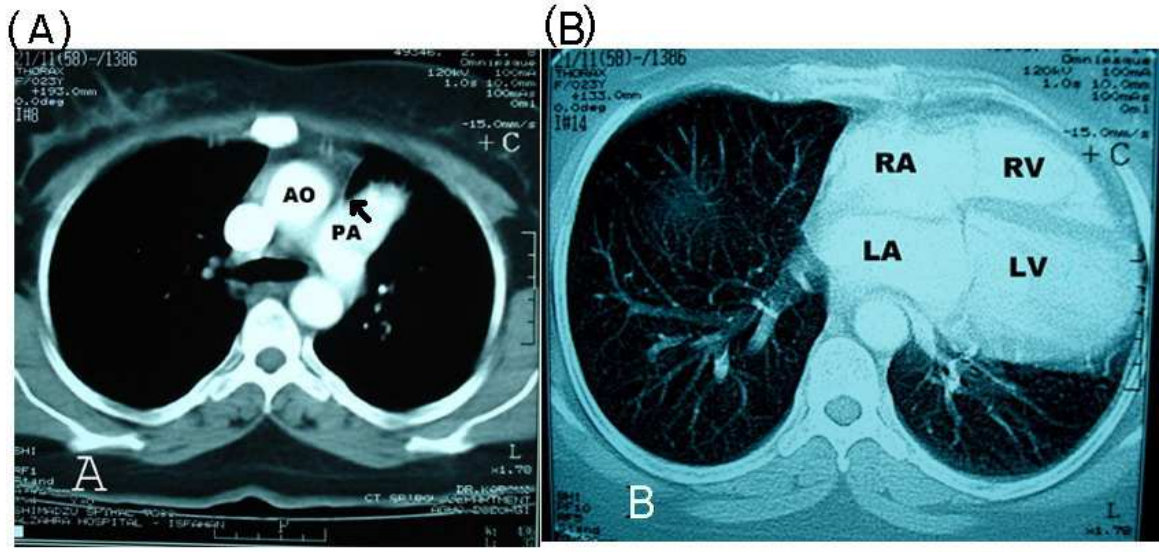

FIGURE 2. (A) Chest CT mediastinal setting shows characteristic interposition of the lung (arrow) between aorta (AO) and pulmonary artery (PA).
(B) Chest CT axial lung setting shows leftward and posterior rotation of the heart and absence of the pericardium laying the posterior portion of the heart.

Conclusion

Congenital complete absence of the left pericardium is a benign disease. Echocardiography may be diagnostic and characteristic especially if unusual echocardiographic window in parasternal long- axis view can be corrected by changing position of the patient from left lateral decubitus to right lateral decubitus.

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