

A rare case about pericardium: Left deviated heart and pericardial agenesis

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ABSTRACT

Congenital absence of the pericardium is not a common condition in daily practice. There are no obvious and clear symptoms. This condition, which is diagnosed incidentally, may cause some complications when not diagnosed. Therefore, imaging techniques, such as echocardiography, are essential. In this article, we present a rare case of pericardial agenesis.

Keywords: Congenital; pericardial agenesis; pericardium.

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Absence of the pericardium is a rare condition which is usually asymptomatic [1]. It is generally diagnosed incidentally during surgical procedures or autopsies. Seldomly, serious complications may occur. Patients describe atypical complaints and are diagnosed by ultrasonographic or radiological examination. Herein, we present a case of pericardial agenesis (PA) that was found incidentally in a patient who presented with dyspnea and palpitation.

CASE REPORT

A 71-year-old female patient with a history of hypertension, diabetes mellitus, and paroxysmal atrial fibrillation admitted to our cardiology department with dyspnea and palpitation. Physical examination showed blood pressure of 130/85 mmHg, pulse rate of 51 bpm, oxygen saturation of 95%, and respiratory rate of 20/min. The cardiovascular examination revealed left-sided apical impulse, and there were no pathological findings in the other systems examination at first evaluation. Electrocardiography

showed normal axis, normal sinus rhythm with 47 heart bpm, and negative T-waves at V1–5 leads (Fig. 1). At the laboratory panel including complete blood count, cardiac biomarkers, electrolytes, typical values were detected. Transthoracic echocardiography (TTE) showed an unusual view with the apex subsiding posteriorly in the thorax the apical window was displaced at the posterior (Fig. 2). A posteroanterior chest radiograph showed loss of the right heart border and on the lateral view a posterior bulging of the heart with the apex (Fig. 3). As a result of these evaluations, total absence of the pericardium was considered. Magnetic resonance imaging (MRI) was planned for further examination. MRI could not be performed due to knee prosthesis, so computed tomography angiography (CT angiography) was performed and revealed that the whole heart was shifted in the left hemithorax with the apex pointing posteriorly and PA (Fig. 4a). Furthermore, chest CT shows interposition of the lung between the aortic arch and pulmonary trunk (Fig. 4b). No additional treatment was planned, and outpatient clinic follow-up was recommended.

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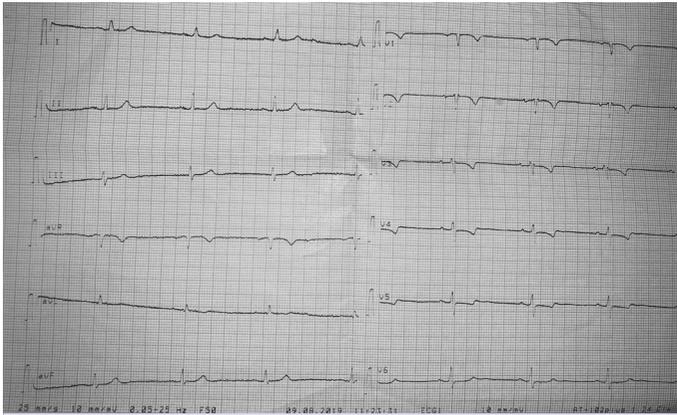


FIGURE 1. Electrocardiogram shows normal axis and negative T-waves in leads V1–5 at presentation.

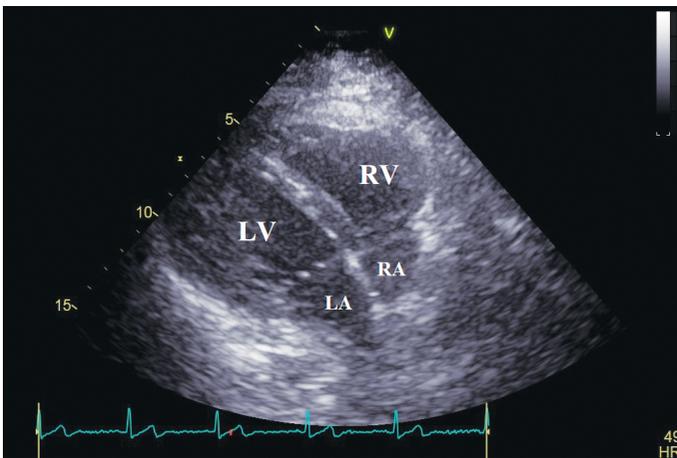


FIGURE 2. The image of the left deviated heart in two-dimensional echocardiography. The apical 4-chamber window showing the abnormal position of the apex, receding to the left (unusual long-axis parasternal view with an oblique).

LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.

DISCUSSION

PA is asymptomatic, especially in the total absence of the pericardium. Complete and partial agenesis occurs at a rate of approximately 1/14,000 and is more common in males at a ratio of 3:1 [2]. PA is a rare clinical entity that is recognized incidentally. The diagnosis of PA is quite difficult, except for cardiac surgery or autopsy [3]. Physical examination may show some findings but may not clearly indicate this disease. ECG is usually normal, but right axis deviation, reduced R-wave progression secondary to leftward displacement partial or complete right bundle branch block may be detected [4, 5]. TTE is the most helpful test in the evaluation.

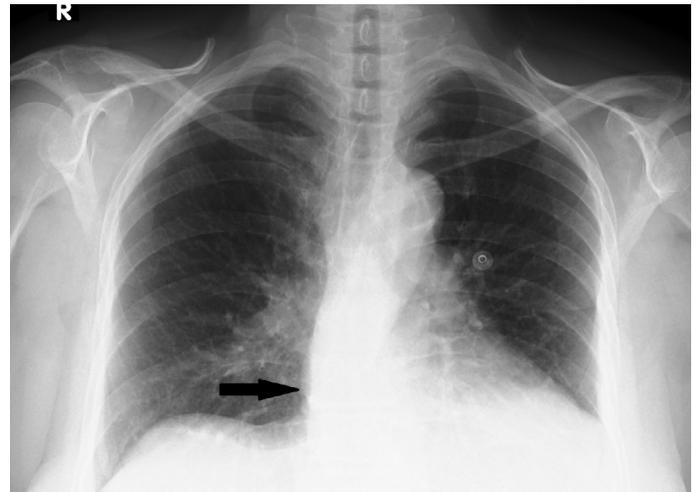


FIGURE 3. Chest X-ray image: Frontal projection showing heart displacement in the left-sided position (arrow).

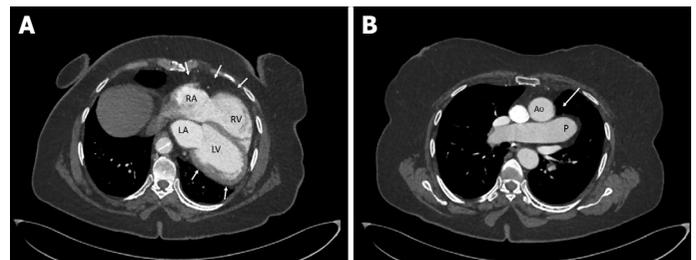


FIGURE 4. Computed tomography angiography imaging and chest X-ray in a patient. **(A)** The chest computed tomography (CT) image shows the absence of pericardium along with the cardiac chambers (white arrows). Chest CT also reveals deviation of cardiac apex to a more lateral position and shift of heart to the left hemithorax. **(B)** The chest CT reveals the interposition of the lung between the aortic arch and pulmonary trunk (white arrow).

LA: Left atrium, LV: Left ventricle, RA: Right atrium, RV: Right ventricle, Ao: Aorta, P: Pulmonary trunk

Because the 1st time, we suspect this disease with the findings during this TTE. There are some characteristic echocardiographic findings such as unusual echocardiographic view, cardiac hypermobility, and abnormal swinging motion of the heart [5, 6]. CT and MRI may be useful for diagnosis, but sometimes they may be insufficient. Typically, there is no interposition of lung tissue between the aorta and the pulmonary artery, but absence of the pericardium allows this condition. This change is evaluated by CT and MRI better than chest X-ray [4]. MRI is accepted as the standard gold method in diagnosis because it can display images synchronized with the cardiac cycle [3]. PA may present

with partial or complete absence. Although both have great prognosis and are mostly asymptomatic, they may rarely cause fatal complications [6]. There is a potential risk of cardiac herniation, strangulation, and incarceration in the partial absence of the left pericardium. In fact, patients may present with many clinical conditions such as arrhythmia and sudden death [7, 8]. Surgical intervention may be necessary for symptomatic patients, but follow-up is recommended in asymptomatic patients. Surgical technique is determined according to the size of the defect. If it's small one, some techniques can be used such as primary closure technique, patch closure, or left atrial appendectomy [7, 9]. Herein, we present a case of PA that was diagnosed incidentally in a patient presenting with atypical complaints.

Learning Points

In conclusion, if patients have characteristic features during echocardiography, we should suspect PA. There is no need for surgical intervention in PA, which is very rare and usually asymptomatic. However, if it causes any cardiac complications, then surgery can be performed. Patients should be treated according to their symptoms. Treatment options should be evaluated according to clinical follow-up.

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