

## Case Report

### *Role of Multimodality Imaging in a Case Series of the Partial Absence of the Pericardium: A Case Report*

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#### ABSTRACT

**Background:** The congenital absence of the pericardium is a rare cardiac entity which could be manifested in isolation or in association with other anomalies. Usually, affected cases are asymptomatic. Hereby, we describe adults with recent palpitations who were diagnosed with the partial absence of the pericardium on chest computed tomography (CT).

**Case Presentation:** The first case was a 49-year-old woman, who presented with palpitations, especially in the left decubitus position, of 1 year's duration. Her physical examination revealed nothing of significance, except for tachycardia. The second case was a 37-year-old woman, who presented with dyspnea. Transthoracic echocardiography suggested the partial absence of the pericardium. The definite diagnosis was made using multidetector CT, which was compatible with the echocardiography findings. An abnormally leftward rotated heart without a tracheal deviation, an elongated and flattened left ventricular border (Snoopy's sign), the absence of the pericardium, an excessive cardiac motion, a prominent pulmonary artery, and cardiac pulsations were seen on the scan slices. A lucent area between the diaphragm and the heart or the aorta and the pulmonary artery was also seen due to an interposed lung tissue. The final case was a 40-year-old man, who presented with dyspnea. In addition to the above findings, dextrocardia and a persistent left superior vena cava were noticeable.

**Conclusions:** Palpitations and sinus tachycardia in adults may be a mere presentation of the partial absence of the pericardium, and multimodality imaging can be applied for the proper identification of this entity. (*Iranian Heart Journal 2018; 19(4): 54-57*)

**KEYWORDS:** Congenital absence of pericardium, Palpitation, Multidetector CT

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The congenital absence of the pericardium is a rare cardiac anomaly with a male preponderance. The maldevelopment of pleuropericardial membranes results in the partial or complete absence of the pericardium,

with the latter anomaly being more common.<sup>1</sup> The congenital absence of the pericardium usually occurs in isolation, but sometimes it occurs with other congenital abnormalities.<sup>2</sup> Patients with this disorder are often

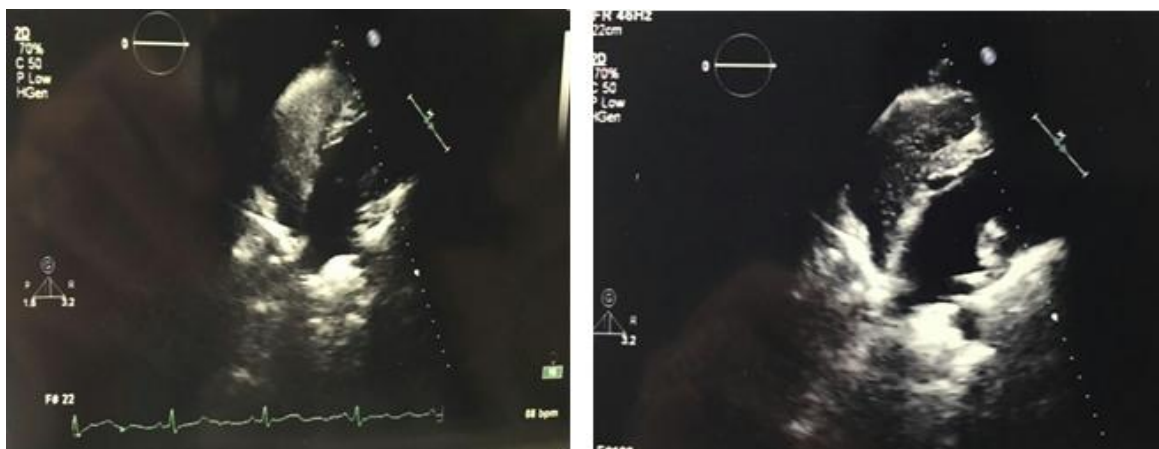
asymptomatic; nonetheless, various clinical presentations have been reported such as palpitations, dizziness, dyspnea, and lancinating chest pain.<sup>3</sup> In cases with the complete absence of the pericardium, the prognosis is excellent, whereas in cases with the partial absence of the pericardium, heart herniation, strangulation, tricuspid regurgitation following chordal traction, ischemia, and sudden cardiac death might occur.<sup>1,4</sup>

Clinical manifestations, electrocardiography (ECG), and chest radiography are not conclusive for the diagnosis and often multi-imaging modalities are required. Paradoxical septal motions, enlarged right ventricles in systole, bulbous ventricles, elongated atria, and teardrop appearances of the left ventricle could be seen on echocardiography. Rightward axis shifts, herniation of the heart through defects, and attachments of the right pericardium to the anterior chest wall are additional findings seen on decubitus CT scan.<sup>5</sup> We herein report 3 cases of the partial absence of the pericardium with recent occurrences of sinus tachycardia.

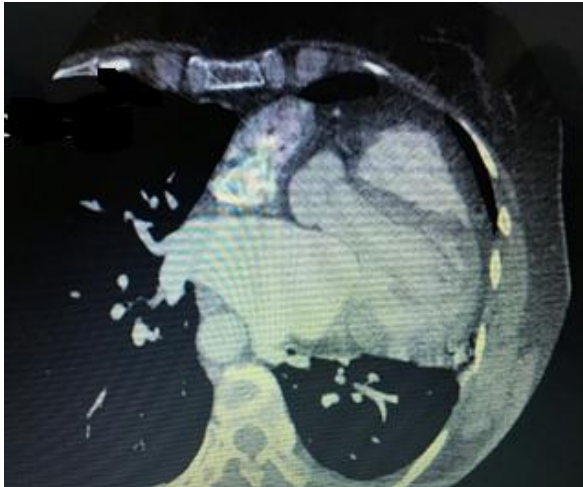
### Case Presentation

The first case was a 49-year-old woman, who referred to us with complaints of palpitations, especially in the left decubitus position, of 1 year's duration. She was a physically active woman without a significant past medical history. Other than tachycardia, nothing else

was found in her physical examination. In 12-lead ECG, sinus tachycardia was detected. All the lab data were within the normal limits. Transthoracic echocardiography suggested the congenital partial absence of the pericardium with a posteriorly displaced heart, straight left-heart borders, and the enlargement of the right ventricle in systole. Contrast echocardiography revealed no bubble passage from right to left side (Fig. 1). The definite diagnosis was made via multidetector CT, which was compatible with the echocardiography findings. An abnormally leftward rotated heart without a tracheal deviation, elongated and flattened left ventricular borders (Snoopy's sign), the absence of the pericardium, an excessive cardiac motion, a prominent pulmonary artery, and cardiac pulsations were seen on scan slices.<sup>6</sup> Additionally, a lucent area was observed between the diaphragm and the heart or the aorta and the pulmonary artery, due to an interposed lung tissue.<sup>7</sup> Reconstructed CT images are depicted in Figure 2. In this case, the recent tachycardia was an implication toward the diagnosis of the congenital partial absence of the pericardium. The usual ECG finding in the pericardial absence is bradycardia with a right bundle branch block. Prominent P waves and a poor R-wave progression have also been previously reported.<sup>8</sup> In our case, sinus tachycardia was the main ECG finding.



**Figure 1.** Transthoracic echocardiography of the patient with the congenital absence of the pericardium



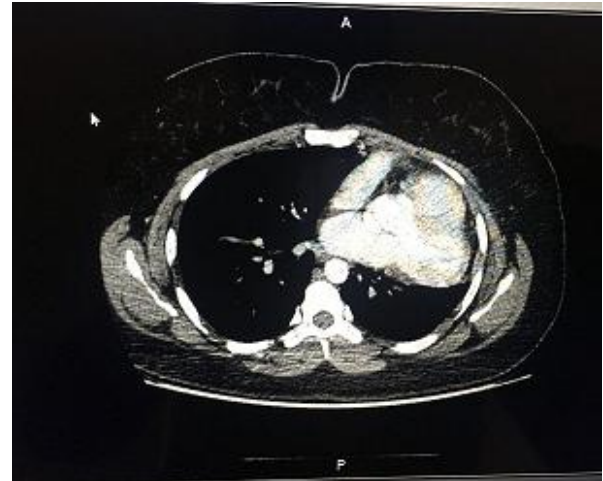
**Figure 2.** Reconstructed computed tomographic angiography imaging of the first patient, with the congenital absence of the pericardium



**Figure 3.** Computed tomographic angiography of the second patient, with the congenital absence of the pericardium

The second case was a 37-year-old woman, who presented with dyspnea. The imaging findings were the same as those in the first case (Fig. 3).

The third and final case was a 40-year-old man, who presented with dyspnea. In addition to the partial absence of the pericardium, he was diagnosed with dextrocardia and a persistent left superior vena cava (Fig. 4).



**Figure 4.** Reconstructed computed tomographic angiography of the third patient, with the partial absence of the pericardium in addition to dextrocardia and a persistent left superior vena cava

## CONCLUSIONS

Palpitations and sinus tachycardia in adults may constitute a mere presentation of the partial absence of the pericardium, and multimodality imaging can be applied for the proper identification of this entity.

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