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Case Report

A case report of congenital absence of the pericardium that was diagnosed by cardiac computed tomography angiogram (CCTA)☆,☆☆

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ABSTRACT

This case report describes a 40-year-old male who presented to the emergency department (ED) with chest pain. Initial diagnostic workup was concerning for a congenital cardiac anomaly, further imaging revealed complete congenital absence of the pericardium (CAP) which is a rare condition. Multimodality cardiac imaging including cardiac computed tomography angiogram (CCTA) was used to confirm the diagnosis of CAP. We briefly discuss various clinical presentations of CAP along with potential complications and other anomalies that could be associated with pericardial agenesis.

Abbreviations: CAP, congenital absence of the pericardium; CMR, cardiac magnetic resonance; CCTA, cardiac computed tomography angiography (CCTA).

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Background

Congenital absence of pericardium (CAP) is a rare cardiac malformation, that affects about 1/10,000 births [1]. CAP can be complete or partial. Complete left-sided defects accounts for about 70% of all pericardial defects. Although CAP is a congenital anomaly, some iatrogenic cases have been reported [2,3]. Patients with complete CAP are usually asymptomatic and require no intervention, while those with partial CAP could be at risk of sudden cardiac death (SCD) requiring surgical intervention [4–7]. We report a case of a 40-year-old male who presented with chest pain and his Chest CT scan revealed presence of CAP.

Clinical vignette

A 40-year-old male with a medical history significant for hyperlipidemia and supraventricular tachycardia (SVT) presented to the emergency department (ED) with a chief complaint of recurrent non-radiating, mild substernal chest discomfort, with no clear aggravating or alleviating factors. Review of systems was unremarkable. Vital signs on evaluation were within normal limits. Physical examination demonstrated no cardiopulmonary abnormalities. Comprehensive laboratory evaluation was unremarkable. Troponin was within the normal range. Electrocardiogram (ECG) revealed sinus tachycardia, right axis deviation, incomplete right bundle branch block (RBBB) with ST and T wave abnormalities involving the anterolateral and inferior leads suspicious for ischemia (Fig. 1). Chest X-ray showed straightening and elongation of left sided cardiac borders suggestive of Snoopy sign

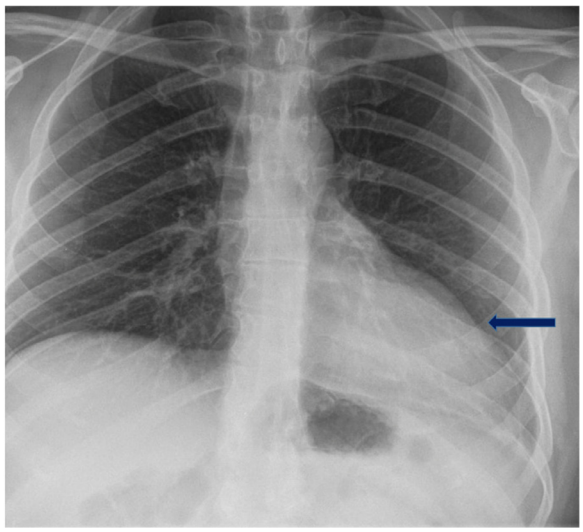

Fig. 2 – Chest X-ray reveals the posterior and leftward rotation of the heart through the straightening and elongation of the left border of cardiac silhouette, Snoopy sign (Arrow). There is indistinct right heart border that covered by the spine. Note the lucent area that indicates interposition of lung tissues between the aorta and pulmonary artery.

(Fig. 2). Initial transthoracic echocardiogram (TTE) was of limited value due to the imaging view being significantly off-axis and thus further evaluation was recommended. Computed tomography angiography (CTA) of the chest was performed to rule out pulmonary embolism (PE), and it showed abnormal orientation and configuration of the heart and mediasti-

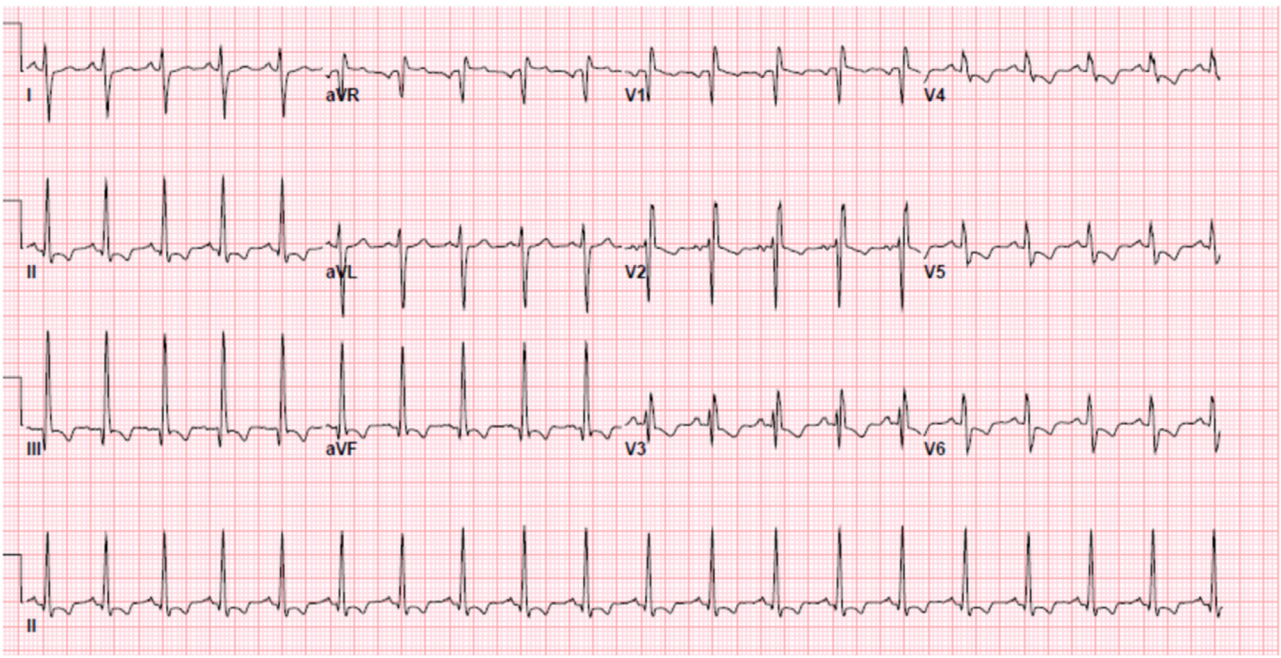

Fig. 1 – Electrocardiography (ECG) shows sinus tachycardia, right axis deviation, incomplete right bundle branch block (RBBB) with ST depression and T wave inversions involving the anterolateral and inferior leads.

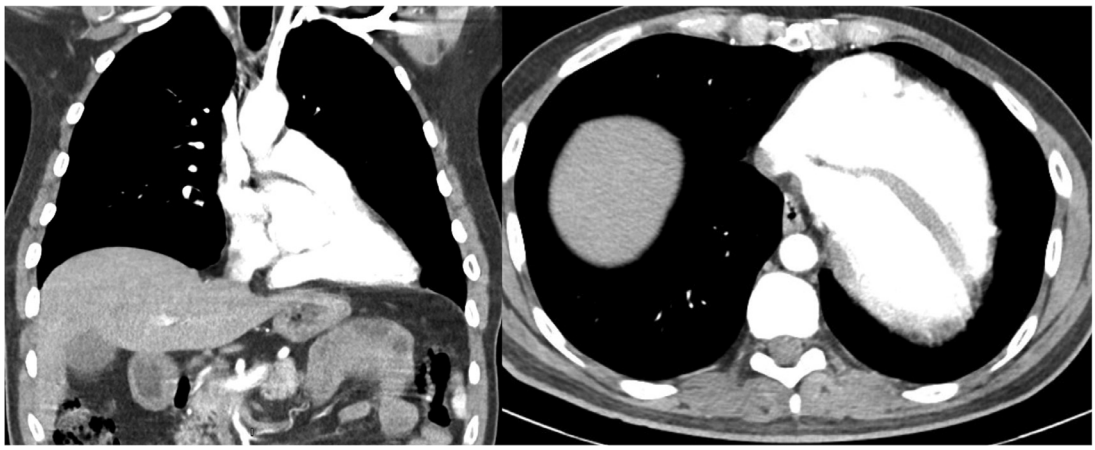

Fig. 3 – (A) Chest CTA coronal setting shows a leftward shift of the heart, absent pericardium and mild flattening and elongation of the left heart border (snoopy sign). (B) Axial chest CTA concerning for absence of the pericardium along the ventricular border and its possible presence only at the level of LV apex and right atrium. There is a levoposition of the heart.

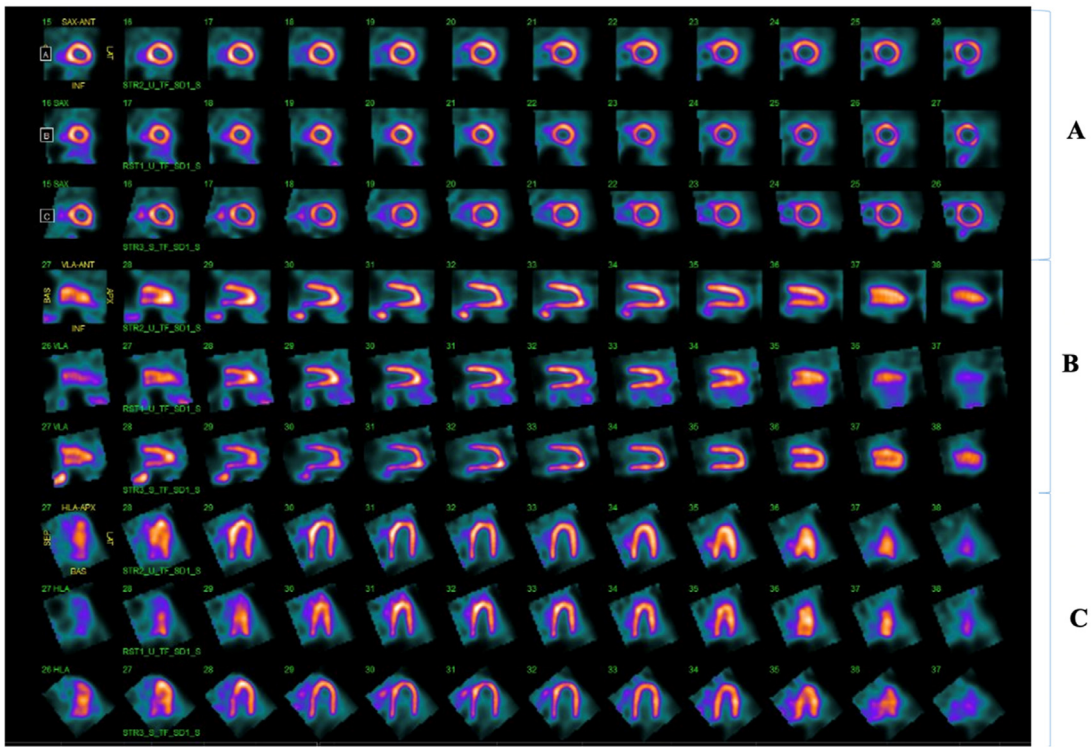

Fig. 4 – Regadenoson myocardial perfusion images revealing presence of transient ischemic dilatation (TID) of 1.7. In each view: (A) Short axis views. (B) Vertical long axis views. (C) Horizontal long axis view. Top row in each: Stress upright images; Middle row in each: Resting images; Bottom row in each: Stress supine images.

nal structures, suggesting a potential congenital heart defect (Figs. 3A and B). According to the ED chest pain protocol, a Regadenoson myocardial perfusion scan was performed that revealed absence of any myocardial perfusion defects at rest or stress, with a resting LVEF of 71%, post stress LVEF of 52%, however; transient ischemic dilatation (TID) of 1.7 was noted (Fig. 4). Given his recurrent symptoms and chest CTA findings, a dedicated gated cardiac computed tomography angiogram (CCTA) for the assessment of anatomic CAD and for better characterization of cardiac morphology was performed. Normal coronary artery origins with the absence of plaques were identified. Almost complete absence of the pericardium with dilated right-sided cardiac chambers was noted (Figs. 5A-D). The left ventricle was rotated toward the posterior left thorax, and the cardiac apex was pointed posteriorly. Interposition of the lung tissue between the aorta and pulmonary artery

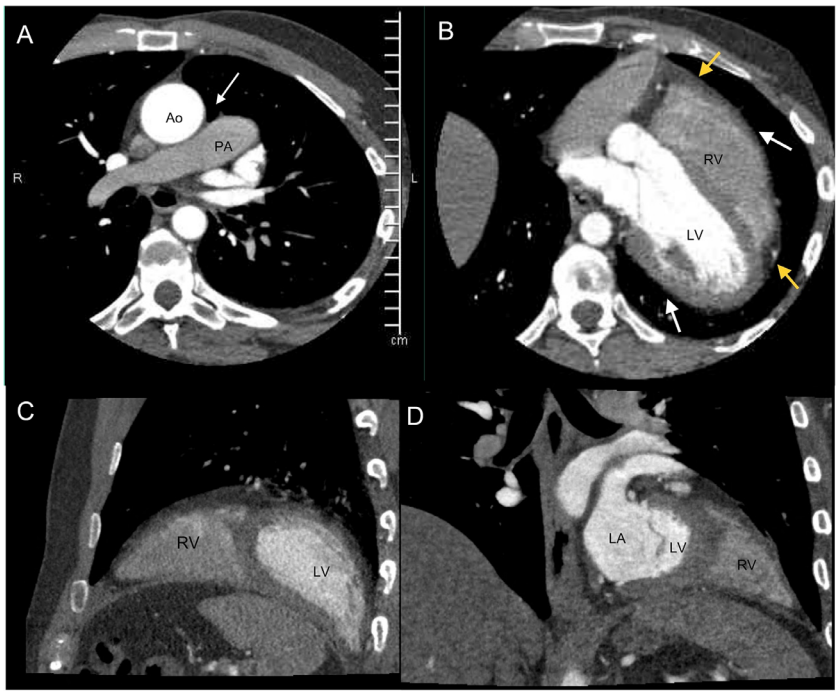

Fig. 5 – Axial cardiac computed tomography angiogram (CCTA) image revealing: (A) the interposition of lung tissue between the aorta and the main pulmonary artery (white arrow). (B) Shows complete absence of the pericardium along the ventricular border (white arrows), and its possible presence only at the level of LV apex and right atrium (yellow arrows). Note the levoposition of the heart. Sagittal (C) and coronal (D) images revealing levoposition of the heart. LV, left ventricle; RV, right ventricle; Ao, aorta; PA, pulmonary artery.

was present, which is a finding characteristic of absent pericardium. A diagnosis of congenital absence of the pericardium (CAP) was made.

Discussion

The pericardium is responsible for a stable position of the heart in the mediastinum and prevents overdistension, and facilitates the atrial filling [8]. CAP was described for the first time in an autopsy by Realdo Colombo in 1559. However, it was diagnosed and reported radiologically for the first time by Ellis in 1959 [9]. Early atrophy of Cuvier's duct which is involved in the development of pleuro-pericardial membranes is thought to be the etiology behind congenital deficiency of the pericardium [10].

CAP can be subdivided based on the defect's size and site to be complete or partial type. The complete CAP could be biventricular (9%), left-sided (70%) or right-sided only (17%). Complete CAP is mostly silent and found accidently; however, some patients present with atypical chest pain, dyspnea and palpitations that are hypothesized to be related to cardiac hypermobility [4–6]. Partial CAP is reported to be associated with serious complications, including ventricle herniation and coronary circulation compression that could lead to sudden cardiac death (SCD), with first reported SCD in 1887 by Boxall [7].

CAP is mostly reported in literature to be a single congenital defect; however, it can be associated with other congenital anomalies in about 30% of cases that needs further investigation. Such rare abnormalities include: bronchogenic cyst [11], thoracic aortic aneurysm [12], pleural agenesis and coronary artery disease [13], pentalogy of Cantrell with complete ectopia cordis [14], diaphragmatic hernia with ectopic liver [15], and straight back syndrome [16].

Diagnosis of CAP can be challenging, and various clinical presentations have been reported. Nisanoglu [17] reported a case of CAP associated with type A aortic dissection that led to hypovolemic shock secondary to spontaneous drainage of blood into the pleural cavity, but the absence of pericardium prevented the development of cardiac tamponade and cardiogenic shock. Brulotte [18] reported a patient with non-ST-elevation myocardial infarction (NSTEMI) who underwent cardiac catheterization with no obstructive CAD. In this patient a CT chest with contrast ruled out PE and CAP was noted. Sempokuya [19] reported a patient with recurrent typical pericarditis with symptoms of pleuritic chest pain and diffuse ST segment elevation on the ECG, who on CT imaging and TTE was noted to have CAP.

In these challenging cases of CAP, history and physical examination are often unremarkable, and ECG findings are mostly non-specific. Chest x-rays reveals posterior and leftward shift of the heart with elongated left border of the cardiac silhouette (Snoopy sign) [20], indistinct right heart border, and interposition of lung tissue with lucency in the aortopulmonary window or between the heart base and diaphragm.

Partial pericardial defect can be seen on the x-ray as a bulge through the heart's contour during diastole. Echocardiograms can detect the increased cardiac mobility and abnormal position and orientation of the heart [21]. Chest CTA and cardiac magnetic resonance imaging (CMR) provide a detailed description of the pericardium and thus are superior to the echocardiogram in that aspect [22]. These advanced imaging shows the abnormal position of the heart and evaluates for other intracardiac anomalies. Abnormal lung tissue interposition is also recognized on these modalities.

Conclusion

CAP is a rare congenital cardiac anomaly. Patients with complete CAP are usually asymptomatic and require no intervention, while those with partial CAP are at risk for herniation of cardiac chambers or coronary artery compression, which can lead to serious complications, including sudden cardiac death (SCD), requiring surgical intervention. Advanced cardiac imaging modalities, including CTA or CMR are required to confirm the diagnosis due to their higher spatial and temporal resolution. In our current case, chest CTA was used to make the final diagnosis of CAP. TID was noted on the nuclear stress test; however, its significance in the absence of perfusion defects in patients with CAP is unknown.

Patient consent

Written and informed consent for publication of this case was obtained from the patient.

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