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Case Report

It's Never Too Late: Spotting Congenital Absence of the Pericardium in an Older Adult

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ABSTRACT

A 75-year-old male with a cardiopulmonary history presented with chest pain and dyspnea. He was hypertensive. An electrocardiogram showed paced rhythm. A high-sensitivity test showed his troponin T level was minimally elevated. Coronary angiography results were unremarkable. Chest radiography revealed an elevated cardiac apex, previously attributed to cardiomegaly. Echocardiography revealed a teardrop shaped heart in a nonstandard apical window. Computed tomography confirmed congenital absence of the left pericardium. Challenges of recognizing a rare condition are highlighted. Congenital absence of the pericardium, an often benign but rarely catastrophic condition, can masquerade for decades before diagnosis, underlining the importance of clinical vigilance in evaluating common cardiac complaints.

RÉSUMÉ

Un homme de 75 ans qui avait des antécédents cardiopulmonaires a éprouvé une douleur thoracique et accusé une dyspnée. Il souffrait d'hypertension. Un électrocardiogramme a montré un rythme électro-entraîné. Le dosage de la troponine T hautement sensible a révélé des concentrations minimalement élevées. Les résultats de l'angiographie coronarienne étaient normaux. La radiographie pulmonaire a révélé un apex du cœur élevé, antérieurement attribué à la cardiomégalie. L'échocardiographie a révélé un cœur en forme de larme dans une fenêtre apicale non standard. La tomodensitométrie a permis de confirmer l'absence congénitale du péricarde gauche. Nous présentons les enjeux liés à l'identification d'une maladie rare. Puisque l'absence congénitale du péricarde, une anomalie souvent bénigne, mais rarement catastrophique, peut demeurer dissimulée durant des décennies avant le diagnostic, nous soulignons l'importance de la vigilance clinique dans l'évaluation des symptômes cardiaques courants.

Case

A 75-year-old male with hypertension, a pacemaker, and chronic obstructive pulmonary disease (COPD) presented with atypical chest pain and dyspnea. There were no congestive symptoms or palpitations. He was hypertensive (161/72 mm Hg), similar bilaterally. There were scant crackles. An electrocardiogram demonstrated an atrial-sensed ventricular-paced rhythm. High-sensitivity measurements showed troponin T to be minimally elevated, 18 ng/L (> 15 ng/L).

Chest radiography demonstrated an elevated cardiac apex with an indistinct right heart border (Fig. 1A), interpreted as cardiomegaly on prior films. A transthoracic echocardiogram was performed, with apical views obtained supine in the left posterior axillary line. The left ventricle was bulbous with elongated atria in a "teardrop" shape (Fig. 1B; view Videos 1 and 2, online). Abnormal septal motion was noted during a paced rhythm. Biventricular size and systolic function were normal. Congenital absence of the pericardium (CAP) was suspected, though not noted on prior studies.

Computed tomography of the chest revealed leftward deviation of the long axis of the heart (Fig. 2A) and lung parenchyma interposition between the ascending aorta and the main pulmonary artery without cardiac herniation (Fig. 2B), consistent with complete left CAP.

Coronary angiography results were unremarkable. Diuretic therapy did not impact symptoms. His blood pressure and symptoms improved. He was discharged with follow-up for COPD and CAP. He is stable at 1 year.

Discussion

CAP is rare, with an estimated prevalence of 0.007%–0.044%.[1] The pericardium may be completely (left, right, or both) or partially (left or right) absent.[2,3] During embryogenesis, the pleuropericardial folds fail to fuse, possibly due to an atrophic left common cardiac vein or cardiac enlargement before fusion completion.[3]

Consistent with our case, left complete CAP is most common[1] with a male predominance.[2] However, the median age at diagnosis is much younger (21 years[4] and 48 years[3] in 2

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Novel Teaching Points

- CAP may masquerade for decades before diagnosis, as highlighted by this case.
- Recognizing clinical clues on history (eg, trepopnea) and common cardiac tests (eg, "Snoopy's" sign on chest radiograph, teardrop shaped heart on echocardiography) may avoid rare catastrophic consequences in this otherwise benign condition.
- Advanced imaging (computed tomography/magnetic resonance imaging) is warranted for both diagnosis (demonstrating interposition of lung tissue between the aorta and pulmonary artery) and prognosis (to exclude high-risk anatomy).

case series). Most patients are asymptomatic,[1] with CAP being detected incidentally during imaging or surgery.[2] Symptoms are nonspecific, including chest pain (most commonly), palpitations, syncope, or dyspnea[2,3] and can have confounders, such as COPD as in our case. Symptoms may be due to the absence of normal pericardial cardiac cushioning, torsion of great vessels, and vascular compression of coronary arteries or pulmonary veins with hemodynamic collapse.[1,3]

Associated congenital anomalies are present in a third of cases.[3] Although some lesions present early (eg, tetralogy of Fallot),[3] others may continue undetected until adulthood (eg, atrial septal defects, bicuspid aortic valve, and ventricular noncompaction[1,3]).

Underrecognized chest radiograph clues include leftward cardiac displacement, loss of the right heart border, and straightening/elongation of the left heart border ("Snoopy's" sign, Fig. 1A).[1]

On echocardiography, the sonographer reported challenging imaging windows, even for a COPD patient, an early common observation in CAP.[2,3] The teardrop shaped heart (elongated atria and bulbous ventricles[2]) within a displaced apical window flagged the suspicion for CAP. Additional echocardiographic features include paradoxical septal motion (present but confounded by paced rhythm), cardiac hypermobility, and apparent right ventricular enlargement (due to leftward shifting of the cardiac chambers[5]) with tricuspid regurgitation.[2] The latter can be confused for right ventricular cardiomyopathy or trigger searches for intracardiac shunts, leading to potential unnecessary tests or treatment.[3] Reduced systolic superior vena cava flow and systolic to diastolic pulmonary vein flow ratios are specific Doppler findings.[2]

Advanced imaging (computed tomography/magnetic resonance imaging) is suggested once CAP is suspected.[1,3] The most consistent finding is interposed lung parenchyma between the aorta and the main pulmonary artery (Fig. 2B).[3] Higher-risk anatomy (eg, partial left CAP) can be excluded, as in our case.[3] Caution should be exercised to avoid overdiagnosis, as the pericardium may not be adequately visualized in patients with minimal epicardial and pericardial fat.[2,3]

Complete bilateral and complete left CAP patients have life expectancy and cardiac function similar to those of controls[1] with no specific treatment.[3] Partial CAP (particularly left-sided) may result in rare complications, including cardiac herniation (most commonly the left atrial appendage), tissue strangulation, coronary compression, and sudden death.[1,2] Clinical suspicion for partial CAP may be heightened by "unique" features including trepopnea (dyspnea lying on one side but not on the other)[3] and abrupt kinking of otherwise normal coronaries on angiography in ischemic patients.[3]

Surgery (pericardiotomy, pericardioplasty, and/or lysis of adhesions) may be warranted for severe symptoms and to treat/prevent serious complications in select patients with partial defects.[1,3] Given these outcomes, a conservative strategy was pursued.

Conclusions

Although CAP is a rare, often benign, congenital anomaly, it merits recognition regardless of age to avoid potentially

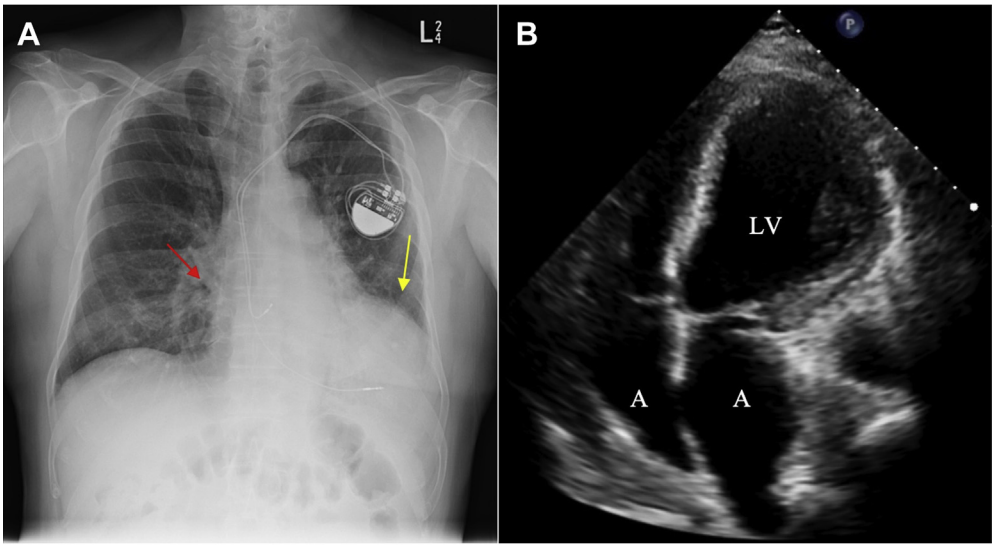

Figure 1. (**A**) Elevated cardiac apex and indistinct cardiac border (**yellow arrow**); loss of the right heart border (**red arrow**). (**B**) bulbous left ventricle (LV) and elongated atria (A).

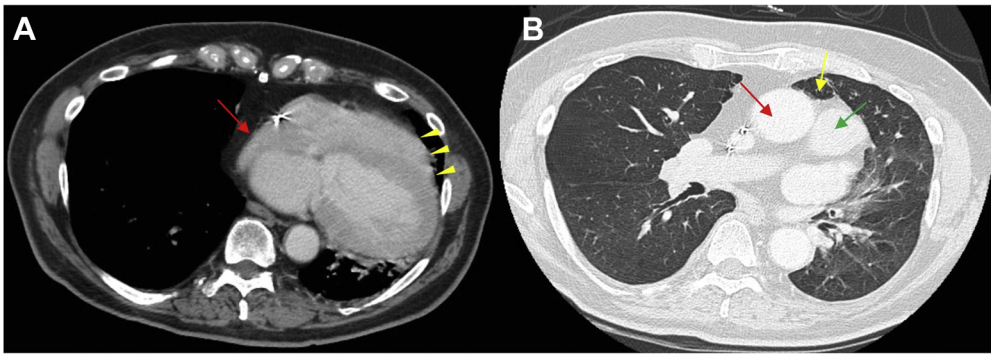

Figure 2. (**A**) Present right pericardium (**red arrow**) and absent left pericardium (**yellow arrowheads**). (**B**) Interposition of lung parenchyma (**yellow arrow**) between aorta (**red arrow**) and pulmonary artery (**green arrow**).

catastrophic consequences. It may also be confounded with other structural heart disease, leading to potentially erroneous treatment. Clinical vigilance and reviewing old data through new lenses are highly recommended when evaluating chronic cardiac complaints.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2020.06.017.