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Review

Congenital absence of the pericardium: case presentation and review of literature

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Abstract

Congenital absence of the pericardium is an uncommon finding that may or may not be symptomatic. Asymptomatic patients are discovered incidentally during cardiac surgery for an unrelated condition or postmortem. However, symptomatic patients may experience non-exertional paroxysmal stabbing chest pain. It may occur with other cardiac or extracardiac abnormalities and a variety of imaging modalities may identify the condition. Complete cases are more rare than partial effects. However, complications are more common with partial absence due to strangulation of the heart into the defect thus requiring surgical intervention.

Keywords: Pericardium; Absence; Congenital

1. Introduction

Congenital absence of the pericardium is an uncommon finding with variable presentations. Asymptomatic patients are discovered incidentally during cardiac surgery for an unrelated condition or postmortem [1]. However, symptomatic patients may experience non-exertional paroxysmal stabbing chest pain [2]. Electrocardiography (ECG), plain chest X-ray (CXR), echocardiography (echo) [1], CT scan [3], and magnetic resonance imaging (MRI) may aid in the diagnosis [4].

2. Case presentation

A 27-year-old healthy male with a history of a murmur presented with mild cough and upper respiratory infection symptoms. He also experienced an occasional brief chest wall throb with exercise that occurred unpredictably and was not associated with chest pain, shortness of breath, or palpitations. His past medical history and family history were unremarkable. He was on no medications and denied illicit drug, tobacco, or alcohol abuse.

On examination, the patient's blood pressure was 110/60 mm Hg and his pulse was 50 bpm with frequent irregularities. There was no jugular venous distension and the lungs were clear to auscultation. There was mild pectus excavatum. Cardiac examination was remarkable for lateral displacement of the apex to the mid-axillary line, with an irregularly irregular rhythm and a grade II/VI systolic murmur heard best at the lower left sternal border with no radiation. The abdomen and lower extremities were unremarkable.

A 12-lead ECG showed a heart rate of 46 bpm with marked sinus arrhythmia poor R wave progression, and an incomplete right bundle branch block (RBBB) (Fig. 1). A chest X-ray revealed levoposition of the heart into the left chest cavity increased visibility of the spine, flattening and elongation of the left ventricular contour (Snoopy sign), and a band of lucency between the heart and diaphragm (Fig. 2). Because of the abnormal chest X-ray and murmur, the patient underwent an echocardiogram. This demonstrated a ''teardrop'' appearance due to elongated atria and relatively bulbous ventricles because of suspension of the heart from its basal pedicle (Fig. 3). Unusual echocardiographic windows and cardiac hypermobility were also present. MRI revealed non-visualization of the pericardium or pericardial fat in the posterior and posterosuperior

Abbreviations: CXR, chest X-ray; ECG, electrocardiogram; Echo, echocardiography; MRI, magnetic resonance imaging; RBBB, right bundle branch block.

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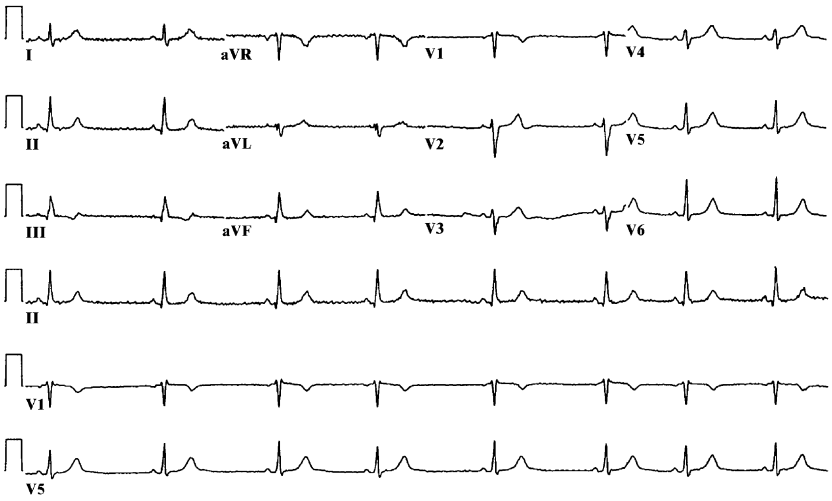

Fig. 1.

aspects of the left ventricle in an area measuring 7.5 cm in the short axis (Fig. 4) and 9.5 cm in the long axis. The patient was diagnosed with congenital absence of the entire left pericardium. Because of mild symptoms, the patient elected a conservative approach.

3. Discussion

Congenital absence of the pericardium is a rare condition that is typically discovered at autopsy or during cardiac surgery [1]. However, a more recent report of 10 cases demonstrated paroxysmal stabbing chest pain as the initial presentation in all cases [2]. Most common is the absence of the entire left side of the pericardium (found in 67% of patients) [1]. Complete absence, partial left absence, and absent right pericardium are very uncommon. Absence of the inferior pericardium is rare in adults, but may be associated with herniation of abdominal organs into the pericardial sac if it is associated with a diaphragmatic defect [5].

Congenital pericardial absence is due to premature atrophy of the left common cardiac vein with insufficient blood supply to the pleuropericardium leading to its agenesis. Male/female ratio is 3:1 [1] and familial occurrence is rare [6]. Thirty percent of patients have associated defects: atrial septal defect, bicuspid aortic valve, patent ductus arteriosus, and tetralogy of Fallot are the most common associated cardiac defects. Extracardiac defects as pulmonary sequestration and bronchogenic cysts may also occur [7]. The

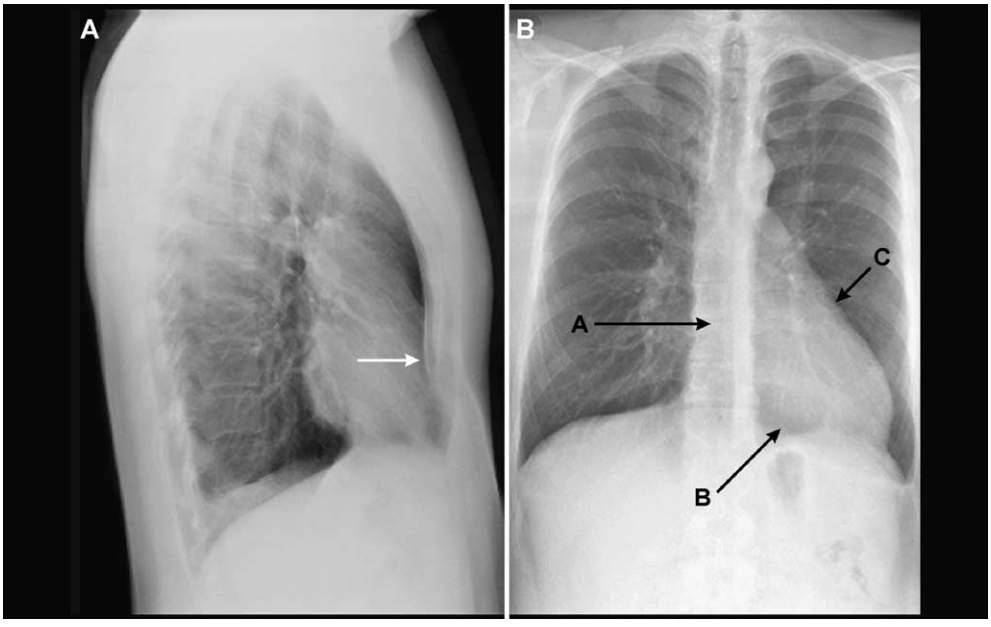

Fig. 2.