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CASE REPORT

Congenital complete absence of pericardium in a young woman with non-specific symptoms

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Image

Abstract Congenital absence of the pericardium is a very rare entity that is usually asymptomatic and hence difficult to diagnose. However, cases of sudden death have been reported in patients with partial pericardial defects (even asymptomatic ones), and such patients require surgical treatment.

We report the case of a 17-year-old patient with complete pericardial agenesis (diagnosed by chance during a cardiological consultation) and briefly review the radiological findings of this entity.

PALAVRAS-CHAVE
Pericárdio;
Ausência;
Tomografia computadorizada de múltiplos detectores;
Ressonância magnética;
Imagem

Ausência congénita completa do pericárdio numa mulher jovem com sintomatologia não específica

Resumo Agenesia congénita do pericárdio é uma entidade muito rara e de difícil diagnóstico, pois na maioria dos casos é geralmente assintomática. No entanto, tem havido relatos de morte súbita em pacientes com agenesia do pericárdio parcial, sendo necessário nestas situações a cirurgia (mesmo em pacientes assintomáticos).

Os objetivos deste trabalho são apresentar o caso de uma paciente de 17 anos com agenesia do pericárdio completo (diagnosticadas por acaso, durante uma visita ao cardiologista) e uma breve revisão dos achados radiológicos desta entidade.

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Introduction

Congenital defects of the pericardium are rare and in most cases asymptomatic.[1–4] They used to be diagnosed as an incidental finding during surgery or autopsy, but the more widespread use of imaging techniques has increased their incidence.[1–4] The most common congenital anomaly is complete absence of the left pericardium, which is usually asymptomatic.[1] However, partial defects can cause symptoms due to ventricular herniation or entrapment and may even cause sudden death, and surgical repair is needed in these cases.[1–4]

We report the case of a 17-year-old woman attended in our institution for non-specific but frequent symptoms, who was diagnosed with congenital complete absence of the pericardium.

Case report

We report the case of a 17-year-old female patient with no relevant medical background, cardiovascular risk factors, or family history of heart disease or sudden death. The patient was referred to the cardiology department of our institution for episodes of dizziness lasting for seconds without spinning sensation, accompanied by cold sweats. Syncope was not present on any occasion and she did not report chest pain, palpitations or dyspnea. Physical examination and vital signs were all normal (blood pressure 102/63 mmHg and heart rate 74 bpm). Lower limbs showed no edema or signs of deep vein thrombosis, and femoral and pedal pulses were preserved. Diagnostic examinations revealed the following:

ECG: sinus rhythm at 71 bpm, PR interval 120 ms, right axis deviation, poor R-wave progression in the precordial leads, inferolateral repolarization abnormalities and negative T waves in V5–V6, II, III, and aVF (Figure 1).
Chest X-ray: displacement of the cardiac silhouette to the left without tracheal deviation, obliteration of the right cardiac border overlapping the spine and imprint of the pulmonary artery (Figure 2).

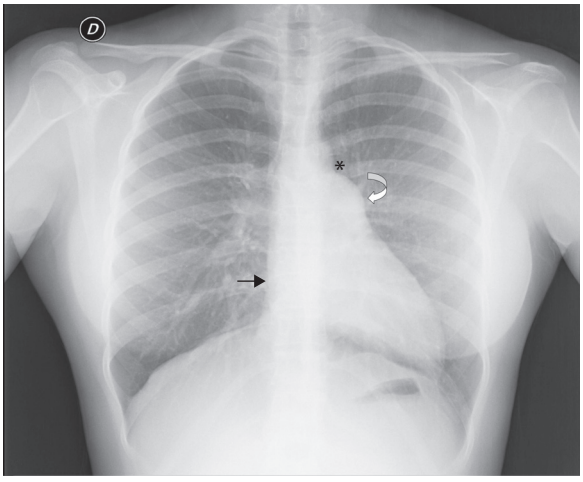

Figure 2 Posterior–anterior chest X-ray showing levoposition of the heart without tracheal deviation, right heart border superimposed on the spine (straight arrow), imprint of the main pulmonary artery (curved arrow), interposition of lung parenchyma between the aortic arch and left pulmonary artery (asterisk) and flattening and elongation of the left ventricular contour (Snoopy sign).

Blood tests: no changes in the various parameters studied.
Holter ECG recording: sinus rhythm with extreme frequencies and circadian variation within normal range; no relevant ventricular or supraventricular arrhythmias, pauses or atrioventricular block.
Echocardiogram: apex offset to the left and posteriorly, particularly noticeable in modified parasternal long-axis view. The left ventricle was non-dilated with preserved systolic function, while the right ventricle was slightly dilated, especially in apical 4-chamber view, forming the bulk of the ventricular apex. The left ventricular filling pattern was normal. The mitral valve opened correctly without a gradient, and tricuspid and aortic valve function was normal. There was mild tricuspid regurgitation with normal pulmonary artery systolic pressure. The inferior vena cava was dilated. The septum showed no alterations and the

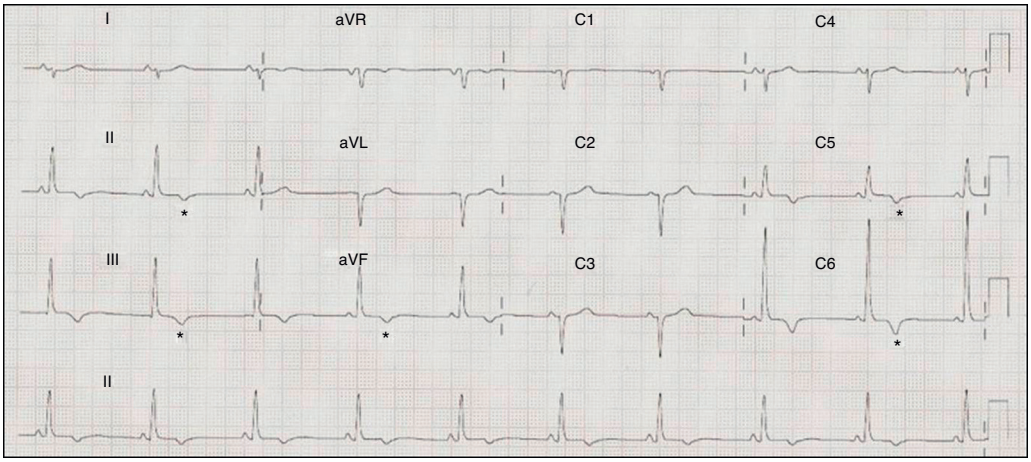

Figure 1 ECG showing sinus rhythm at 71 bpm, PR interval 120 ms, right axis deviation, poor R-wave progression in the precordial leads, inferolateral repolarization abnormalities and negative T wave in V5-V6, II, III and aVF (asterisk).