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Brief Report

Congenital absence of pericardium: a nomad heart

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Abstract Congenital pericardial absence is a rare cardiac defect. Although most patients are asymptomatic, recognising this condition is clinically important as it can cause serious complications. We report the case of an asymptomatic 33-year-old woman seeking medical attention due to a family history of sudden cardiac death. The investigation led us to the diagnosis of congenital absence of the pericardium. The role of imaging in the diagnosis of this rare cardiac malformation is briefly discussed.

Keywords: Congenital absence of percurdium; cardiac magnetic resonance; cardiac anomaly

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Case report

A 33-year-old asymptomatic woman was referred to the cardiology clinic after the sudden death of her brother, whose autopsy revealed a previously undiagnosed hypertrophic cardiomyopathy. Physical examination was unremarkable, except for leftward displacement of the apical impulse. Her 12-lead electrocardiogram showed incomplete right bundle branch block and poor R-wave progression. Chest radiography suggested levoposition of the heart, with no projection of the cardiac silhouette on the right side of the spine, and a band of lucency between the heart and the diaphragm (Fig 1). The transthoracic echocardiogram required the use of atypical lateral views but was considered normal except for the unusual cardiac orientation. Finally, cardiac MRI was performed to confirm the absence of phenotypic signs of hypertrophic cardiomyopathy and ascertain the reason for the other uncommon findings.

Cardiac MRI confirmed the leftward cardiac displacement and showed marked mobility of the heart with respiratory movements, with the cardiac apex pointing laterally and anteriorly in end-inspiration and slightly posteriorly in end-expiration (Fig 2a and b).

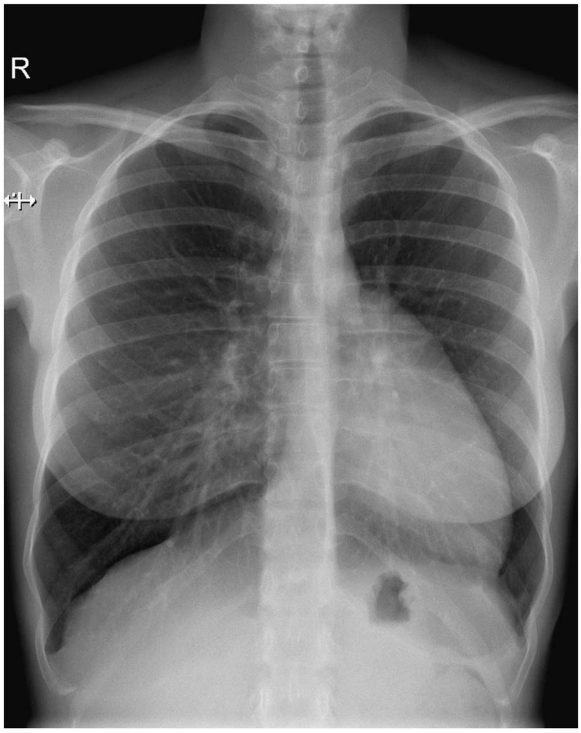

Figure 1.
Chest radiography showing leftward displacement of the heart (with no projection of the cardiac silhouette on the right side of the spine) and a band of lucency between the heart and the diaphragm.

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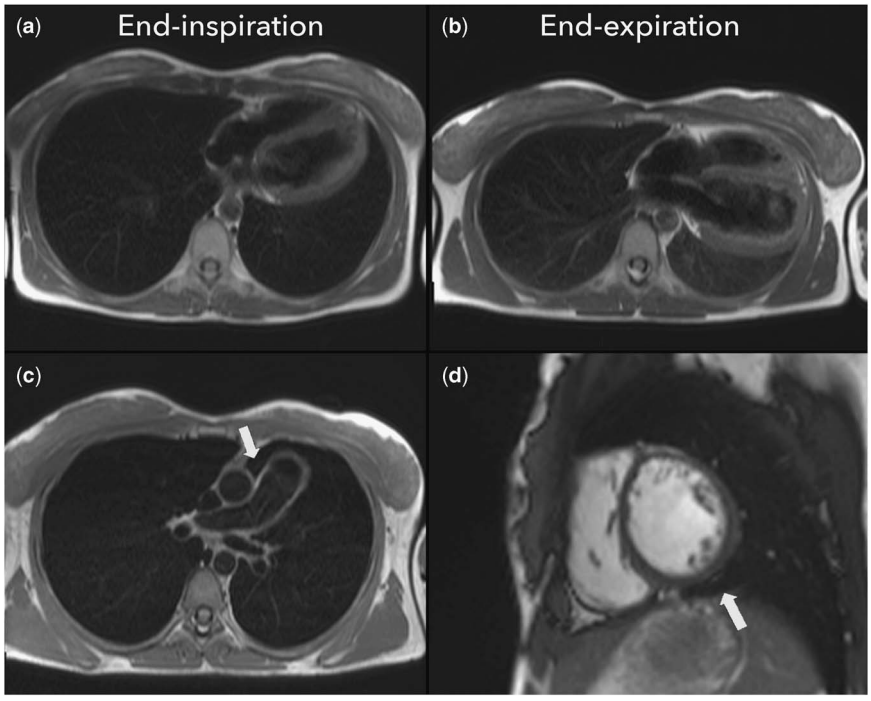

Figure 2.
*(**a** and **b**) Axial T2-weighted half-Fourier acquisition single-shot turbo spin-echo (HASTE) in end-inspiration and expiration, respectively, showing the marked cardiac mobility with respiratory movements; (**c**) axial segmented turbo spin-echo image showing the interposition of lung tissue between the aorta and the main pulmonary artery; (**d**) balanced steady-state free precession (b-SSFP) in left ventricular short-axis view, depicting the presence of lung tissue between the diaphragm and the inferior surface of the heart.*

The parietal pericardium could not be visualised over the left side of the heart. Cardiac MRI depicted the interposition of lung parenchyma between the aorta and the pulmonary artery and between the inferior surface of the heart and the diaphragm (Fig 2c and d), thus establishing the diagnosis of congenital absence of the pericardium. There were no signs of herniation or incarceration of cardiac structures, and no other developmental anomalies were identified. There were no phenotypic signs of hypertrophic cardiomyopathy either. Given the low risk of complications and as the patient was asymptomatic, a conservative approach was adopted.

Discussion

Congenital absence of the pericardium is a rare cardiac anomaly. It was first described in the 16th century and is thought to result from premature atrophy of the left common cardiac vein with insufficient blood supply to the pleuropericardium, leading to its agenesis in the early embryonic stage.[1] Patients are usually asymptomatic, but may sometimes present with positional chest pain, syncope, or even sudden death as a result of cardiac herniation through partial pericardial defects.[2] The chest radiograph and echocardiogram are very useful in suggesting the diagnosis, which is usually confirmed with cardiac MRI or computed tomography. Rather than documenting the absence of the pericardium itself (which can be very challenging in the absence of adjacent fat), these techniques establish the diagnosis by showing indirect signs that are considered pathognomonic of this condition, such as the interposition of lung tissue between the aorta and the pulmonary artery and between the diaphragm and the inferior surface of the heart.[3] Prognosis is usually benign, and most cases are managed conservatively. Surgical pericardioplasty can be considered in highly symptomatic patients or when partial absence of the pericardium leads to herniation or strangulation of the cardiac or vascular structures.[2,4]

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Conflicts of Interest

None.

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