Left-sided congenital absence of pericardium confirmed by decubitus cardiac magnetic resonance imaging

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Accepted: February 4, 2024 Early publication date: February 9, 2024 A 52-year-old man presented with chest discomfort. Physical examination and cardiac serum markers were unremarkable. A 12-lead electrocardiogram showed right axis deviation and an incomplete right bundle branch block (Supplementary material, *Figure S1*). Chest radiography showed a leftward shift of the cardiac silhouette (Figure 1A). Transthoracic echocardiography in the parasternal long-axis view showed that the left ventricular (LV) apex was posteriorly directed (*Video S1*). Electrocardiography-gated cardiac computed tomography (CCT) showed significant stenosis in the proximal segment of the left anterior



Figure 1. A. Chest radiography posteroanterior view showing a leftward shift of cardiac silhouette. **B.** Threedimensional volume-rendered reconstruction of cardiac computed tomography (CCT) showing significant stenosis in the proximal segment of the left anterior descending coronary artery (LAD, white arrow). **C.** Axial supine CCT image showing leftward, posterior positioning of the heart in the thoracic cavity. **D.** Multiplanar reconstruction short-axis CCT image showing the absence of the upper half of the surrounding pericoronary adipose tissue in the mid to distal segment of the LAD (middle: yellow arrow; right: blue arrow), unlike the proximal segment of the LAD, which was completely surrounded by the pericoronary adipose tissue (left: white arrow). Arrow positions correspond to panel **B. E.** Cardiac magnetic resonance imaging (MRI) steady-state free-precession sequence, coronal view in the left lateral decubitus position, showing the heart location adjacent to the left thoracic wall. **F.** Cardiac MRI steady-state free-precession sequence, coronal view in the right lateral decubitus position, showing the heart position recovered to the midline in the thoracic cavity, resulting in interposition of left lung tissue between the heart and left thoracic wall (arrowheads) descending coronary artery (LAD; Figure 1B: white arrow). Notably, CCT showed leftward, posterior positioning of the heart in the thoracic cavity (Figure 1C). CCT did not show lung interposition between the pulmonary trunk and the aortic arch or between the left hemidiaphragm and the base of the heart. CCT multiplanar reconstruction images showed the absence of the upper half of the surrounding pericoronary adipose tissue in the mid to distal segment of the LAD (Figure 1D middle: yellow arrow; right: blue arrow), unlike the proximal segment of the LAD, which was completely surrounded by the pericoronary adipose tissue (Figure 1D left: white arrow). We suspected congenital absence of the pericardium (CAP) and performed cardiac magnetic resonance imaging (MRI). In the left lateral decubitus position, cardiac MRI showed the heart adjacent to the left thoracic wall (Figure 1E; Video S2–S4). Conversely, in the right lateral decubitus position, cardiac MRI showed the heart position recovered to the midline in the thoracic cavity, resulting in interposition of the left lung tissue between the heart and left thoracic wall (Figure 1F, arrowheads; Video S5–S7). Based on this dynamic alteration in cardiac position depending on posture, a definitive diagnosis of complete left CAP was made. Subsequently, percutaneous coronary intervention (PCI) was performed for a 90% stenotic lesion in the proximal site of the LAD. His chest discomfort completely disappeared after PCI.

CAP can be subdivided into complete and partial types based on defect size and site [1]. Complete CAP could be biventricular, left-sided, or right-sided only. Complete CAP is mostly silent and found incidentally, while partial CAP can present with positional chest pain and has a higher risk of serious complications such as herniation of cardiac structure and coronary circulation compression [2]. Complete left CAPs are most common, with a reported prevalence of 70% [1]. A diagnosis of CAP can be made by combining several non-invasive imaging findings. A leftward shift and levorotation of the heart is an imaging finding that can be a diagnostic clue pointing to CAP. Direct visualization of the pericardium is sometimes difficult in the LV lateral and posterior walls even in normal cases, because epicardial and pericardial fat is often negligible or absent in this region [3]. Therefore, lack of visibility of the pericardium on cardiac MRI or CCT does not prove the absence of the pericardium. Positional change is a simple and reliable intervention for echocardiographic diagnosis of CAP [4]. Although the diagnostic usefulness of decubitus imaging in CAP with CCT has been reported, it requires a double radiation dose [5]. In our case, decubitus cardiac MRI demonstrated dynamic alteration in cardiac position that led to a definitive diagnosis of complete left CAP. Decubitus cardiac MRI can provide convincing objective information about CAP without radiation exposure.

Supplementary material

Supplementary material is available at https://journals. viamedica.pl/polish_heart_journal.

Article information

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