A 52-year-old male presented to our facility with dyspnea and dizziness. His past medical history was significant for smoking and hyperlipidemia. An electrocardiogram (ECG) showed a normal sinus rhythm, a rightward axis, low voltage, and poor R-wave progression (Figure 1 A). Transthoracic echocardiography revealed an abnormal atrial-ventricular axis; however, it was difficult to image the heart from the usual parasternal long axis because the apex was shifted laterally and posteriorly and the atria was elongated (Figure 1 B). In addition, the chest x-ray displayed levoposition of the heart with an obscured right heart border, an elongated and flattened left heart border (“Snoopy sign”), and a radiolucency between the main pulmonary artery and aorta (Figure 1 C).

**Figure 1.**
Congenital absence of the pericardium. (A) Electrocardiogram demonstrating rightward axis, low voltage, and poor R-wave progression due to leftward shift of the heart and precordial transition zone. (B) Transthoracic echocardiography, approached apical four-chamber view. Note the lateral position of the apex and the elongated right atrium (teardrop appearance). (C) Posterioranterior chest radiograph showing leftward displacement of cardiac silhouette, causing the right heart borders to be obscured, and an elongated heart border. There is a radiolucent space between the aorta and main pulmonary artery demonstrating interposed lung tissue (arrow). (D) Computed tomography (CT) showing the laterally displaced left ventricular apex. (E) CT showing the interposition of lung tissue between the main pulmonary artery and the ascending aorta (arrow).
Consequently, the patient underwent computed tomography (CT) to determine if there was a congenital absence of the pericardium (CAP). The CT confirmed the leftward displacement of the heart and the absence of the pericardial layer (Figure 1 D). An interposition of lung tissue between the main pulmonary artery and the aorta—a characteristic sign for CAP—could be seen in the axial view (Figure 1 E).

Congenital absence of the pericardium is a very rare disease that can be categorized as a complete or partial absence of the pericardium, with the latter having a higher risk for complications. Often found incidentally, CAP has a nonspecific clinical presentation, with possible symptoms being chest pain, dizziness, and shortness of breath. In our case, the diagnosis was suspected because of the unexplained shifted heart position in the chest.

Although CT and magnetic resonance imaging are the gold standard for diagnosing CAP, certain findings on an ECG, echocardiography, and chest x-ray can also indicate its presence. Nevertheless, the rarity of CAP can make it difficult to detect; therefore, clinicians should familiarize themselves with the different findings that can lead to a prompt diagnosis.

REFERENCES