Congenital Absence of the Pericardium

A 24-year-old asymptomatic man presented for preoperative chest radiography. Posteroanterior chest radiograph (A) shows levoposition of the heart with loss of the right heart border, superolateral displacement of the cardiac apex, and lucency between the heart and diaphragm. Axial contrast-enhanced CT (B) shows no identifiable pericardium or pericardial fat, consistent with congenital absence of the pericardium (CAP).

CAP is a rare diagnosis in which the pericardium fails to form, most commonly due to premature atrophy of its vascular supply. There is a male predominance of 3:1. CAP may be complete or partial with the most common presentation being a left-sided partial absence. Complete CAP is usually clinically insignificant. However, partial absence of the pericardium may present with chest pain or arrhythmia, or be complicated by fatal herniation and incarceration of the ventricles or atrial appendages.1

Chest radiography findings of CAP include levoposition of the heart, a posteriorly and superiorly displaced cardiac apex, and abnormally interposed lung parenchyma. Definitive diagnosis of CAP is made on cardiac CT or MRI and demonstrates three findings: 1) failure to visualize the hypointense pericardial line between myocardium and pericardial fat; 2) levoposition of the heart; and 3) abnormal interposition of lung parenchyma between the heart and diaphragm or in the aorto-pulmonary space. Additionally, transesophageal echocardiogram may disclose paradoxical motion of the interventricular septum, falsely enlarged cardiac chambers due to the shifting heart, and absent systolic separation of the pericardial layers.2 Treatment is surgical and generally reserved for patients who have debilitating symptoms or herniation.

REFERENCES