Congenital Absence of the Pericardium

Subha V. Raman, MD; Curt J. Daniels, MD; Steven E. Katz, MD; James M. Ryan, MD; Mark A. King, MD

A 37-year-old, physically active man presented for evaluation of nonexertional chest pain. He described it as sharp, left-sided, and nonradiating. The review of systems was unremarkable. He had had no previous thoracic surgery. Physical examination revealed a nonpalpable apical impulse, with no other abnormal findings.

A chest radiograph (Figure 1) demonstrated a prominent pulmonary artery shadow, with leftward and posterior rotation of the cardiac apex. Because this configuration can be seen with congenital absence of the pericardium, an ECG-gated cardiac MRI examination was performed to look for pericardial tissue and to rule out other causes of cardiac displacement. Figure 2 shows 3 images obtained using a T1-weighted, double-inversion recovery pulse sequence on a conventional 1.5 Tesla MR scanner (General Electric Medical Systems). Figure 2A was obtained in the axial plane in the lower thoracic region; no pericardium is present over most of the heart, and the characteristic displacement of the heart into the left hemithorax with no structural abnormalities is seen. Figure 2B, which is from a more cranial axial section, demonstrates the pathognomonic presence of lung parenchyma in the usually lung-free aortopulmonary space (arrow). Finally, Figure 2C was obtained in the coronal plane and illustrates the altered relationship of the right and left ventricles to each other and to the spine. The patient has continued his activities without limitation and is not contemplating surgical intervention at this time.

Figure 1. Posteroanterior (A) and lateral (B) chest roentgenograms.
Figure 2. Cardiac MR images in the axial (A, B) and coronal (C) planes.
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