A 19-year-old black mother complained of intermittent episodes of mainly right-sided, sharp, nonpleuritic, anterior precordial chest pain, unassociated with shortness of breath or dyspnea on exertion. No precipitating factors were noted. There was no history of cardiovascular problems. She was well-developed, well-nourished and in no acute distress. Blood pressure was 110/70; pulse 74; temperature 37°C. Cardiac auscultation disclosed no murmurs, with an occasional third heart sound heard over the apex. Peripheral pulses were normal; the extremities revealed no cyanosis, clubbing, or edema.

A presumptive radiological diagnosis can be made at this time (fig. 1).

Figure 1
PA chest radiograph.
Radiological Diagnosis: Partial Absence of the Left Pericardium

Because of the presumptive diagnosis of partial left pericardial defect, angiocardiography and a diagnostic left pneumothorax were performed. The former demonstrated a normal pulmonary artery segment. The levo-phase showed the left atrial appendage occupying the abnormal segment of the left heart border (fig. 2). Its position was more superior than normal and the appendage was dilated in comparison to the size of the left atrium. The degree of herniation through the pericardium varied with the motion of the left atrium. The left ventricle was normal.

After the injection of 450 cc of carbon dioxide into the left pleural space, gas within the pericardial sac defined sharply the herniation of the left atrial appendage through the pericardial defect. A left lateral decubitus study shifted the carbon dioxide into the right pericardial area.

The rent in the pericardium was repaired by surgery (fig. 3).

Discussion

Pericardial defects are three times more common in males than females.3,4 Thirty percent of the reported cases have been associated with other varied congenital anomalies, although these associations may be coincidental. Isolated right-sided defects, total pericardial absence, and diaphragmatic pericardial defects are considered rare.2,5,11 Total absence of the left pericardium, is the most common defect and accounts for 70% of the patients. The partial, or foramen defect described in our patient is less common.

Figure 2

Angiocardiogram demonstrating the left atrial appendage in diastole (arrows), herniated through the left pericardial defect.

Figure 3

Diagram of appearance of the partial left pericardial defect with herniation of the left atrial appendage, as seen at surgery.

The embryological basis of pericardial defects has been debated and well described in the reviews of Sunderland12 and Rusby.13 Premature atrophy of the left duct of Cuvier, which supplies nourishment of the developing pleuro-pericardial membrane, is thought to be the cause of incomplete formation of the left pericardium.14

Complete absence of the left pericardium, which does not decrease life expectancy, presents characteristic radiographic findings.2,16 These findings include a shift of the heart to the left, often with the right heart border overlying, or left of the vertebral bodies. The inferior portion of the cardiac silhouette is flattened and stretched over the dome of the left hemidiaphragm with varying degrees of interposed radiolucency. Three prominences are seen along the left heart border: 1) the aortic knob, 2) an elongated, prominent and clearly identifiable pulmonary artery segment, often with increased translucency between this segment and the aortic knob, and 3) a prominent left ventricular segment. Further evaluation by angiocardiography and/or left pneumothorax is no longer considered necessary for confirmation when these characteristic findings are present.

Recognition of a partial left pericardial defect may be difficult. Although most patients are asymptomatic,
bradycardia, and nonspecific chest pain may be present. Fisher attributes chest pain to the following: 1) undue torsion or strain on great vessels which serve as the only anchor for the heart, 2) lack of a cushioning effect of the pericardium, 3) tension on pleuro-pericardial adhesions, and 4) pressing of the rim of remaining pericardium on cardiac vessels. This latter factor is well-demonstrated in a case report by Lajos, in which the anterior descending coronary artery was partially occluded, and in reports of ischemic changes which may respond to nitroglycerin. Three patients with a partial defect have been reported who have died from ventricular strangulation.

The radiographic appearance of the partial left pericardial defect is less characteristic. The usual case presents no roentgenographic abnormalities. In this event diagnosis is impossible without a simultaneous left pneumothorax. The entity can be suspected only when there is herniation of the left atrial appendage through the defect or when gas enters the pericardial sac secondary to a pneumothorax. The remainder of the cardiac silhouette is normal.

The differential diagnosis of an abnormal mid-left heart contour includes many different possibilities. Hilar lymphadenopathy secondary to infection, lymphoma, metastases, or a primary neoplasm must certainly be considered (fig. 4). Pulmonary artery abnormalities including poststenotic dilatation, idiopathic dilatation, or pulmonary artery aneuysm may all show similar radiographic findings (fig. 5). An atypical thymus located in this position in the mediastinum may occasionally be confusing (fig. 6). The presence of a bronchogenic cyst in this area may or may not be related to a pericardial defect. Finally, a left atrial abnormality such as idiopathic hypertrophy, enlargement secondary to mitral stenosis, or a left-to-right shunt may rarely present a similar radiographic appearance.

In addition to routine chest radiographs, useful
Additional Indexing Words:

Fluoroscopy
Angiocardiography
Pneumothorax

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