288 Pericardial Disease

Eugene Braunwald

NORMAL FUNCTIONS OF THE PERICARDIUM

The normal pericardium is a double-layered sac; the visceral pericardium is a serous membrane that is separated by a small quantity (15–50 mL) of fluid, an ultrafiltrate of plasma, from the fibrous parietal pericardium. The normal pericardium, by exerting a restraining force, prevents sudden dilation of the cardiac chambers, especially the right atrium and ventricle, during exercise and with hypervolemia. It also restricts the anatomic position of the heart, and probably retards the spread of infections from the lungs and pleural cavities to the heart. Nevertheless, *total* absence of the pericardium, either congenital or after surgery, does not produce obvious clinical disease. In *partial* left pericardial defects, the main pulmonary artery and left atrium may bulge through the defect; very rarely, herniation and subsequent strangulation of the left atrium may cause sudden death.

ACUTE PERICARDITIS

Acute pericarditis, by far the most common pathologic process involving the pericardium (Table 288-1), has four principal diagnostic features:

1. *Chest pain* is usually present in acute infectious pericarditis and in many of the forms presumed to be related to hypersensitivity or autoimmunity. The pain of acute pericarditis is often severe, retrosternal, and left precordial, and referred to the neck, arms, or left shoulder. Frequently the pain is pleuritic, consequent to accompanying pleural inflammation (i.e., sharp and aggravated by inspiration and coughing), but sometimes it is steady, constricting, radiates into either arm or both arms, and resembles that of myocardial ischemia; therefore, confusion with acute myocardial infarction (AMI) is common. Characteristically, however, pericardial pain may be relieved by sitting up and leaning forward and is intensified by lying supine (Chap. 19). Pain is often absent in slowly developing tuberculous, postirradiation, and neoplastic, uremic, and constrictive pericarditis.

   The differentiation of AMI from acute pericarditis may become perplexing when, with acute pericarditis, serum biomarkers of myocardial damage such as troponin and creatine kinase-MB rise, presumably because of concomitant involvement of the epicardium in the inflammatory process (an epi-myocarditis) with resulting myocyte necrosis. However, these elevations, if they occur, are quite modest given the extensive electrocardiographic ST-segment elevation in pericarditis. This dissociation is useful in differentiating between these conditions.
2. A *pericardial friction rub* is audible at some point in about 85% of patients with acute pericarditis, may have up to three components per cardiac cycle, is high-pitched, and is described as rasping, scratching, or grating (Chap. 267). It is heard most frequently at end expiration with the patient upright and leaning forward.
3. The *electrocardiogram* (ECG) in acute pericarditis without massive effusion usually displays changes secondary to acute subepicardial inflammation (Fig. 288-1). It typically evolves through four stages. In stage 1, there is widespread elevation of the ST segments, often with upward concavity, involving two or three standard limb leads and V_2 to V_6, with reciprocal depressions only in aVR and sometimes V_1. Also, there is depression of the PR segment below the TP segment, reflecting atrial involvement. Usually there are no significant changes in QRS complexes. After several days, the ST segments return to normal (stage 2), and only then, or even later, do the T waves become inverted (stage 3). Weeks or months after the onset of acute pericarditis, the ECG returns to normal (stage 4). In contrast, in AMI, ST elevations are convex, and reciprocal depression is usually more prominent; these changes may return to normal within a day or two. Q waves may develop, with loss of R-wave amplitude, and T-wave inversions are usually seen within hours *before* the ST segments have become isoelectric (Chaps. 294 and 295).
4. *Pericardial effusion* is usually associated with pain and/or the ECG changes mentioned above, as well as electrical alternans.

TABLE 288-1 CLASSIFICATION OF PERICARDITIS

Clinical Classification

- I. Acute pericarditis (<6 weeks)
  - A. Fibrinous
  - B. Effusive (serous or sanguineous)
- II. Subacute pericarditis (6 weeks to 6 months)
  - A. Effusive-constrictive
  - B. Constrictive
- III. Chronic pericarditis (>6 months)
  - A. Constrictive
  - B. Effusive
  - C. Adhesive (nonconstrictive)

Etiologic Classification

- I. Infectious pericarditis
  - A. Viral (coxsackievirus A and B, echovirus, mumps, adenovirus, hepatitis, HIV)
  - B. Pyogenic (pneumococcus, *Streptococcus, Staphylococcus, Neisseria, Legionella*)
  - C. Tuberculous
  - D. Fungal (histoplasmosis, coccidioidomycosis, *Candida,* blastomycosis)
  - E. Other infections (syphilitic, protozoal, parasitic)
- II. Noninfectious pericarditis
  - A. Acute myocardial infarction
  - B. Uremia
  - C. Neoplasia
    - 1. Primary tumors (benign or malignant, mesothelioma)
    - 2. Tumors metastatic to pericardium (lung and breast cancer, lymphoma, leukemia)
  - D. Myxedema
  - E. Cholesterol
  - F. Chylopericardium
  - G. Trauma
    - 1. Penetrating chest wall
    - 2. Nonpenetrating
  - H. Aortic dissection (with leakage into pericardial sac)
  - I. Postirradiation
  - J. Familial Mediterranean fever
  - K. Familial pericarditis
    - 1. Mulibrey nanism[a]
  - L. Acute idiopathic
  - M. Whipple's disease
  - N. Sarcoidosis
- III. Pericarditis presumably related to hypersensitivity or autoimmunity
  - A. Rheumatic fever
  - B. Collagen vascular disease (systemic lupus erythematosus, rheumatoid arthritis, ankylosing spondylitis, scleroderma, acute rheumatic fever, granulomatosis with polyangiitis [Wegener's])
  - C. Drug-induced (e.g., procainamide, hydralazine, phenytoin, isoniazid, minoxidil, anticoagulants, methysergide)
  - D. Postcardiac injury
    - 1. Postmyocardial infarction (Dressler's syndrome)
    - 2. Postpericardiotomy
    - 3. Posttraumatic

[a]An autosomal recessive syndrome characterized by growth failure, muscle hypotonia, hepatomegaly, ocular changes, enlarged cerebral ventricles, mental retardation, ventricular hypertrophy, and chronic constrictive pericarditis