Clinicalpathologic Description of Myocarditis

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Histologic evidence of myocarditis was demonstrated in 35 of 448 patients submitted to endomyocardial biopsy over 5 years. Analysis of the histologic findings and clinical course of these patients resulted in a new clinicopathologic classification of myocarditis in which four distinct subgroups are identified. Patients with fulminating myocarditis became acutely ill after a distinct viral prodrome, have severe cardiovascular compromise, multiple foci of active myocarditis by histologic study and ventricular dysfunction that either resolves spontaneously or results in death. Patients with acute, chronic active and chronic persistent myocarditis have a less distinct onset of illness.

Patients with acute myocarditis present with established symptoms. Idiopathic myocarditis is an inflammatory disease of the myocardium of unknown etiology. Although the clinical and histopathologic features of the disease have been extensively studied, a unifying characterization of the disease has failed to emerge. Historically, cases of subacute (11), lethal (4-10) and progressive (1,4-6) myocarditis have been observed and the disease has been variously described utilizing electrocardiography (2), echocardiography (7,12), serologic studies (13,14) or endomyocardial biopsy (4,8,13,15). In an effort to provide uniform criteria for the pathologic diagnosis of myocarditis, a panel of cardiac pathologists developed a classification of this disease based on histologic features of endomyocardial biopsy specimens. Known as the Dallas criteria (16), this system has been criticized for interobserver variability (17) and may be subject to sampling error (1,18). The failure to develop a clinical description of myocarditis to accompany the pathologic classification has impaired the development of therapeutic trials and fostered controversy as to the very existence of the disease (18).

The purpose of this study was to identify the clinical spectrum of myocarditis and to categorize this disease into four subgroups. This classification is supported by animal models and in humans by clinical and pathologic experience and, in addition, is analogous to the accepted classification of viral hepatitis (19,20). It is hoped that recognition of the clinical substrates of histologically documented myocarditis will allow a better understanding of the anticipated course of patients with this disease. Then, as in hepatitis (20), we may better define an individual patient's suitability for immunosuppressive therapy.

Methods

Study patients. Between December 1, 1983 and July 1, 1988, 448 patients underwent diagnostic endomyocardial biopsy to evaluate cardiac dysfunction. On histologic analysis, 60 patients (17,25) exhibited active or borderline myocarditis as defined by the Dallas criteria (16).

Two separate classifications are used for the first and subsequent biopsies. On the first biopsy, active myocarditis is defined by myocardial necrosis or degeneration, or both, associated with an inflammatory infiltrate adjacent to the degenerating or necrotic myocytes. Borderline myocarditis is diagnosed when the inflammatory infiltrate is too sparse or when damage to myocytes is not demonstrable. No myocarditis implies that the myocardium is either entirely normal or shows nonspecific changes.

On subsequent biopsies, angione myocarditis means that myocyte damage or necrosis in association with inflammatory infiltrate persists. In resolving myocarditis, the inflammatory infiltrate is substantially reduced and is not ini-