Acute Heart Failure due to Fulminant and Giant Cell Myocarditis

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Abstract

Acute or fulminant nonischemic, dilated cardiomyopathy (DCM) is an uncommon cause for heart failure with a highly variable prognosis that is in part dependent on histopathology and in part on clinical presentation. Once common causes of acute DCM are excluded using standard clinical tests, the specific inflammatory and infectious causes of DCM should be systematically evaluated and treated. Specific histopathologic forms include fulminant lymphocytic myocarditis, which has an excellent prognosis with standard heart failure care. By contrast, giant cell myocarditis, which may be fulminant or acute, has a poor prognosis and frequently requires heart transplantation or immunosuppression for long-term survival. Noninvasive tests that may support the diagnosis of fulminant or acute myocarditis include cardiac magnetic resonance imaging and biomarkers of cardiac injury. Certain clinical, hemodynamic, and echocardiographic variables predict risk of death or transplant in acute myocarditis. This article will compare the presentation, prognosis, and treatment options for several uncommon causes for acute heart failure and suggest certain clinical scenarios in which the likelihood of specific histopathologic disorders is high enough to warrant endomyocardial biopsy if noninvasive tests are inconclusive.

Akute Herzinsuffizienz bei fulminanter Myokarditis und Riesenzellmyokarditis

Zusammenfassung


Introduction

The syndrome of acute heart failure is frequently associated with a newly diagnosed dilated, hypokinetic left ventricle (DCM) with or without right ventricular dysfunction. Common causes of acute DCM that should be excluded by medical history, physical examination, electrocardiography, echocardiography, and sometimes coronary angiography include acute ischemia, heart rhythm-related, pericardial, valvular, peripheral vascular, congenital, and familial disorders [1]. A careful medical history often reveals that acute heart failure turns out to actually be an acute manifestation of a chronic cardiomyopathy. Once the common disorders are excluded, specific inflammatory and infectious causes for DCM should be considered. These disorders include acute viral myocarditis and a number of idiopathic myocarditides such as the eosinophilic disorders, giant cell myocarditis (GCM), cardiac sarcoidosis (or idiopathic granulomatous myocarditis), and acute lymphocytic myocarditis [2]. Although relatively little is known about the pathogenesis of myocarditis, descriptive clinical studies have revealed a strong correlation between histopathologic forms of myocarditis and prognosis [3].

Fulminant Lymphocytic Myocarditis

Fulminant lymphocytic myocarditis (FLM, Figure 1a) is a clinicopathologic entity defined by the sudden onset of heart failure within 2–3 weeks of a distinct episode of a viral illness [4]. The degree of left ventricular dysfunction is usually severe, and the
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Acute myocarditis is lacking. Since the risks of per-viral clearance in humans following fulminant, versus clearance is an attractive paradigm, data to document response. Although the concept of successful viral trials (in adults) aimed at altering this acute immune inating the virus from the heart. This may be one ex-

In the clinical setting of FLM, the inflammation that contributes to heart failure may be beneficial by elim-

Amabile et al. recently reported a case series of eleven children with FLM [7]. The median age was 1 year (range 0–9 years), and the mean left ventricu-
lar ejection fraction (LVEF) was 22%. A virus was identified in five patients: parvovirus B19 (n = 2), Epstein-Barr (n = 1), varicella zoster (n = 1), and cox-

The etiology of FLM and the reasons for the ex-

The role for immune therapy with steroids and/or intravenous immunoglobulin in children has not been assessed in controlled trials, but uncontrolled case series suggest an association with good long-term prognosis. EMB may exclude the uncommon and sometimes clinically similar disorders of necrotizing eosinophilic myocarditis (NEM) and GCM which usually respond to the specific therapies discussed below. One important limitation of EMB for lymphocytic myocarditis is the low sensitivity that is likely due to patchy involvement of the myocardium and variability in interpretation [9–11]. Therefore, a normal or nondiagnostic biopsy does not exclude lymphocytic myocarditis.

In the setting of nondiagnostic findings on EMB, cardiac magnetic resonance imaging (CMR) can be useful to guide EMB to areas of active inflammation [12] and to distinguish ischemic from nonischemic acute cardiomyopathy. Laissy et al. prospectively ex-

Subendocardial delayed enhancement on CMR is not typically seen in acute myocarditis but may be seen in myocarditis patients who have fully recovered after fulminant myocarditis for the sole purpose of determining viral clearance seems not justified, we will likely never know why patients with fulminant myocarditis have such a good prognosis.

However, EMB at the time of presentation in suspected FLM is indicated for two reasons. If lymphocytic myocarditis is present, most adult patients seem to improve with standard care and no immuno-suppression. The role for immune therapy with ste-
roids and/or intravenous immunoglobulin in children has not been assessed in controlled trials, but uncontrolled case series suggest an association with good long-term prognosis. EMB may exclude the uncommon and sometimes clinically similar disorders of necrotizing eosinophilic myocarditis (NEM) and GCM which usually respond to the specific therapies discussed below. One important limitation of EMB for lymphocytic myocarditis is the low sensitivity that is likely due to patchy involvement of the myocardium and variability in interpretation [9–11]. Therefore, a normal or nondiagnostic biopsy does not exclude lymphocytic myocarditis.

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