Giant Cell Myocarditis Manifested as Fulminant Myocarditis

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ABSTRACT

A 42 year-old female with fulminant myocarditis became acutely ill following a distinct viral prodrome 2 weeks before her hospitalization. She had severe ventricular dysfunction and multiple foci of giant cell myocarditis on the histopathological study. She experienced severe hemodynamic compromise that required high-dose vasopressors, immunoglobulin and extracorporeal membrane oxygenation (ECMO). Although she was managed with aggressive pharmacologic and mechanical support and also heart transplantation, she died at hospital day 12. (Korean Circulation J 2006;36:159–161)

KEY WORDS : Myocarditis ; Giant cell myocarditis.

Introduction

Giant cell myocarditis (GCM) is a rare and fatal form of myocarditis, and is characterized by diffuse myocardial necrosis with multinucleated giant cell. Although it is suspicious that GCM may be a sequel of viral infection, the relationship between GCM and a certain viral infection has not been documented yet. Here, we report a GCM, manifested by fulminant myocarditis (FM) whose myocardial biopsy demonstrated severe myocardial necrosis and multinucleated giant cells.

Case

A 42-year-old woman was admitted because of recurrent syncope. Two weeks before, she had mild fever for 5 days. Four days earlier, she had palpitation, chest discomfort and dizziness. At emergency room, cardiac enzymes were elevated. Coronary angiogram showed no thrombus and no significant stenosis, however, recurrent ventricular tachycardia was noticed during angiography. Mechanical circulatory support was started with an extracorporeal membrane oxygenation (ECMO) because of recurrent ventricular tachycardia and progressive cardiogenic shock. At hospital day 5, thrombectomy was done because of huge thrombus that obstructed mitral valve inflow. Biopsy specimen of left atrium during thrombectomy, showed severe inflammation and diffuse myocardial necrosis with multinucleated giant cells (Fig. 1). Despite 208 hours of ECMO support and intravenous immunoglobulin (500 mg/kg/day) for 3 days, left ventricular function didn’t recover. Finally, she received heart transplantation at day 10. In the explanted heart, severe myocardial necrosis and inflammation with giant cells were found in all 4 chambers (Fig. 2). She died at day 12 because of renal failure and metabolic acidosis.

Discussion

GCM is a rare and fatal disorder of an unknown origin with no proven treatment. Although there are only case reports of acute viral myocarditis in Korea and a few recent reports clearly saying that FM is associated with coxsackievirus type B3 (CVB3), adenovirus, the causative virus of FM has not been well clarified.

The clinical course of patients with GCM varies; some patients present acutely or with a fulminant disease which progress to heart failure, ventricular arrhythmias, or heart block and others present as dilated cardiomyopathy with late deterioration. Lieberman et al. classified myocarditis as either fulminant or acute (nonfulminant) on the basis of clinicopathological criteria, including the severity of the illness on presentation. Patients with FM become worse rapidly after a history of distinct viral illness, have cardiovascular collapse, fin-
Findings of active myocarditis on histological study and ventricular dysfunction, which either resolves spontaneously or results in death. Paradoxically, patients with FM, although more severely ill on presentation, were more likely to recover left ventricular function than those with acute myocarditis. This is a very rare case that severe myocardial necrosis and inflammation with giant cells were found in all 4 chambers of the explanted heart.

Heart transplantation (HTL) is the treatment of choice for GCM. HTL has been used with acceptable morbidity and mortality as primary therapy for the management of GCM. However, since the risk of death during the acute phase is high, a bridge to recovery with circulatory support devices is often useful in FM patients. Regardless of the type, early mechanical circulatory support (MCS) helps save life and prevents multi-organ failures in patients with FM. Because other management with immunoglobulin, antiviral agent or monoclonal antibody remain to be confirmed, aggressive hemodynamic support with MCS is considered as the best management for patients with FM.8)

Fig. 1. Massive myocardial necrosis, inflammation (A, ×200) and multinucleated giant cells (B, ×1000) were identified in left atrium at day 5 (hematoxylin and eosin).

Fig. 2. Pathology of explanted right atrium, right ventricle, left atrium, and left ventricle at day 10 (A through D, ×400).
REFERENCES


