Case report - Transplantation

Acute myocarditis supported by extracorporeal membrane oxygenation successfully bridged to transplantation: a giant cell myocarditis

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Abstract

Giant cell myocarditis is a rare and fatal heart disease in previously healthy young patients. We report the case of a 43-year-old patient presenting unstable acute congestive heart failure as a consequence of myocarditis who was supported for four days by an extracorporeal membrane oxygenation. While no cardiac recovery was observed and no viral and autoimmune causes of myocarditis were found, he underwent successful orthotopic heart transplantation in emergency. Giant cell myocarditis was diagnosed on the explanted heart. The patient has been on a triple-immunosuppression therapy with no signs of recurrence of the disease or rejection 16 months after surgery. This experience is compared with published cases and implication of diagnosis and treatment are discussed.

Keywords: Myocarditis; Mechanical cardiac support; Heart transplantation

1. Introduction

Giant cell myocarditis (GCM) is a rare form of myocarditis of unknown origin. The disease usually occurs in previously healthy young people and is typified by rapidly progressive cardiac dysfunction, often requiring cardiac transplantation because of its poor prognosis. We describe a patient who had rapidly progressive heart failure and needed circulatory support by extracorporeal membrane oxygenation (ECMO). The outcome was favorable after transplantation. GCM was diagnosed from the explanted heart and a review of the literature is discussed.

2. Case report

A 43-year-old previously healthy man was admitted to a general hospital with a 2-week history of breathlessness, fatigability and abdominal pain. He was first diagnosed for a cholecystitis and underwent a cholecystectomy. Postoperatively he presented unstable hemodynamics and respiratory failure and was therefore transferred to our hospital for diagnosis. He had no chest pain, but had marked shortness of breath. Arterial oxygen saturation was 90% under 10 l/min nasal O2, blood pressure was 90/65 mmHg with a regular heart rate of 120 beats/min. An electrocardiogram (ECG) showed a right bundle branch block with lateral ST modification. An echocardiogram showed a globally hypokinetic heart and a left ventricular ejection fraction of 10%. The coronarography was normal and the angiography confirmed a poor left ventricular ejection fraction. He was diagnosed for acute heart failure, initially stabilized by medical therapy, which secondarily deteriorated to cardiogenic shock. Troponin level remained in normal range whereas serum transaminases and BUN were elevated (alanine aminotransferase was seven times the normal range, aspartate aminotransferase was eight times the normal range and serum BUN concentration was under twofold the upper limit of the normal range). It was therefore decided to implant a peripheral ECMO by femorofemoral cannulation, which permitted to stabilize hemodynamic conditions while peripheral organ function returned to normal range. However, no cardiac recovery was observed within the first four days of circulatory assistance with a mean flow rate of 3.07 l/min. Endomyocardial biopsies performed during ECMO implantation were irrelevant. Also, autoimmune and viral causes of myocarditis were investigated and remained negative. Therefore, chances to recover were thought to be small and because hepatic and renal functions began to deteriorate, it was decided to transplant the patient in emergency rather than bridge to another assist device.

The histological diagnosis was confirmed to be GCM (Fig. 1). The postoperative immunosuppression did not differ from our other cardiac transplant patients and consisted of

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Giant cell myocarditis was first described by Saltykow in 1905 [1] and is histologically characterized by diffuse intramyocardial inflammatory infiltrate consisting of lymphocytes, eosinophils and plasma cells, diffuse myocardial necrosis with multinucleated giant cells, as shown in our case on Fig. 1 [2]. GCM can be idiopathic when any association with another illness is found, or can be secondary to other diseases with a myocardial granulomatous component such as sarcoidosis, mycobacteria or fungal infections and autoimmune diseases. Differentiation of idiopathic GCM from cardiac sarcoidosis might be challenging and perhaps these two entities represent two ends of a disease spectrum as suggested by Stoica et al. [3].

Several experiments suggest that GCM is an autoimmune disorder dependant on CD4-positive T lymphocytes [4]. Until 1996, the multicenter Giant Cell Myocarditis Registry recorded 63 cases from 36 centers. Median survival of such patients was 5.5 months from the onset of symptoms until death or transplantation [2].

GCM is an uncommon disorder which typically affects young to middle-aged adults, with a slight male preponderance [2,5,6]. Congestive heart failure is the most common cardiac presentation. On occasion, the initial manifestations may be quite dramatic, including rapidly progressive hemodynamic deterioration, intractable arrhythmias and sudden death. Although the early symptoms of GCM may not be different from other types of acute myocarditis, the condition progresses rapidly to death, often within days to months. Because of this fatal nature of the disease, early right ventricular EMB in previously healthy patients presenting unexplained acute heart failure should be performed to allow accurate diagnosis and adapted treatment with immunosuppression or heart transplantation [2,5,7]. They must be repeated because GCM which may be focal can be undiagnosed if EMB are performed just once, as demonstrated by our case.

Several treatment options have been described in the literature [2,5-8]. Immunosuppressive therapy with regimens including cyclosporine, azathioprine or both, but not corticosteroids alone, may be considered but only prolongs the time to transplantation. Indeed, survival of patients presenting GCM and receiving a double immunosuppressive therapy is less than one year [2]. The rapid and precipitous clinical deterioration into congestive heart failure can be bridged to transplantation with mechanical circulatory support, as demonstrated by our case using a peripheral ECMO. ECMO permits to stabilize the patient regarding hemodynamic conditions and peripheral organ perfusion during a short period of time. It is also less expensive than mechanical assist devices such as Thoratec pVAD or Cardiowest TAH used by Davies et al. in patients presenting GCM myocarditis who were further transplanted within 2 to 53 days after implantation [8]. In case of no myocarditis etiology being found by EMB and if no heart recovery is observed within the first 7 to 10 days after ECMO implantation, the patient can be either bridged to another mechanical assist device or transplanted in emergency. We preferred to transplant our patient even only after four days of ECMO circulatory support for several reasons: first, because no cardiac recovery was observed and no etiology (EMB, autoimmune and viral causes) was found; second, because hepatic and renal functions began again to deteriorate while the patient was under ECMO, requiring a bridge to a mechanical assist device. However, a ‘Super Emergency’ heart allocation program is dedicated in France to patients who are unstable under intravenous inotropes or ECMO and who therefore should be bridged to a long-term mechanical assist device, which we decided to use in this particular case.

Heart transplantation may remain the only possibility for prolonged survival, which is almost similar to the overall survival rates for ischemic or dilated cardiomyopathy patients [5,7,8]. The recurrence of GCM in cardiac transplants has been described in only 5 cases with good response to increased immunosuppression, which differs dramatically from that in the native heart [9]. The post-transplantation care of these patients should probably be different from other heart transplant patients with a more frequent follow-up maintained indefinitely to assess recurrence of GCM.

4. Conclusion

GCM is a rare and fatal disorder presenting as acute congestive heart failure. Because of the fatal nature of this disease, early and repeated endomyocardial biopsies in previously healthy patients presenting unexplained acute heart failure should be performed to allow accurate diagnosis and adapted treatment. In case of a rapid clinical deterioration, ECMO can be safely used as a bridge to transplantation which remains a reliable therapy despite the risk of post-transplantation recurrence of GCM requiring a more frequent follow-up after transplantation maintained indefinitely.

Fig. 1. Histological aspect of the explanted heart (hematoxylin and eosin, original magnification ×100). 1: mononuclear cell infiltrate, 2: giant cells, 3: degenerative cardiomyocytes.
References