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CASE REPORT

Total artificial heart implantation for biventricular failure due to eosinophilic myocarditis

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Abstract Idiopathic hypereosinophilic syndrome is a condition of unknown etiology characterized by proliferation of eosinophils and their infiltration into tissues. Although cardiac involvement is rare, eosinophilic myocarditis can lead to life-threating fulminant congestive heart failure. Treatment of patients with eosinophilic myocarditis is challenging as heart failure can be caused by biventricular dysfunction. To our knowledge, this is the first case reported in the literature describing a patient with acute severe biventricular heart failure caused by eosinophilic myocarditis with mural left ventricular apical thrombus who was successfully treated with implantation of a total artificial heart as a bridge to heart transplant.

Keywords Heart failure · Circulatory support device · Cardiomyopathy · Artificial organs

Introduction

Idiopathic hypereosinophilic syndrome is a condition of unknown etiology characterized by infiltration of eosinophils into the tissues [1]. Although rare, cardiac tissue can be involved; eosinophilic myocarditis may result in life-threatening congestive heart failure, accounting for 33–43% of deaths seen in patients with this syndrome [2]. In this report, we describe a patient with acute biventricular heart failure caused by eosinophilic myocarditis who was successfully treated with implantation of a total artificial heart (TAH) as a bridge to heart transplant.

Case report

A previously healthy 20-year-old man presented with worsening dyspnea, decreased appetite, fatigue, and epigastric discomfort for 9 days. Physical examination showed bilateral lower extremity edema. The patient's blood pressure was noted as 92/70. Electrocardiogram revealed normal sinus tachycardia with V5 and V6 ST-segment depression and left axis deviation. His leukocyte count was 8900/μL, and the eosinophil count was moderately elevated at 1602/μL, constituting 18% of the overall leukocyte count. Plasma levels of creatinine kinase, troponin I, and brain natriuretic peptide were increased to 1.46 mg/dL, 0.33 U/L, and 1654 pg/mL, respectively. Transthoracic echocardiography showed a normal-size left ventricle chamber with a large left ventricular apical mural thrombus, measuring 2.0 cm in thickness (Fig. 1). The ejection fraction was 35–40%, and the right ventricle was dilated, measuring up to 47 mm, with severely decreased systolic function. The right atrial pressure was 20 mmHg. Severe eccentric functional mitral regurgitation and moderate tricuspid regurgitation were noted. Computed tomography scan showed an enlarged left and right atrium and right ventricle. A moderate volume of ascites was also seen. The intrahepatic inferior vena cava was distended with dilated hepatic veins. The patient was admitted with a diagnosis of acute biventricular heart failure of unknown origin. The right heart catheter exam

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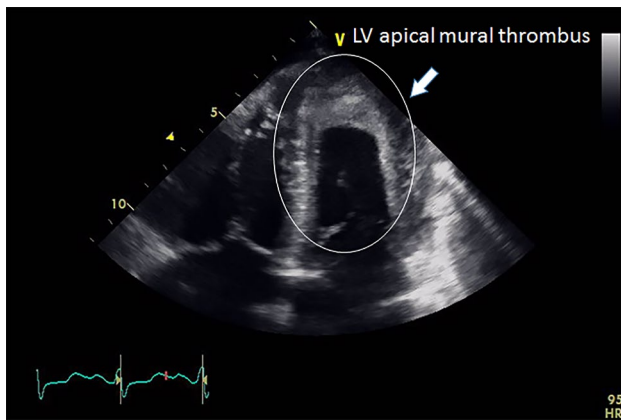
Fig. 1 Preoperative transthoracic echocardiogram shows a large apical mural thrombus in the left ventricle (LV), reducing the size of the chamber

revealed increased pulmonary capillary wedge pressure, pulmonary artery pressure, right ventricular pressure, and central venous pressure [33 mmHg, 68/36 (50) mmHg, and 66/10 (25) mmHg, 20 mmHg, respectively]. Cardiac index was significantly reduced to 1.18 L min/m^2, and pulmonary vascular resistance was increased to 7.35 wu. No stenosis was seen on coronary angiogram. Endomyocardial biopsy was performed; the results showed severe endocardial fibrosis with eosinophilic infiltration, and the endocardium was severely thickened and rich in elastic fibers (Fig. 2). The bone marrow showed trilineage hematopoiesis and increased eosinophils. Blast cells were not increased, and there were no abnormal lymphoid or mast cell infiltrates that would suggest leukemia, lymphoma, or mastocytosis. The clonality of the eosinophils was excluded with the use of molecular studies. Furthermore, laboratory screening tests for the following infectious agents were negative: *Coccidiodes immitis, Trypanosoma cruzi, Strongyloides stercoralis, Toxoplasma gondii*, Epstein–Barr virus, hepatitis

A virus, hepatitis B virus, cytomegalovirus, varicella zoster virus, *Treponema pallidum* (syphilis), *Mycobacterium tuberculosis*, and ova and parasites. The eosinophilia was successfully treated with steroids. Despite maximal medical inotropic support, decompensated biventricular heart failure developed in the patient. As a result, a TAH (SynCardia, Syncardia Systems, Inc., Tucson, Arizona, USA) was implanted as a bridge to transplant after biventriculectomy. The intraoperative lung biopsy showed no evidence of eosinophilic infiltration. The patient's postoperative course was uneventful, and he was extubated on postoperative day (POD) 1. He was ambulatory and discharged home on POD 37 after receiving complete instructions regarding the TAH. At the 6-month outpatient follow-up, he had no major complications, and his creatinine kinase had improved to normal levels (0.77 mg/dL). Physical examination showed no edema in his lower extremities, and a computed tomography scan showed no evidence of ascites. His pulmonary hypertension improved (21/10 mmHg).

On histologic examination, the heart showed left ventricular mural scarring, and the thick endocardial thrombus reduced the size of the lumen of the left apical ventricle approximately (Fig. 3). In addition, hypertrophic changes were noted in cardiomyocytes, and dense mural myocardial replacement fibrosis was seen (Fig. 4).

Discussion

Patients with hypereosinophilia usually have an eosinophil count of more than 1.5×10^9/L (1500/μL) [normal, $<0.5 \times 10^9$/L (500/μL)] [3]. However, hypereosinophilic syndrome is diagnosed when there is evidence of end organ damage due to eosinophilic infiltration, with no identifiable cause such as parasitic infection, drug use, connective tissue disorders, vasculitis, malignancy, or allergies [4]. Almost all organs are prone to eosinophilia-associated
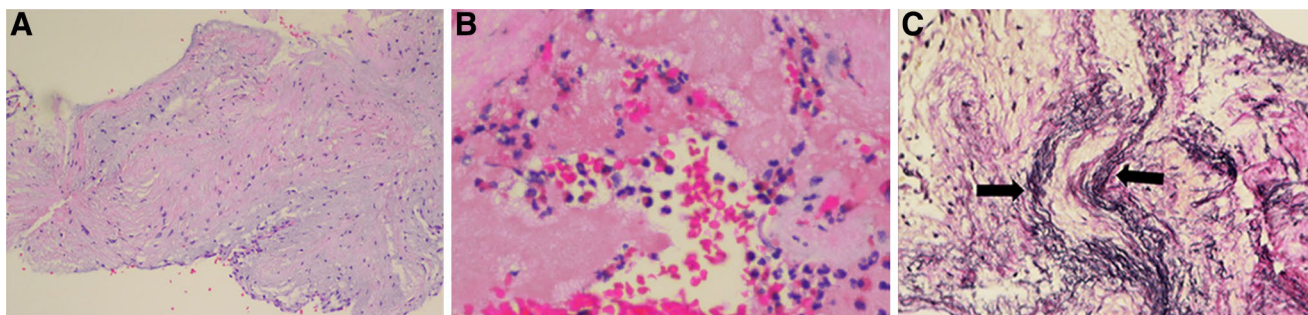

Fig. 2 Endomyocardial biopsy findings. **a** Hematoxylin and eosin stain showing adherent fibrin thrombus containing eosinophils. **b** Photomicrograph of hematoxylin and eosin stained-section under high-power magnification showing eosinophils in the fibrin clot. **c** Verhoff's Van Gieson stain for elastic fibers confirming the presence of excessive elastic fibers (*dark purple* strands indicated by *arrows*) in the region of severe endocardial thickening
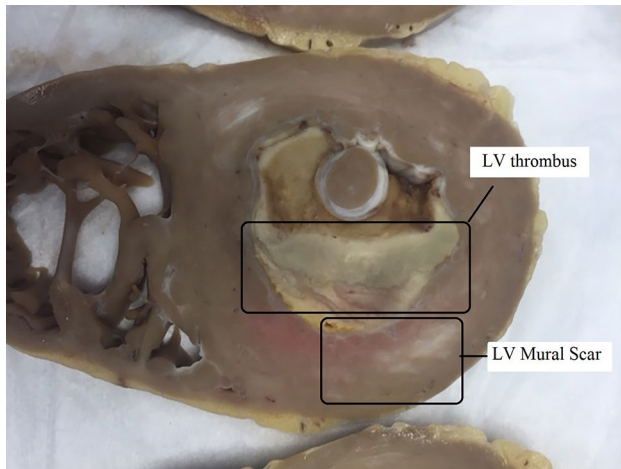
Fig. 3 Gross specimen of the left ventricle (LV) shows that an organizing thrombus in the left ventricular chamber reduced the size of the lumen. Left ventricular mural scarring is seen
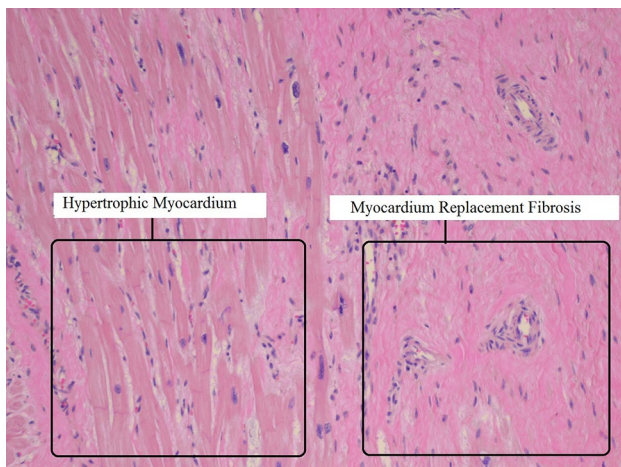
Fig. 4 Hematoxylin and eosin staining of the left ventricular myocardium shows the presence of hypertrophic cardiomyocytes in the left ventricle (LV). Dense mural myocardial replacement fibrosis is seen

damage, but the heart, nervous system, skin, and respiratory tract are most frequently targeted [5].

In eosinophilic myocarditis, there are three stages of cardiac involvement. The first is an acute necrotic stage caused by myocardial infiltration of eosinophils; eosinophilic granules form in the endocardium and myocardium, degranulate, and release toxic proteins, leading to myocardial injury. However, patients are generally asymptomatic in this stage. The second or intermediate phase, called the thrombotic stage, is characterized by the development of mural thrombi as well as thrombus formation along the damaged endocardium; patients usually become symptomatic in this phase. The third stage is the fibrotic phase in which granulation tissue undergoes fibrosis. Only minimal deposition of eosinophils is found in the myocardium during this last phase [1, 5].

Our patient was a previously healthy 20-year-old man who had worsening symptoms over the 9 days before presentation. We made the diagnosis of eosinophilic myocarditis based on the results from laboratory studies and endomyocardial biopsy. The laboratory data showed a significantly elevated eosinophil count, and the endomyocardial biopsy specimen revealed endocardial fibrosis with eosinophilic infiltration. Medical history and screening tests eliminated medication side effects, allergic responses, infection, hematologic disease, malignancy, connective tissue disorders, and endocrinopathy as the cause of the eosinophilia. Treating the eosinophilia with steroids decreased the eosinophil count to normal levels. Despite being on maximal medical therapy, the patient required immediate biventricular mechanical circulatory support because of acute decompensated biventricular function with evidence of moderate ascites and bilateral extremity edema. Heart transplantation was not indicated at that time because of severe pulmonary vascular resistance and pulmonary hypertension.

Right ventricular failure in the early postoperative period after left ventricular assist device (LVAD) implantation is a major cause of morbidity and mortality [6–8]. The patient had multiple preoperative independent risk factors predictive of postoperative right ventricular failure after LVAD support as indicated in previous reports [6–8]. Because of the estimated high mortality from postoperative right ventricular dysfunction, we chose biventricular mechanical support over LVAD use. In general, the use of a biventricular assist device has the advantage over a TAH in sparing the ventricles for potential recovery of function. In our case, the whole left ventricular apex was obliterated by a thick mural thrombus caused by disease-related damage to the endocardium (Fig. 1). Although a biventricular assist device could be implanted with a left ventricular thrombectomy, the formation of another thrombus with its accompanying risks could not be ruled out. Recurrence of a left ventricular thrombus may cause LVAD thrombosis or embolic events, which can result in major complications with a high mortality rate. Therefore, in our case, we chose TAH implantation as a bridge to transplant to eliminate any potential major secondary complications caused by the disease. Good short-term outcome (up to 18 months) has been achieved after cardiac transplantation in a patient with hypereosinophilic syndrome with no evidence of early myocardial involvement on echocardiography and myocardial biopsy [9].

In conclusion, we have shown that eosinophilic myocarditis was associated with acute decompensated biventricular dysfunction in our patient. Moreover, we report here the successful treatment of fibrotic end-stage eosinophilic

myocarditis with the presence of a large mural left ventricular apical thrombus with TAH implantation as a bridge to transplant.

Compliance with ethical standards

Conflict of interest The authors have no conflicts of interest to disclose.

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