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Challenges of Managing Giant Cell Myocarditis: A Case Report on the Mechanical Support Perspective

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INTRODUCTION

Giant cell myocarditis is a rare and fatal disease which may result in heart failure, complete heart block, or ventricular arrhythmias. We describe a patient who previously had been discharged from our institution with a left ventricular assist device and immunosuppressive therapy for management of his giant cell myocarditis. His subsequent course was complicated by further deterioration of heart function which required multiple mechanical circulatory support devices. He successfully received a heart transplant which later had recurrence of giant cell myocarditis. This case highlights the challenges of left and right sided mechanical assist devices in managing giant cell myocarditis.

CASE PRESENTATION

AG is a 33-year-old Hispanic man with a history of heart failure with reduced ejection fraction secondary to giant cell myocarditis implanted with a HeartMate3 left ventricular assist device (LVAD) who presented after experiencing low flow alarms.

The patient presented with a three-week history of diffuse abdominal pain and low-flow alarms on his LVAD. Device parameters were as follows: flow of 3.3 L/min, speed of 5500 rpm, PI of 4.2, and Power of 3.9 Watts. Echocardiography was significant for right ventricular dilation and hypertrophy and severely decreased right ventricular function. CT abdomen/pelvis showed hepatic venous congestion and a thick-walled descending colon. There were new findings of fluid overload with new pleural effusions, ascites, and anasarca. The patient was admitted due to worsening right heart failure and increasing inotrope requirements. He also tested positive for SARS COV-2 on admission PCR testing despite being asymptomatic and afebrile. Right heart catheterization showed RA pressure of 25mmHg, RV 30/25 mmHg, PA 30/25/28, PCWP 25mmHg with a CO of 2.7, CI of 1.5, SVR of 2516, and PVR of 1.1. The patient was started on milrinone for inotropic support. He also began workup for right

ventricular assist device as a bridge to heart transplantation and received a CT angiogram which showed scarring of R IJ from a prior Protek Duo temporarily placed on a previous admission. This resulted in unfavorable subclavian anatomy for a new Protek Duo.

Due to concern for worsening RV dysfunction, RHC was repeated five days later which showed RA 27mmHg, PA 34/23, PCWP 23 with CI of 1.57, CO of 2.87, PVR of 1.04, and SVR of 2033. Milrinone dose was increased and dobutamine was added. The patient continued to clinically deteriorate so an Impella RP was placed One day after placement of the device, AG had darkening of his urine, elevated LDH, and decreased hemoglobin and platelet count, consistent with device-induced hemolysis. A subsequent right IJ angioplasty was performed to allow for placement of a Protek Duo. The patient's undocumented immigration status had in the past resulted in limited opportunities for transplantation, however acquisition of health insurance since then allowed for the patient to be a candidate for an orthotopic heart transplant.

DISCUSSION & KEY POINTS

Giant cell myocarditis (GCM) is a rare and fatal disease resulting in significant mortality due to heart failure, complete heart block, and ventricular arrhythmias. Patients with GCM often present at middle-age in fulminant heart failure or arrhythmias, and early diagnosis is critical for management. Even for patients diagnosed with GCM during their lifetimes and managed with immunosuppressive therapy, transplant-free survival at 5-years is only 48%¹. While the pathophysiology of GCM is not entirely understood, it is characterized by T-lymphocyte myocardial inflammation that can be diagnosed on gold-standard endomyocardial biopsy². Diffuse myocardial inflammatory infiltrates with multinucleated giant cells can also be targeted for biopsy with cardiac MRI assistance. Imaging findings are consistent with myocardial fibrosis; there is myocardial strain and late-gadolinium enhancement visible at affected areas of the myocardium³.

Immunosuppression of T-mediated inflammation with prednisone, azathioprine, and cyclosporine can mitigate the disease and improve survival. Patients should also be optimized with goal directed medical therapy for heart failure, but unstable hemodynamic presentation can limit the extent of medical management. Despite immunosuppression and medical management, mortality is high and serious ventricular tachycardias continue to be the single most prominent reason for death in patients with GCM⁴. Immunosuppression and ICD placement are mainstays of GCM therapy, but this report focuses on mechanical circulatory support (MCS) devices and their role in GCM management.

There are a variety of options for MCS when managing GCM. These options can be temporary or durable, but destination therapy is limited to orthotopic heart transplant. The decision for types of MCS can be influenced by the patient's clinical status, degree of cardiac dysfunction, institutional preferences, and patient's listing status for transplant. Because of the rare and fulminant presentation of GCM, it is common for patients to be severely symptomatic prior to biopsy-proven diagnosis. In one case series of seven GCM patients the time interval from referral to device placement or transplant ranged from 2 days to 4 months, and GCM was not diagnosed in any patient prior to intervention⁵. There is favorable evidence for VA-ECMO as the first temporary MCS option⁶. In addition to ECMO, intra-aortic balloon pumps (IABP) or other trademarked devices are feasible as temporary bridge-therapy. More durable MCS options include LVAD, BiVAD, or total artificial heart (TAH) implants.

In the case of AG, the patient had a previous admission with right ventricular dysfunction requiring a Protek Duo RVAD. The Protek Duo Cannula has been shown to be a safe and effective option for short term Bi-Ventricular support in conjunction with a secondary LV device⁷. This is a dual lumen cannula device percutaneously inserted via the internal jugular vein and positioned to have the inflow lumen in the right atrium and the outflow lumen positioned in the main pulmonary artery. A disadvantage of short-term percutaneous RV assist devices is the possibility of venous scarring resulting in difficulty of device re-insertion. For AG, venous angioplasty was a safe and effective technique to allow for Protek Duo re-insertion in a patient with unfavorable anatomy who has failed Impella RP placement.

After hemodynamic stabilization, AG was able to successfully receive an OHT. His clinical course was then complicated by recurrence of GCM. Studies have shown that approximately 12% of patients can have

GCM recurrence after transplant despite continuing immunosuppressive therapy⁸. The rarity, severity, and acuity of giant cell myocarditis results in limited ability to conduct randomized control trials. Despite these barriers to establishing better evidence-based management, the use of MCS, and immunosuppressive therapy are still cornerstones of therapy as bridge-therapy to cardiac transplantation.

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