Rapidly Progressive Giant Cell Myocarditis Requiring Cardiac Transplant

Madison Rosen, MD1, Andrea Healy, DO1, Elena Casanova, CRNP2, Benjamin Horn, DO2, Mara Caroline, MD2
1Department of Internal Medicine, Lankenau Medical Center, Wynnewood, PA
2Department of Cardiology, Lankenau Medical Center, Wynnewood, PA

BACKGROUND

Giant cell myocarditis (GCM) is a rare and rapidly progressive myocarditis that frequently presents with cardiogenic shock. Its estimated prevalence is 0.13 per 100,000 patients however, this is likely an underestimate due to its high mortality rate. We describe a case of GCM that presented with complete heart block (CHB) after NSTEMI and later, cardiogenic shock requiring mechanical support and ultimately heart transplant.

CASE

A 52-year-old female with no significant past medical history presented to our hospital with persistent substernal chest pain and dyspnea. Of note, one week prior to presentation, she had an NSTEMI and was found to have 80% RCA stenosis for a DES was placed. Her post-operative course was complicated by persistent complete heart block requiring pacemaker placement. On admission to our institution, BP was 94/47, HR 65 bpm, satting 100% on RA. ECG demonstrated A sensed V paced rhythm. Labs were notable for troponin elevation to 14 ng/mL (previously 7), BNP 2261, transaminitis. She had a bedside echo performed demonstrating new biventricular dysfunction and EF 10%.

DECISION MAKING

Right heart catheterization and coronary angiography were performed which demonstrated elevated right and left heart filling pressures (RA 24, PA 44/31 (mean 35 mm Hg) and PCW 29 mm Hg, Pa sat 44% (CO 2.0 L/min, CI 1.3 L/min); coronary angiography with patent coronaries and RCA stents. The patient was subsequently placed on IABP and dobutamine. Her clinical status stabilized however, <24 hours later she suddenly decompensated without any changes to her medications and she developed lower limb ischemia. She was transitioned to VA-ECMO + Impella. She was maintained on this platform awake and not intubated to facilitate urgent transplant evaluation. Given concern for GCM, she was immediately transferred to the nearest transplant center where endomyocardial biopsy (EMB) was performed and transplant workup was initiated. Biopsy demonstrated giant cell myocarditis. She was started on high dose IV steroids, tacrolimus, and thymoglobulin. She was unable to be weaned from VA-ECMO and received OHT 8 days later. She was medically managed with tacrolimus, mycophenolate and steroids. Patient was ultimately weaned from ventilator, and ECMO was decannulated 10 days post op. Repeat TTE prior to discharge demonstrated EF 50-55%. She was discharged in less than 45 days and has good functional status and no evidence of rejection or recurrence on repeat EMB 2 months post op.

Toolbox for treatment of GCM

- Corticosteroids
- Cyclosporine or tacrolimus
- Inotropes
- Mechanical support
- Transplant
- When stable: HF GDMT, ICD

Giant cell myocarditis (GCM) is a distinct form of myocarditis with rapidly progressive disease. It frequently leads to cardiogenic shock requiring inotropic and mechanical support.

Cardiac transplantation was used as salvage therapy in a patient with rapid onset giant cell myocarditis and cardiogenic shock refractory to medical therapy and mechanical support with VA-ECMO.

REFERENCES


FIGURE 1

1. ECG after NSTEMI with 1st degree AV block and bifascicular block prior to pacemaker placement.

FIGURES 2 & 3

2. Figures 2 & 3 (apical four chamber) and 3 (parasternal long) - TTE showing hypokinetic LV, EF 10%.

FIGURE 4

4. Pathology of EMC biopsy showing inflammatory infiltrates and multinucleated giant cells.