Acute Necrotizing Eosinophilic Myocarditis Treated with Anti-IL5 Inhibitor and Mechanical Support

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INTRODUCTION

Acute necrotizing eosinophilic myocarditis (ANEM) is a rare disease, usually seen in adolescents and young adults. It is characterized by eosinophilia, fulminant heart failure and a high mortality rate[1]. Etiologies of ANEM include drug reactions, parasitic infections, hematologic cancers, hypereosinophilic syndrome, or eosinophilic granulomatosis with polyangiitis, while still others remain idiopathic[2]. Treatment has traditionally focused on cardiac support, precipitating trigger withdrawal, and aggressive steroid immunosuppression. However, in an already immunocompromised patient, the use of a steroid-sparing agent has not been well studied.

CLINICAL VIGNETTE

A 30 year old female from Sierra Leone presented with several days of chest pain and fevers. EKG showed pronounced anterior ST elevations and a troponin I was elevated to 10.77 ng/ml. Echocardiogram revealed globally reduced left ventricular function with an ejection fraction (EF) of 25%. She was transferred to our tertiary care center with concern for myocarditis and a new diagnosis of AIDS (CD4 56 cells/uL). She experienced sustained ventricular tachycardia with hemodynamic instability on arrival and received IV methylprednisolone. Angiography revealed normal coronary arteries, biventricular failure and cardiogenic shock. She underwent emergent placement of extracorporeal membrane oxygenation (ECMO) in the subclavian veno-arterial configuration. Peripheral eosinophil count was 0 but an endomyocardial biopsy demonstrated diffuse eosinophilic invasion of the myocardium, and a diagnosis of ANEM was made. An exhaustive investigation did not identify any precipitant. Due to the patient's already immunocompromised status secondary to AIDS, subcutaneous mepolizumab, an anti-interleukin(IL)-5 monoclonal antibody, was administered as a steroid sparing agent.

Figure 1: Presenting ECG

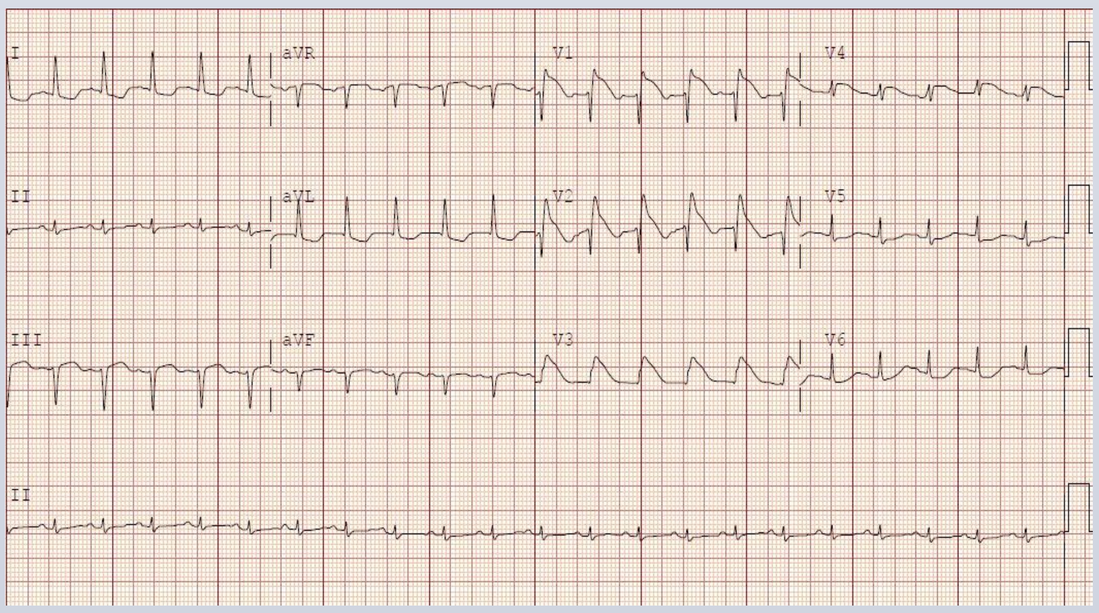
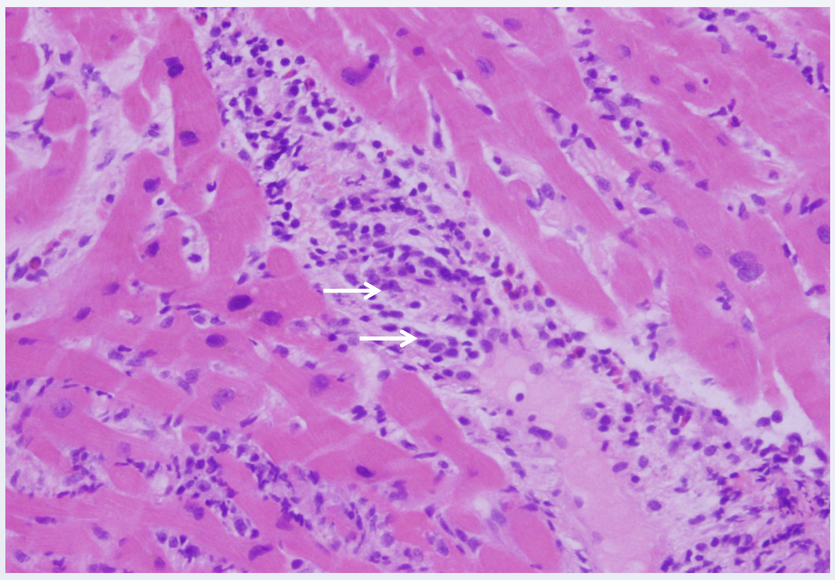


Figure 2: Endomyocardial biopsy



Necrotizing eosinophilic myocarditis: There is a mixed inflammatory infiltrate including numerous eosinophils (arrows) hematoxylin and eosin 20x original magnification)

Her course was complicated by pneumothorax, stroke without permanent neurologic deficit, and subclavian artery dissection requiring covered stent repair. ECMO support was discontinued after 6 days when her echocardiogram showed improved LV function. She was initiated on HIV treatment with a combination of abacavir, dolutegravir, and lamivudine. Prior to discharge a cardiac MRI noted an EF of 48% and an anterior LV aneurysm without evidence of delayed enhancement. Medical therapy was initiated with anticoagulation, beta-blocker and angiotensin-converting enzyme inhibitor. As an outpatient she continued a steroid taper and monthly IL-5 injections without recurrence of heart failure. She remains disease free at one year follow up.

DISCUSSION

Given the rare occurrence of ANEM there is limited guidance on medical therapy. In this vignette, the patient presented with both ANEM and severe immunocompromise. In an attempt to reduce potential life threatening complications with steroid suppression, mepolizumab was administered for direct eosinophilic suppression. Mepolizumab is a humanized monoclonal antibody of IgG. This antibody targets IL-5 and prevents its interaction with the IL-5 receptor[3]. This biologic agent was initially developed to treat allergic asthma but has also been used with success in other systemic eosinophilic diseases such as Churg–Strauss and hyper-eosinophilic syndromes[3]. The successful use of mepolizumab to treat ANEM offers promising possibilities. Its novel use as a steroid-sparing agent makes it an attractive potential therapy for this rare disease.

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Disclosures:

I will discuss off label use and/or investigational use of the following drugs: mepolizumab.
No relevant financial relationships exist related to my role in this session.
Author Seif: No relationships to disclosure
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