

## MINI-FOCUS ISSUE: IMAGING

INTERMEDIATE

## CASE REPORT: CLINICAL CASE

# Eosinophilic Myocarditis in a Patient With Sickle Cell Disease



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**ABSTRACT**

A 23-year-old man with sickle cell disease treated with splenectomy and allogenic stem cell transplantation presented with recurrent chest pain, elevated cardiac enzymes, and unremarkable electrocardiography. His work-up revealed eosinophilia, raising concern for eosinophilic myocarditis. Cardiac magnetic resonance imaging showed patchy late gadolinium enhancement of the left ventricular free wall, suggestive of myocarditis. He was treated with high-dose intravenous steroids followed by oral prednisone, with improvement in his symptoms and eosinophilia and a decrease in cardiac enhancement on follow-up imaging. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:918-21) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

**HISTORY OF PRESENTATION**

A 23-year-old man presented to the emergency department with profound nausea and vomiting. He was found to have peripheral eosinophilia (6,900 /mm<sup>3</sup>), and computed tomography of the abdomen revealed duodenal wall thickening. Upper endoscopy with duodenal biopsy demonstrated eosinophilic infiltrates. He was treated with oral prednisone and discharged. One week later he presented to the emergency department, this time with intermittent exertional chest pain for 1 day. His peripheral blood count was notable for leukocytosis

(30,400/mm<sup>3</sup>) with differential count showing 37% neutrophils, 9% lymphocytes, 3.9% monocytes, 0.3% basophils, and 48.7% eosinophils with doubling of his absolute eosinophil count (14,800/mm<sup>3</sup> from 6,900/mm<sup>3</sup>) over the course of 1 week. His C-reactive protein level was 208 mg/l, creatinine 0.88 mg/dl, aspartate transaminase 92 IU/l, and alanine transaminase 36 IU/l. His troponin levels were elevated compared to the prior week (21.96 ng/dl from <0.006 ng/dl) along with B-type natriuretic peptide level 96.9 pg/ml, but electrocardiography (**Figure 1**) did not show signs of ischemia or injury. Transthoracic echocardiography showed preserved biventricular systolic function without regional wall motion abnormalities or pericardial effusion.

**LEARNING OBJECTIVES**

- To identify patients presenting with myocarditis in the appropriate clinical context.
- To understand the emerging role of cardiac MRI in the diagnosis of myocarditis.

**PAST MEDICAL HISTORY**

Sickle cell disease, treated with splenectomy 15 years earlier and allogenic stem cell transplantation 11 years earlier.

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## DIFFERENTIAL DIAGNOSIS

The differential diagnosis included pulmonary embolism, myocardial infarction, and viral and autoimmune myocarditis.

## INVESTIGATIONS

Cardiac magnetic resonance imaging (MRI) demonstrated patchy midwall and subendocardial late gadolinium enhancement of the left ventricular free wall and midventricular inferoseptal segment, with sparing of the right ventricle and preserved biventricular systolic function (Figure 2). Prior extensive work up of eosinophilia, including helminthic or parasitic infection (stool for ova, parasites, strongyloids), lympho- and myeloproliferative disorder (flow cytometry, breakpoint cluster region–Abelson proto-oncogene, IgE levels, B<sub>12</sub> level, cysteine-rich hydrophobic domain 2 deletion, bone marrow biopsy), and macrocytosis (tryptase level), was unremarkable.

## MANAGEMENT

Based on the elevated eosinophil count, criteria for “clinically suspected myocarditis” and Lake Louise criteria for cardiac MRI, a clinical diagnosis of eosinophilic myocarditis was made. He was started on high-dose intravenous methylprednisolone

(1,000 mg once daily) followed by oral prednisone (60 mg once daily), with rapid improvement in his symptoms and eosinophil count. He was discharged home with a recommendation to avoid strenuous physical activity. His steroids were gradually tapered over the course of 3 months without any recurrent symptoms.

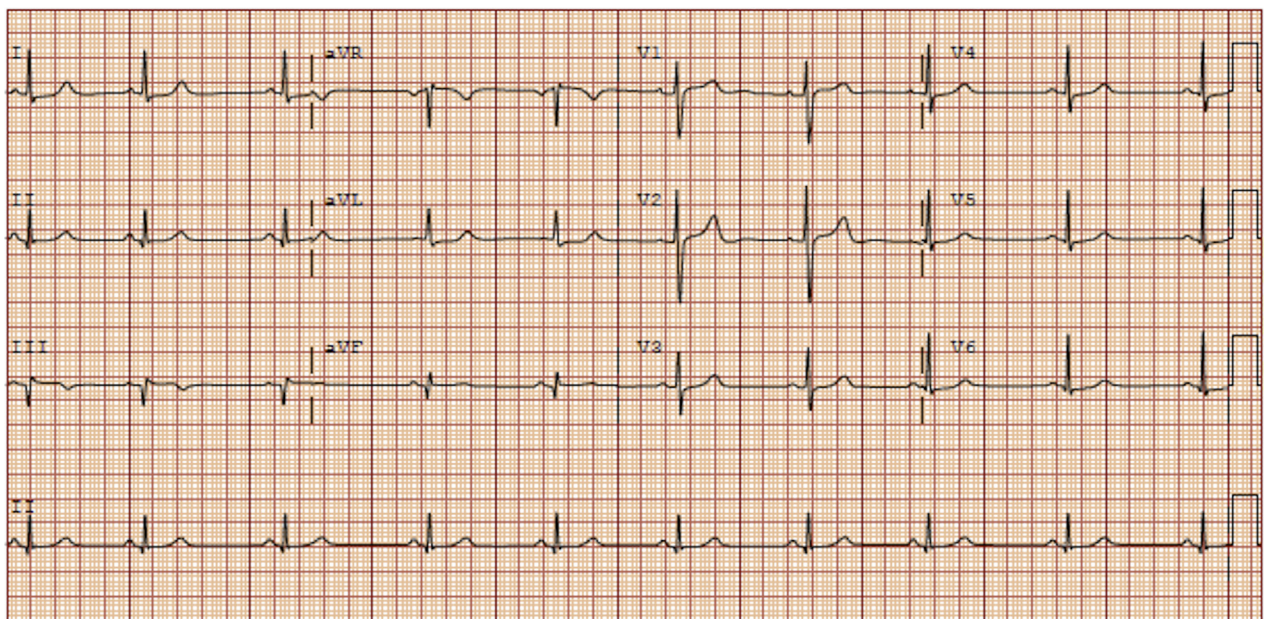
## DISCUSSION

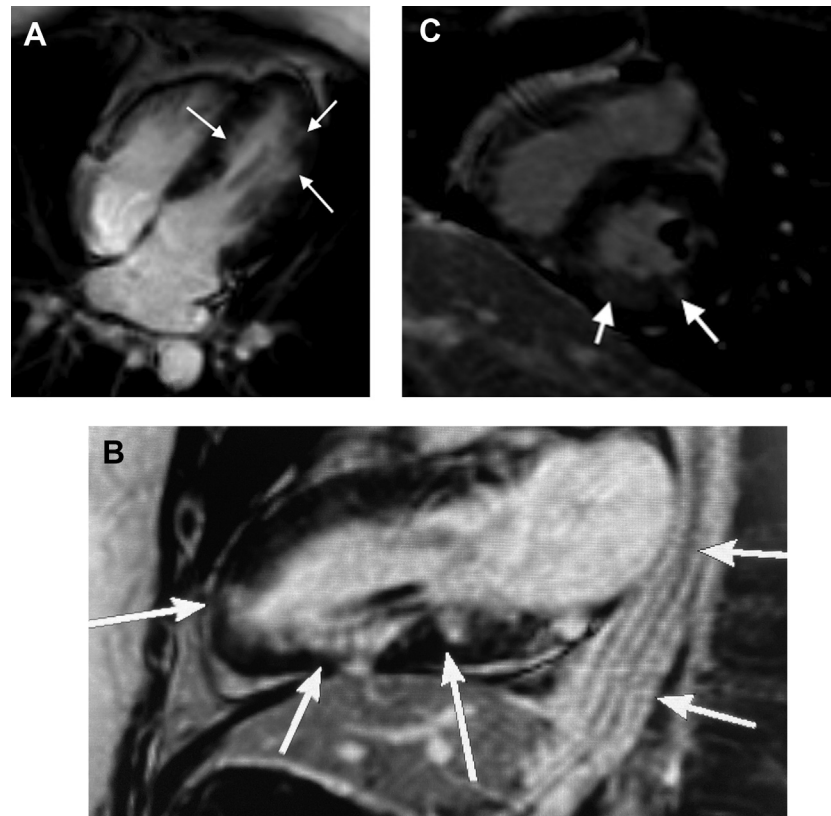
Eosinophilic myocarditis occurs because of infiltration of myocardium from peripheral eosinophilia, causing local inflammation. Multiple conditions, including hypereosinophilic syndrome, immune-complex disorders (e.g., Churg-Strauss syndrome), myeloproliferative disorders, malignancies, and parasitic infections, have been reported to cause eosinophilic myocarditis through persistent eosinophilia. In a significant number of cases, the etiology of eosinophilia remains unknown. This patient had exhaustive work-up without revealing the cause of eosinophilia. It is possible that he received clonal-disorder potentially eosinophilic granulomatosis with polyangiitis from his donor brother, who has progressively worsening asthma that required intensive care unit hospitalization. Unfortunately, we did not receive recent medical records of the donor from the family for our review to make any conclusion. Endomyocardial biopsy has been the criterion

## ABBREVIATIONS AND ACRONYMS

MRI = magnetic resonance imaging

FIGURE 1 Electrocardiogram



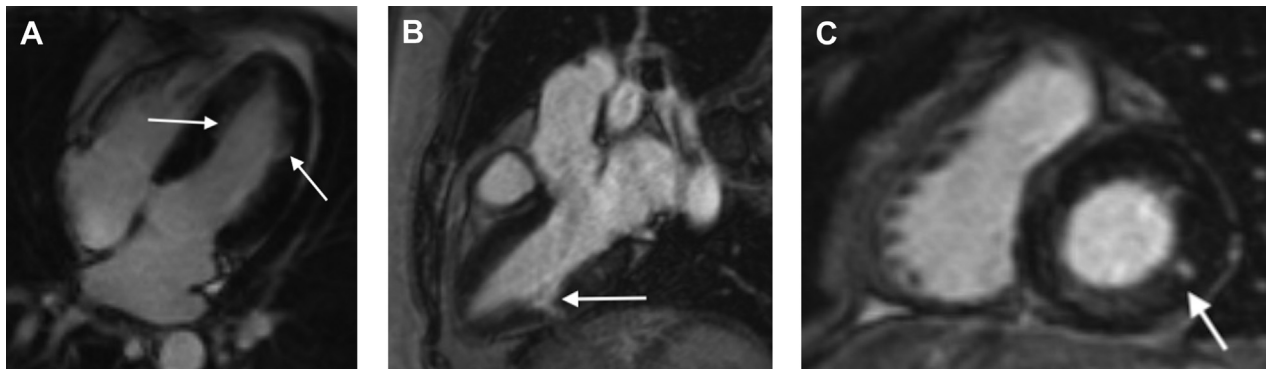
**FIGURE 2** Cardiac Magnetic Resonance Imaging

Late gadolinium enhancement of left ventricle (arrows): (A) apical 4-chamber view, (B) vertical long-axis view (arrows also indicate a thickened esophagus consistent with esophagitis, as seen in prior computed tomogram), and (C) short-axis view.

standard for definitive diagnosis of myocarditis. The yield of endomyocardial biopsy may vary owing to patchy involvement of the myocardium and the phase of myocarditis. Its yield is higher during the acute phase, but during the subacute phase sensitivity may be as low as ~54% (1). An expert consensus recommends cardiac MRI for diagnosis of myocarditis in clinically suspected patients with at least 1 clinical criterion and at least 1 diagnostic test consistent with myocarditis (2,3). Baccouche *et al.* (4) reported good correlation between endomyocardial biopsy and cardiac MRI findings in patients with acute presentation of myocarditis with elevated troponin levels. Cardiac MRI has excellent sensitivity (87.5%) and specificity (96.2%) to diagnose acute myocardial inflammation (5). A recent registry of acute myocarditis patients suggests an increasing role of cardiac MRI based on initial clinical presentation, stable vital signs, and preserved ejection fraction. In addition, there were

no adverse long-term outcomes in patients who did not undergo endomyocardial biopsy (6). Cardiac MRI provides additional prognostic information in acute myocarditis. Late gadolinium enhancement during acute myocarditis is associated with worse clinical outcomes, that is, increased heart failure hospitalizations, resuscitated cardiac arrest, and sudden cardiac death (7). Cardiac MRI can provide additional information about the primary pathologic process, associated with hypereosinophilia, leading to myocardial damage, for example, hypereosinophilic syndrome is commonly associated with diffuse sub-endocardial late gadolinium enhancement, biventricular thrombi, and hypokinesis, whereas eosinophilic granulomatosis with polyangiitis spares the right ventricle with predominant midwall involvement of the left ventricle. In appropriate clinical settings, cardiac MRI can be an excellent surrogate for endomyocardial biopsy in the diagnosis

**FIGURE 3** Interval Cardiac Magnetic Resonance Imaging



Involvement of late gadolinium enhancement (arrows): (A) apical 4-chamber view, (B) vertical long-axis view, and (C) short-axis view.

of eosinophilic myocarditis during the acute presentation.

#### FOLLOW-UP

On 3-month follow up, the patient's cardiac MRI showed significant decrease of intramural and sub-endocardial enhancement (Figure 3) without evidence of LV thrombus or restrictive physiology. He was gradually tapered off steroids without any recurrent symptoms.

#### CONCLUSIONS

Eosinophilic myocarditis is a rare entity that can have dire consequences if not diagnosed quickly.

Cardiac MRI can be a surrogate for endomyocardial biopsy in the diagnosis of myocarditis, and can lead to rapid diagnosis and prompt management.

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**KEY WORDS** cardiac magnetic resonance, left ventricle, necrosis