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Eosinophilic myocarditis in platelet-derived growth factor receptor α -rearranged myeloid neoplasm with central nervous system involvement: a case report

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Abstract

Eosinophilic myocarditis is a rare inflammatory condition caused by eosinophilic infiltration of the myocardium, often associated with systemic or hematologic disorders.

We report a 31-year-old man with a newly diagnosed platelet-derived growth factor receptor α -rearranged myeloid neoplasm who presented with acute chest pain and elevated cardiac biomarkers. Echocardiography showed Loeffler-type apical involvement, coronary computed tomography angiography excluded obstructive coronary disease, and cardiac magnetic resonance (CMR) demonstrated acute non-ischemic inflammatory myocardial injury. During hospitalization, neurological symptoms revealed central nervous system involvement by the underlying myeloid neoplasm. Corticosteroids and targeted hematologic treatment resulted in marked clinical and radiologic improvement.

On long-term follow-up, CMR showed normal cardiac function with complete resolution of the inflammatory myocardial abnormalities and no residual fibrosis.

This case highlights the value of early multimodality cardiac imaging, etiologic diagnosis, and coordinated cardio-hematologic management in recognizing potentially reversible eosinophilic myocardial injury and limiting progression to irreversible cardiac sequelae.

Key words: eosinophilic myocarditis, acute myocarditis, cardiac magnetic resonance, multimodality imaging, myocardial injury, PDGFRA-rearranged myeloid neoplasm.

Introduction

Myocarditis is an inflammatory disease of the myocardium with heterogeneous etiologies and clinical presentations. According to the 2025 European Society of Cardiology (ESC) Guidelines, myocardial and pericardial inflammation frequently coexist within the clinical spectrum of inflammatory myocardial–pericardial syndrome (IMPS) [1].

Eosinophilic myocarditis (EM) is a rare histopathologic subtype, characterized by eosinophilic infiltration of the myocardium, usually associated with peripheral eosinophilia [2]. Underlying causes include hypersensitivity reactions, autoimmune and vasculitic disorders, infections, and, less commonly, hematologic malignancies. Among the latter, myeloid/lymphoid neoplasms with eosinophilia and tyrosine kinase gene fusions represent a rare yet clinically relevant subgroup [3-5]. Platelet-derived growth factor receptor alpha (PDGFRA) is a receptor tyrosine kinase; PDGFRA rearrangements, most commonly FIP1L1-PDGFRA, result in constitutive kinase activation, driving clonal eosinophilic proliferation and conferring marked sensitivity to imatinib [3-6].

We describe early eosinophilic cardiac involvement in a patient with a PDGFRA-rearranged myeloid neoplasm who subsequently developed CNS involvement. The educational value of the case lies in the rapid application of the contemporary ESC diagnostic pathway, integrating multimodality cardiac imaging with etiologic and molecular characterization to guide timely treatment and longitudinal cardiac surveillance [1].

Case Report

A 31-year-old man presented to the emergency department with acute-onset chest pain at rest, radiating to the left shoulder and associated with low-grade fever and abdominal discomfort. Several weeks earlier, he had developed vomiting, diarrhea, severe asthenia, recurrent papular skin lesions, persistent eosinophilia, and marked splenomegaly, prompting hematologic evaluation. Bone marrow biopsy showed approximately 90% cellularity with marked eosinophilic and mast-cell expansion, about 1% CD34-positive blasts, and mild reticulin fibrosis (MF-1), consistent with a chronic-phase myeloid neoplasm with eosinophilia. BCR-ABL1 and JAK2 testing were negative. Molecular analysis subsequently identified the FIP1L1-PDGFRA fusion. At that stage, echocardiography showed no left ventricular hypertrophy and preserved left ventricular ejection fraction (65%). No cardiovascular risk factors were reported, and disease-directed therapy had not yet been started.

On admission, physical examination showed sinus tachycardia, splenomegaly (approximately 8 cm below the left costal margin), and a truncal rash, without murmurs, pulmonary congestion, or peripheral edema. Blood pressure was 144/95 mmHg, heart rate 115 bpm, oxygen saturation 97% on room air, and body temperature was normal.

Electrocardiography (ECG) showed sinus tachycardia, a short PR interval, ST-segment depression in the anterolateral leads, and T-wave inversion in the inferior leads.

Laboratory testing revealed leukocytosis (24,800/ μ L) with marked eosinophilia (absolute eosinophil count 12.7×10^9 /L), mild anemia (12.7 g/dL), thrombocytopenia (77,000/ μ L), elevated high-sensitivity troponin I (2557–2608 pg/mL), and NT-proBNP (2009 pg/mL), with preserved renal function and mild transaminase elevation.

Transthoracic echocardiography demonstrated preserved left ventricular systolic function with increased wall thickness and echogenicity of the apical segments, resulting in systolic apical obliteration and a pseudo-hypertrophic appearance suggestive of Loeffler-type involvement. A mild pericardial effusion (5 mm) was also observed (Figure 1).

Given the presenting symptoms, ECG abnormalities, and laboratory findings, the differential diagnosis included non-ST-segment elevation myocardial infarction (NSTEMI) and myocarditis. The patient was admitted to the coronary care unit.

Considering his low cardiovascular risk profile, coronary computed tomography angiography (CCTA) was performed to exclude an ischemic cause. ECG-synchronized acquisition with prospective tube-current modulation was obtained at a mean heart rate of 82 bpm after administration of 80 mL of iodinated contrast medium (350 mg I/mL) at 7 mL/s. CCTA demonstrated a left-dominant coronary circulation, a trifurcating left main coronary artery, and a small-caliber right coronary artery, with no obstructive coronary stenoses.

CMR showed preserved biventricular volumes and global systolic function, with only minimal apical hypokinesia. Multiple areas of non-ischemic late gadolinium enhancement involved the mid-to-apical lateral wall and cardiac apex, with subtle associated edematous changes, consistent with acute/subacute inflammatory myocardial injury. A pericardial effusion was also present, without pericardial thickening (Figures 2 and 3). These findings, in the clinical context of marked eosinophilia and PDGFRA-rearranged disease, supported the diagnosis of acute eosinophilic myocardial involvement. During hospitalization, a brief episode of non-sustained ventricular tachycardia was recorded and treated with beta-blocker therapy.

In the absence of clinical or microbiological evidence of an infectious cause, high-dose intravenous corticosteroids were administered.

Low-molecular-weight heparin was introduced during the acute phase because of the apical endocardial involvement and associated thrombotic risk. Imatinib was initiated at 100 mg/day as PDGFRA-directed therapy. Because systemic clinical improvement remained incomplete in the setting of rapidly progressive multisystem disease, the dose was subsequently increased to 400 mg/day.

The patient later developed lower limb hypoesthesia and weakness, followed by visual symptoms. Brain magnetic resonance imaging revealed multiple leptomeningeal and parenchymal lesions. Cerebrospinal fluid immunophenotyping demonstrated immature myeloid cells, suggesting neurological dissemination of the underlying myeloid neoplasm. Given the small size and high-risk location of the cerebral lesions, stereotactic biopsy was not pursued. Multidisciplinary review considered the overall neurological picture consistent with CNS involvement by the myeloid neoplasm, including possible myeloid sarcoma and leptomeningeal disease. Imatinib was discontinued, and treatment was intensified with high-dose cytarabine for five consecutive days, followed by dasatinib 100 mg/day. The patient subsequently underwent allogeneic hematopoietic stem-cell transplantation.

Follow-up brain imaging showed disappearance of the previously enhancing parenchymal lesions and cerebrospinal fluid clearance. Bone marrow aspirate showed normal trilineage maturation, eosinophils accounting for 7% of cells, and no undifferentiated elements, consistent with hematological response.

At early cardiology follow-up, the patient was asymptomatic and hemodynamically stable. Left ventricular systolic function remained preserved, although apical pseudo-hypertrophic changes persisted, together with a circumferential pericardial effusion of up to approximately 21 mm without chamber compression or hemodynamic compromise. Continued echocardiographic surveillance was therefore planned.

Following allogeneic transplantation, longer-term CMR demonstrated complete resolution of the previously observed myocardial inflammatory abnormalities, and no evident residual myocardial fibrosis. At approximately one-year cardiology follow-up, the patient remained asymptomatic and hemodynamically stable; echocardiography showed preserved left ventricular ejection fraction (65%), normal right ventricular function, and only minimal circumferential pericardial separation. Beta-blocker therapy was discontinued.

Discussion

Eosinophilic myocarditis is a rare but potentially life-threatening condition caused by eosinophil-mediated myocardial injury [2]. Classically, cardiac involvement may progress through necrotic, thrombotic, and fibrotic stages, ultimately leading to restrictive cardiomyopathy if the underlying process remains uncontrolled [4]. Clinical manifestations are heterogeneous and include chest pain, heart failure, arrhythmias, and sudden cardiac death [7]. Hematologic malignancies represent an uncommon but important cause of EM.

The principal educational value of this case lies not in the individual rarity of its hematologic, cardiac, or neurological manifestations, but in their rapid integration within a contemporary diagnostic and therapeutic pathway. In accordance with the 2025 ESC approach to inflammatory myocardial-pericardial syndromes, initial assessment combined ECG and cardiac biomarkers with echocardiography, exclusion of obstructive coronary disease by CCTA, and myocardial tissue characterization by CMR [1]. In a young patient presenting with chest pain and marked troponin elevation, this strategy rapidly excluded an ischemic mechanism and demonstrated acute non-ischemic myocardial inflammation. The coexistence of marked peripheral eosinophilia, a PDGFRA-rearranged myeloid neoplasm, and Loeffler-type apical changes provided the etiologic context for a clinical diagnosis of eosinophilic myocardial involvement without delaying disease-directed treatment.

Management of EM is primarily etiology-driven. Current ESC Guidelines recommend intravenous corticosteroids as first-line therapy, with appropriate caution when an infectious cause is possible, together with specific treatment of the associated condition [1,8,9]. PDGFRA-rearranged neoplasms are particularly sensitive to imatinib. Legrand et al. identified imatinib as the treatment of choice for FIP1L1-PDGFRA-associated eosinophilic neoplasms and showed that an initial dose of 100 mg/day followed by lower-dose maintenance may provide durable complete hematologic and molecular responses [6]. Accordingly, our patient initially received imatinib 100 mg/day together with corticosteroid therapy.

Because clinical improvement remained incomplete in the setting of rapidly progressive multisystem disease, imatinib was increased to 400 mg/day. This represented treatment intensification in response to the aggressive clinical course rather than the standard initial dosing strategy for PDGFRA-rearranged disease. With subsequent CNS involvement, however, treatment was further intensified with high-dose cytarabine, and imatinib was replaced by dasatinib because of its activity against PDGFRA and more favorable CNS penetration[10]. The patient subsequently underwent allogeneic hematopoietic stem-cell transplantation.

Despite treatment, relapse and long-term cardiac sequelae remain potential concerns in EM, thus requiring serial cardiovascular imaging. Early follow-up showed preserved systolic function despite persistent apical pseudo-hypertrophic changes and pericardial effusion. At longer-term follow-up, CMR demonstrated complete resolution of the inflammatory myocardial abnormalities without evident residual fibrosis, while echocardiography at approximately one year confirmed preserved biventricular function and only minimal residual pericardial separation.

Although robust clinical evidence establishing a causal relationship between early treatment and prevention of fibrosis is lacking, this favorable trajectory is consistent with the rationale for recognizing and treating eosinophilic myocardial injury before progression to irreversible fibrotic restrictive disease. Neurological imaging and bone marrow assessment also documented treatment response.

Overall, this case illustrates how a guideline-based multimodality imaging pathway can rapidly connect an acute cardiac presentation to an underlying systemic disorder. CCTA enabled timely exclusion of an ischemic mechanism, CMR characterized myocardial inflammation and subsequently documented its resolution, and molecular diagnosis provided an actionable therapeutic target. The resulting cardio-hematologic approach allowed cardiac inflammation and the underlying clonal disorder to be addressed in parallel, with a favorable long-term cardiac outcome.

Conclusions

This case highlights the value of rapid guideline-based multimodality imaging in patients with systemic eosinophilic disorders presenting with acute myocardial injury. Early etiologic recognition and coordinated cardio-hematologic management may facilitate timely targeted treatment and limit progression toward irreversible cardiac sequelae.

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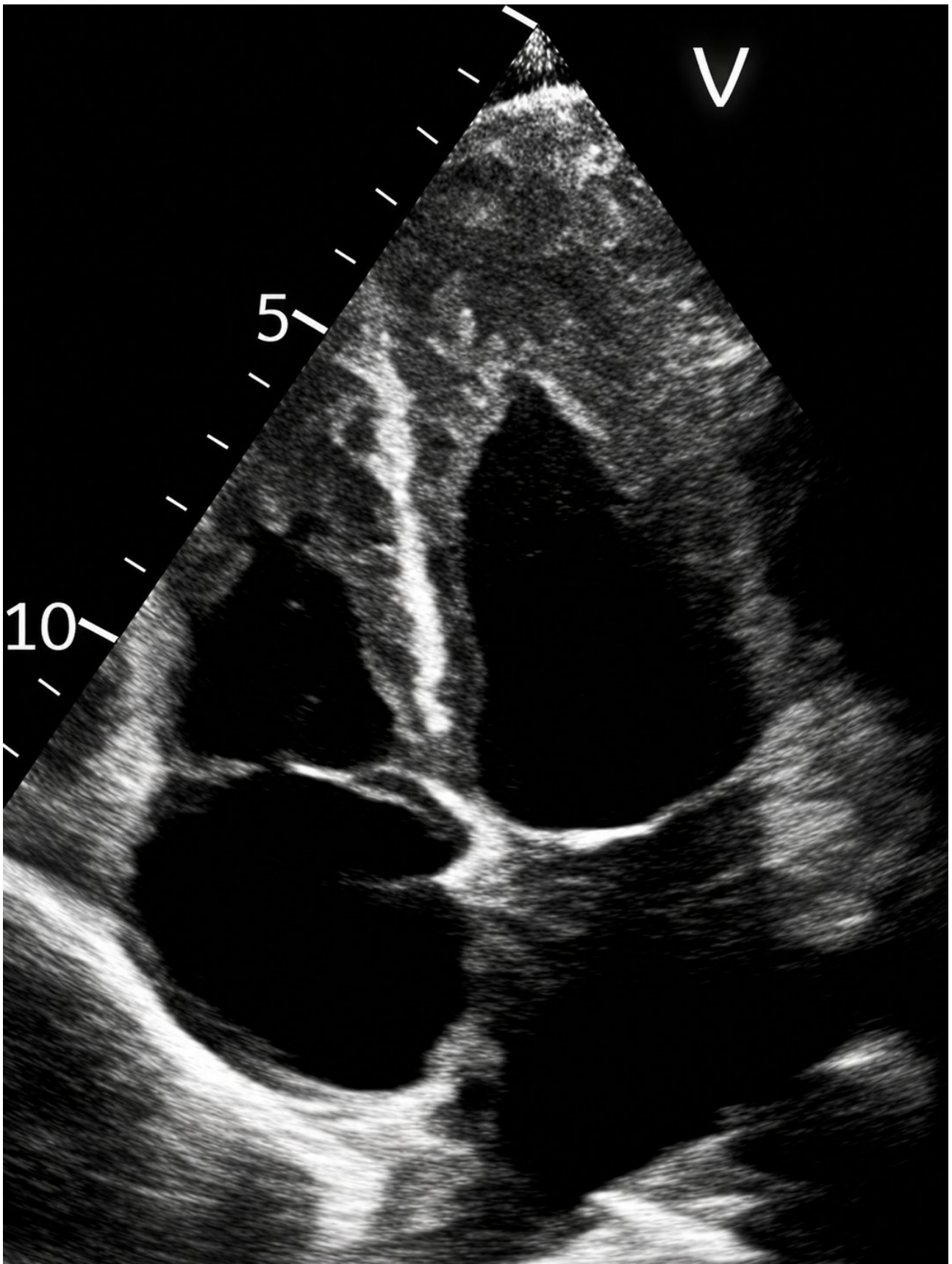


Figure 1. Transthoracic echocardiography, apical four-chamber view: increased wall thickness of the apical left ventricular segments, resulting in systolic obliteration of the apex. Findings suggestive of Loeffler endocarditis (pseudo-apical hypertrophy).

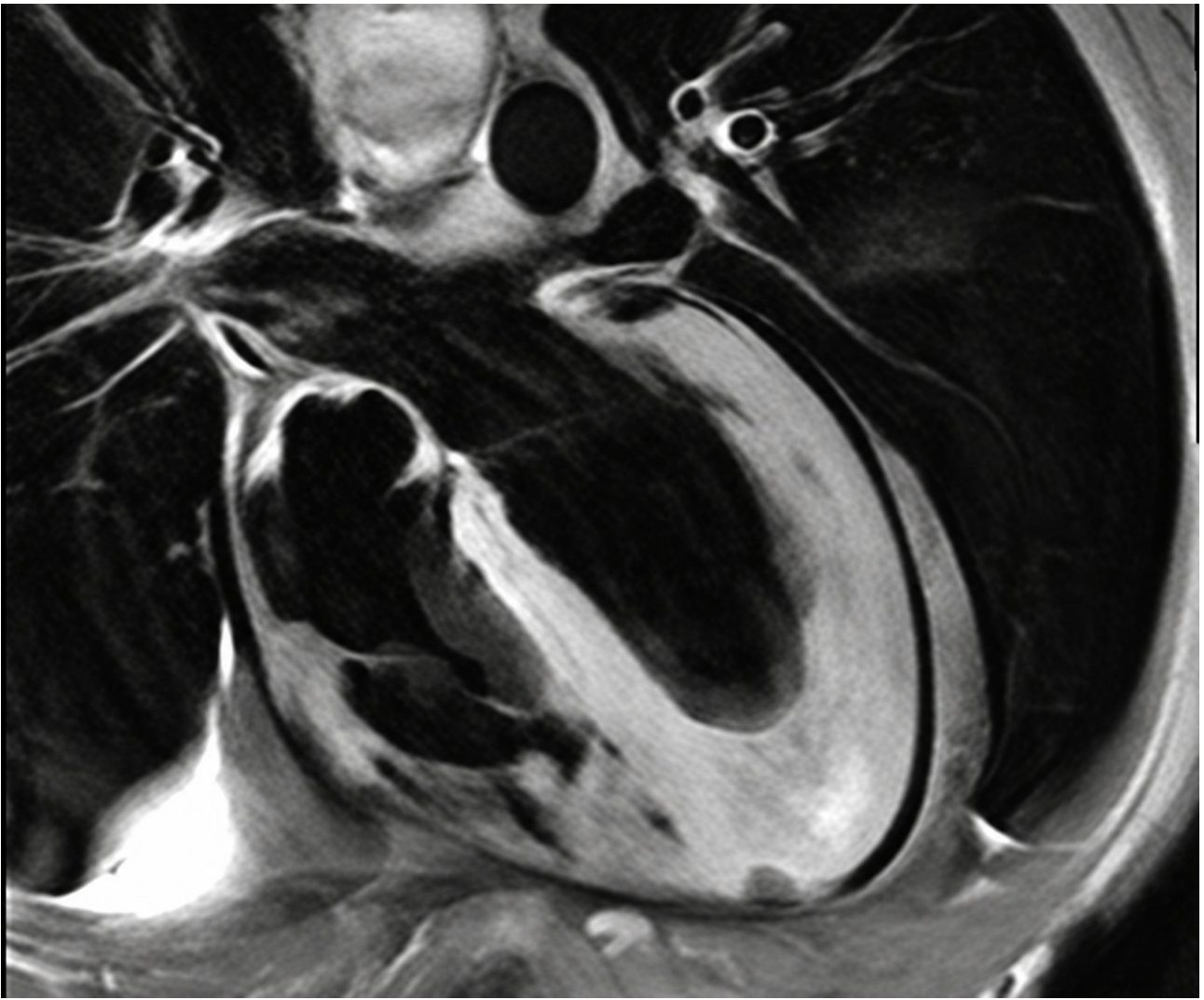


Figure 2. CMR T2-weighted short tau inversion recovery (STIR) sequence, four-chamber long-axis view: diffuse high signal intensity within the left ventricular apical and lateral walls, consistent with myocardial edema.

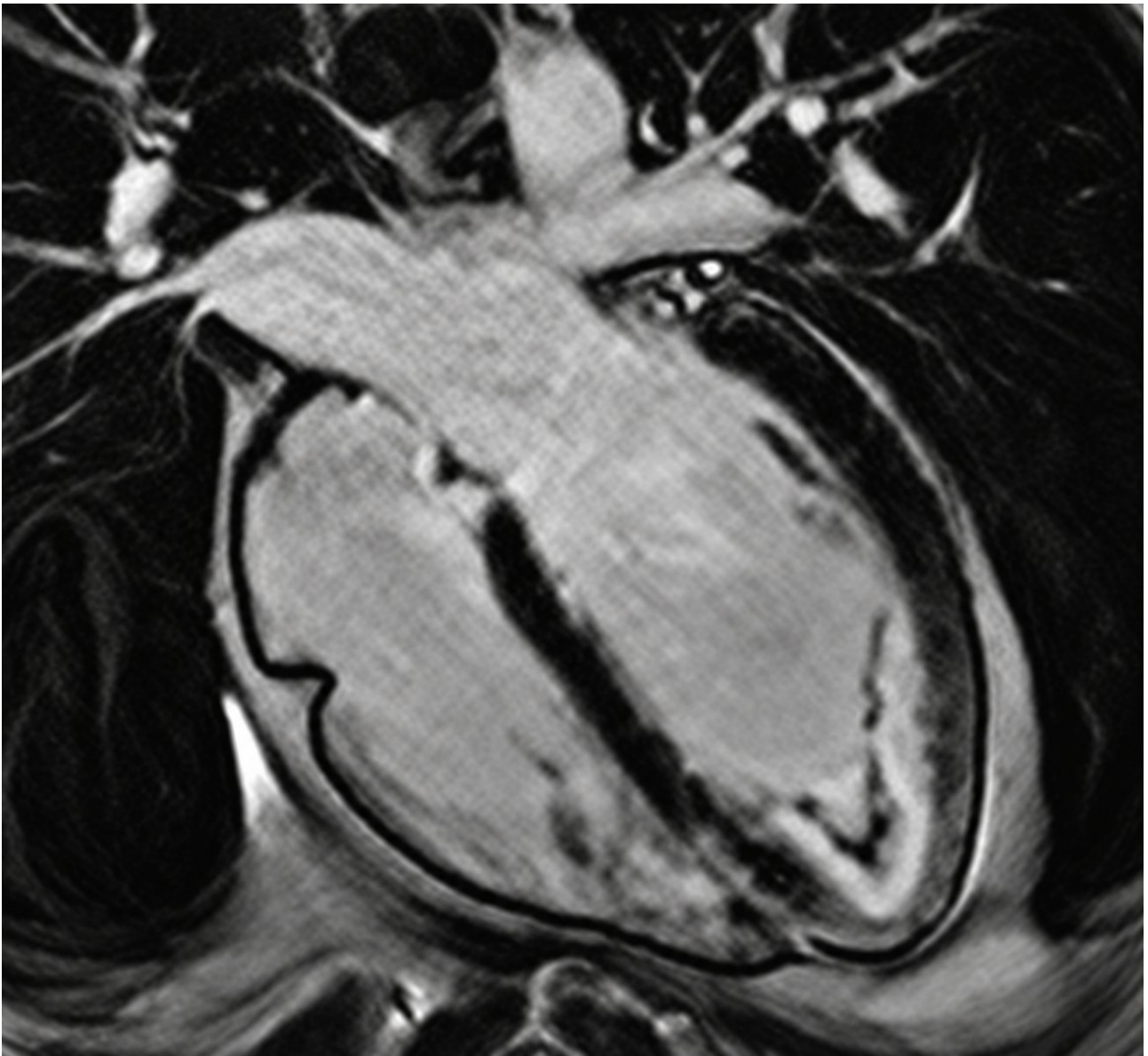


Figure 3. CMR LGE sequence: non-ischemic late gadolinium enhancement involving the left ventricular lateral wall and apex.