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Acute Severe Non-Ischemic Cardiomyopathy in Systemic Sclerosis with AntiPolymerase III Antibodies: A Case of Suspected Autoimmune Myocarditis

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Abstract

In patients with systemic sclerosis, cardiac involvement often goes underrecognized, even though it is a clinically significant finding that can lead to morbidity and mortality in affected patients. This case describes a 52-year-old woman who was diagnosed with systemic sclerosis and anti-RNA polymerase III antibody and recently developed a cardiac manifestation of acute severe systolic heart failure. The patient had elevated troponins in the absence of obstructive coronary artery disease. Initially, the patient had an ejection fraction of 25% with global hypokinesis, but coronary angiography displayed that the coronary arteries were normal, pointing towards non-ischemic cardiomyopathy. Additionally, the patient endorsed proximal muscle weakness and myalgias, creating a clinical picture of an overlap with inflammatory myositis. The patient was treated with inotropic support and high-dose corticosteroids, which led to significant clinical improvement. This case study displays the significance of understanding the role of autoimmune myocarditis as a potential cause of acute cardiomyopathy in systemic sclerosis and being able to differentiate this clinical presentation from acute coronary syndrome.

Keywords: Systemic sclerosis, Myocarditis, Cardiomyopathy, Troponin, Hypokinesis, Anti-RNA polymerase III

Introduction

Systemic sclerosis (SSc) is an autoimmune connective tissue disorder characterized by the accumulation of extracellular matrix components, such as collagen, leading to fibrosis of the skin and visceral organs, as well as microvascular injury. While renal and pulmonary involvement have been well documented, adverse cardiac events caused by systemic sclerosis

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are not as common. Clinical evidence describes that cardiac involvement is associated with an increased risk of death [1, 2]. Although cardiac involvement may go unrecognized, common manifestations include myocardial disease, pericardial disease, conduction disease, and dysrhythmias [3].

Autoimmune myocarditis in systemic sclerosis is difficult to diagnose due to its nonspecific presentation and its close resemblance to acute coronary syndrome with elevated cardiac biomarkers. The incidence of myocarditis is difficult to determine because the gold-standard diagnostic test, endomyocardial biopsy, is infrequently used [4]. In patients with systemic scleroderma, about 4-25% have anti-RNA polymerase III antibodies. Patients who have this antibody are more strongly associated with serious manifestations of systemic scleroderma, such as scleroderma renal crisis, heart problems, gastric irregularities, and high rates of systemic sclerosis-related mortalities [5, 6]. Additionally, these internal organ manifestations of systemic sclerosis, mixed with inflammatory myopathies, may further increase the risk of myocardial involvement [7].

This case presents an acute non-ischemic cardiomyopathy in a patient with systemic sclerosis, displaying the diagnostic challenges and clinical ramifications of autoimmune cardiac involvement.

Case Presentation

A 53-year-old woman with a past medical history of systemic sclerosis, type 2 diabetes mellitus, hypothyroidism, prior pulmonary embolism on anticoagulation, and dyslipidemia presented with progressive weakness, dizziness, diarrhea, and exertional chest discomfort. The patient came to the hospital endorsing worsening fatigue over a period of several months and

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new proximal muscle weakness, which included difficulty raising her arms and performing activities of daily living.

Blood pressure was monitored multiple times a day and it was significant for tachycardia and hypotension with systolic blood pressures in the 80-90 mmgHg range. On physical examination, the patient endorsed generalized weakness but no neurological deficits. Pertinent laboratory values included elevated troponin levels (>3000 ng/L), creatine kinase, and brain natriuretic peptide (BNP). Furthermore, the patient's inflammatory markers were elevated and in their past history, an autoimmune workup revealed the patient was positive for antinuclear antibody (ANA) and anti-RNA polymerase III antibodies.

The electrocardiogram obtained for the patient showed sinus tachycardia with diffuse ST-segment abnormalities, while transthoracic echocardiography revealed a reduced left ventricular ejection fraction of 25% with global hypokinesis and moderate right ventricular dysfunction. Subsequent left heart catheterization displayed normal coronary arteries, thus ruling out obstructive coronary artery disease.

Given that coronary artery disease was effectively ruled out due to various diagnostic tests and the presence of systemic autoimmune features, the patient was diagnosed with acute non-ischemic cardiomyopathy with a high suspicion for autoimmune myocarditis. Since the patient had persistent hypotension that was consistent with a low-output state, this prompted the clinical team to begin a dobutamine infusion to assist with hemodynamic support.

Due to a rising suspicion of autoimmune myocardial involvement, the patient was started on high-dose intravenous methylprednisolone followed by oral prednisone. Guideline-directed medical therapy (GDMT) for heart failure, including sacubitril/valsartan, spironolactone, and an SGLT-2 inhibitor, was started as tolerated by the patient, but beta-blockers were not initiated due

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to the patient's persistent hypotensive values. Cardiac MRI was completed twice, but the results were deemed inconclusive due to the patient's inability to hold her breath for an ample amount of time.

During the patient's hospital course, she showed improvement in her symptoms and a reduction in cardiac biomarkers. Upon discharge, she was scheduled for cardiology and rheumatology follow-up, as well as a wearable defibrillator due to reduced ejection fraction.

Discussion

This case study evaluates the role of systemic sclerosis in this patient's presentation of severe non-ischemic cardiomyopathy, likely secondary to autoimmune myocarditis. Systemic sclerosis heart involvement accounts for a major proportion of systemic sclerosis-associated mortality, resulting in manifestations such as myocardial inflammation, fibrosis, congestive heart failure, and arrhythmias. Although early signs often go unrecognized, a lack of ideal screening and treatment guidelines plays a major role in the progression of heart problems in systemic sclerosis [3, 8]. Myocarditis in individuals with this autoimmune disorder presents with elevated troponin levels and electrocardiographic abnormalities that closely mirror the presentation of acute coronary syndrome, thus making it difficult to differentiate the two diagnoses [4].

In this patient, since angiography results revealed the absence of obstructive coronary artery disease and instead displayed global hypokinesis, it supports a non-ischemic etiology. Moreover, the patient's positive anti-RNA polymerase III antibody suggests a more severe disease phenotype with potential for extensive involvement of internal organs [6].

The additional symptoms within this patient's clinical presentation, such as increased creatine kinase and overlap with inflammatory myopathy, suggest an increased chance of cardiac myopathy [7]. This overlap plays an increased role in myocardial inflammation and dysfunction.

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Cardiac MRI plays a major role in evaluating potential myocarditis in patients with underlying autoimmune disease. It helps identify myocardial edema and fibrosis [9, 10]. In patients with autoimmune myocarditis, corticosteroids, a form of immunosuppressive therapy, have been proven to be favorable in select cases [11].

This case reveals the need to consider autoimmune etiologies in patients with acute heart failure and elevated cardiac enzymes in the absence of coronary artery disease. Recognition and treatment of autoimmune myocarditis will improve and prevent further degradation of cardiac function.

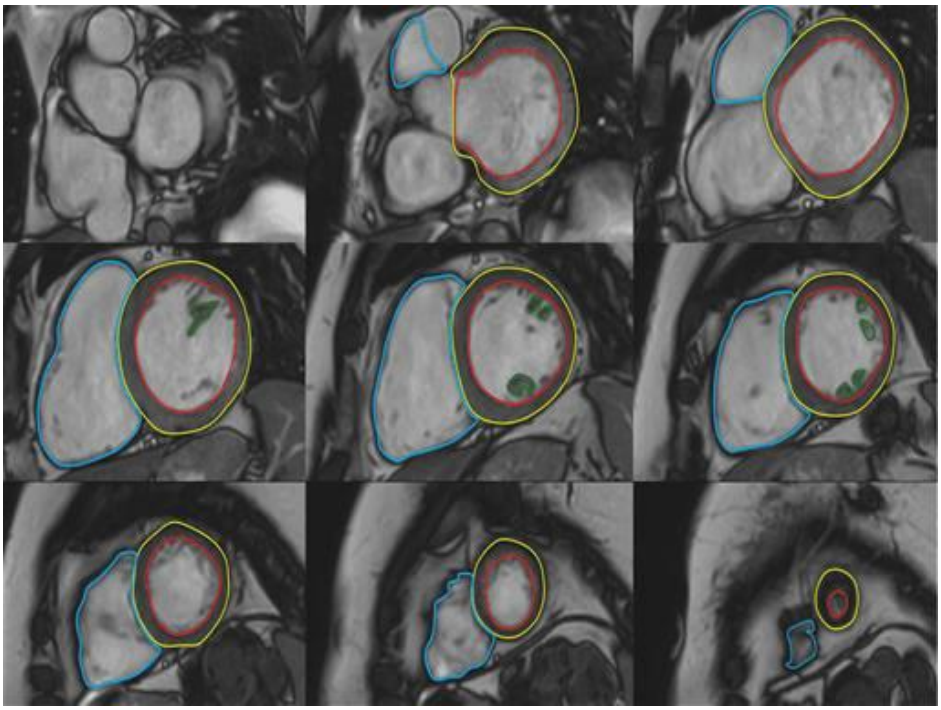

Figure 1: Normal cardiac MRI demonstrating preserved myocardial architecture without late gadolinium enhancement. Reproduced from Kawel-Boehm et al. (2020) under Creative Commons licensing[12].

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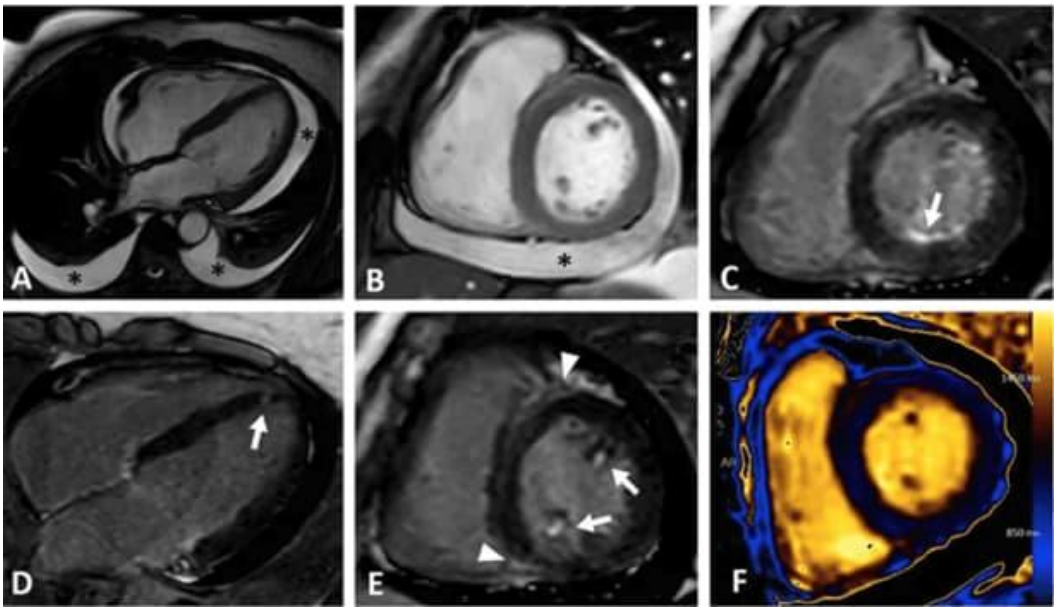

Figure 2: Cardiac MRI demonstrating myocardial fibrosis and late gadolinium enhancement in systemic sclerosis-associated myocarditis. Reproduced from Vitale et al. (2025) under CC BY 4.0 license[13].

Conclusion

Autoimmune myocarditis is an uncommon cause of acute cardiomyopathy in patients with systemic sclerosis. Early recognition and multidisciplinary management may improve clinical outcomes in autoimmune patients with heart failure and elevated troponin levels without evidence of coronary artery disease.

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