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RESEARCH ARTICLE

MYOCARDITISREVEALING AN ADULTSTILLONSETDISEASE

M. Jaafari, H. Dahmani, A. Krimech, H. Ouaouicha and E. Salengro
Cardiology Department, Lucie & Raymand Aubrac Hospital Center, Villeneuve Saint-Georges, France.

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Abstract

A 21-year-old male patient presented with a combination of fever, palpitations, a maculopapular rash, and pharyngitis. Laboratory tests revealed an elevated high-sensitivity troponin I concentration of 3000 ng/L, indicating potential cardiac involvement. A transthoracic echocardiogram demonstrated a reduction in global longitudinal strain, suggesting impaired cardiac function. Further investigation with cardiac magnetic resonance imaging confirmed the diagnosis of acute myocarditis. After a thorough clinical evaluation, the patient was diagnosed with adult-onset Still's disease. Treatment was promptly initiated, consisting of intravenous corticosteroids and Kineret (anakinra), aimed at managing the inflammatory response and preventing further complications.

Introduction:

Myocarditis is a rare and potentially life-threatening condition characterized by inflammation of the heart muscle, oftenresulting in arrhythmias, heartfailure, or suddencardiacdeath. Whilemyocarditis can becaused by infectious agents such as viruses, bacteria, or parasites, it can also arise in the context of systemicinflammatorydiseases. Adult-onsetStill'sdisease (AOSD) is a rare autoinflammatorydisorderthat can presentwith a wide range of symptoms, includingfever, rash, arthritis, and, in rare cases, myocarditis. Recognizingmyocarditis as an early manifestation of systemicdiseases like AOSD is crucial for timelydiagnosis and management. In this report, we explore the intersection of myocarditis and adult-onsetStill'sdisease, emphasizing the importance of consideringsystemicinflammatory conditions in the differentialdiagnosis of myocarditis.

Case Presentation:

A 21-year-old Caucasian man with no prior medical history presented to the emergency department with a one-week history of fever (39.5°C), fatigue, pharyngitis, odynophagia, a maculopapular rash, arthralgia, and myalgia. The patient did not report any chest pain.

His medical background was unremarkable, with no known allergies, cardiovascular risk factors, or recent travel abroad.

Upon arrival, his vital signs were stable, including a blood pressure of 120/70 mmHg and an oxygen saturation of 97% on room air. He exhibited no neurological symptoms. Physical examination revealed a fixed, pseudo-urticarial, non-pruritic, erythematous maculopapular rash localized to the extremities.

Corresponding Author:M. Jaafari
Address:Cardiology Department, Lucie & Raymand Aubrac Hospital Center, Villeneuve Saint-Georges, France.

Initial blood tests showed marked neutrophilic leukocytosis, with a white blood cell (WBC) count of 15,000 cells/uL (neutrophils 11,000/uL), along with hyperferritinemia (3,000 ng/mL) and an elevated C-reactive protein (CRP) level of 280 mg/L.

All specific cultures and serology antibody for infectious agents were negative. A Chest CT demonstrated a pneumonia opacity. At this point, we treated first with empiric antibiotic therapy by third generation cephalosporine and erythromycin, but there was no improvement.
Three days later, the patient presented a chest pain associated to a shortness of breath. Auscultation detected no murmurs. Electrocardiogramshowed sinus rhythm, heart rate of 78beats/min; diffuse ST elevation; and normal QTinterval corrected for heart rate(456ms). High-sensitivity troponin I concentration was raised (3000ng/l). The patient was transferred to intensivecare unit.

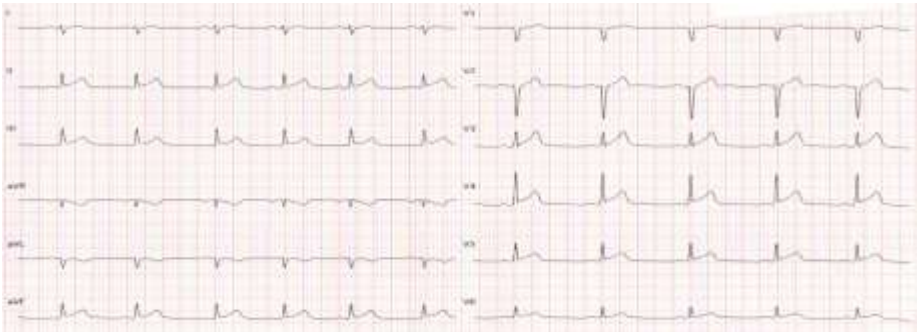

Figure 1: Electrocardiogram showing diffuse ST elevation.

A critical care echocardiography revealed hypokinesia of the lateral wall of the left ventricle without ventricular dilation or pericardial effusion, along with reduced global longitudinal strain.

Cardiac MRI was used to help make the diagnosis of myocarditis. It confirmed subacute myocarditis with involvement of the lateral segments of the left ventricular myocardium. Left ventricular ejection fraction (LVEF) was 51%. The left ventricle was not dilated, and the right ventricle shows good systolic function. A limited pericardial reaction is present in the inferolateral basal region. No myocardial iron overload is detected.

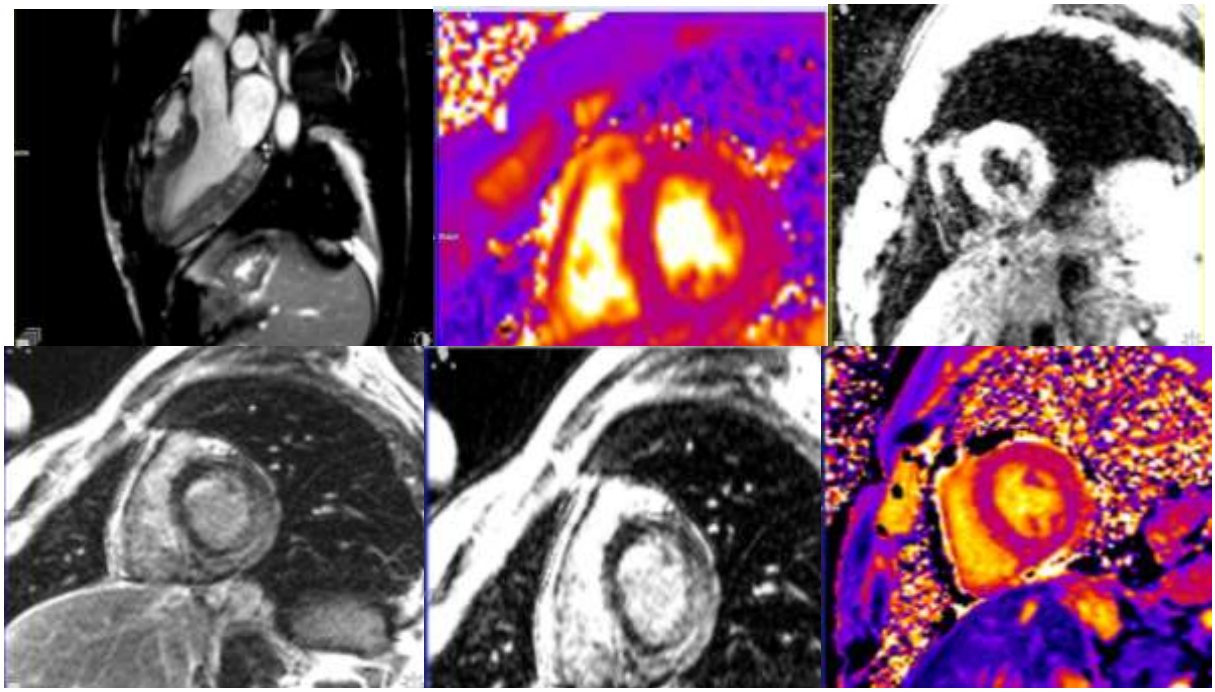

Figure 2: MRI images showing extensive acute myocarditis.

Our patient clinical presentation was characteristic of AOSDin line with the Yamaguchicriteria (fever, arthralgia, rash, leukocytosis). Kineret was started at the standard dose (anti-IL1 drug—100 mg/d).

Discussion:

First described in 1971, adult-onset Still's disease (AOSD) is a rare multisystemic disorder considered as a complex multigenic autoinflammatory syndrome, characterized by several distinctive features. The condition is marked by prolonged daily fever, evanescent skin rash, polyarthralgia, neutrophilic polynucleosis, hyperferritinemia, and a decreased glycosylated fraction of ferritin [1-2]. While the exact cause of AOSD remains unclear, it is believed that environmental exposures, particularly viral infections, can act as triggers for the disease.

The pathogenesis of AOSD involves the activation of an uncontrolled inflammatory response, primarily mediated by innate immune cells such as polynuclear cells and macrophages. Exogenous factors, known as pathogen-associated molecular patterns, or endogenous factors, referred to as damage-associated molecular patterns, can both serve as danger signals that initiate the aberrant inflammatory reaction. As a consequence, proinflammatory cytokines are released, leading to systemic inflammation and, in severe cases, even macrophage activation syndrome[2]. Hence, AOSD is considered an IL-1, IL-6, and IL-18-driven disease in which one of the major events in the pathogenesis is a dysregulation of inflammasome complex and a related overproductionof active IL-1b promoted by IL-18.

Cardiac manifestations associated with Still's disease exhibit a wide range of involvement, affecting all three layers of the heart. The pericardium is the most commonly affected layer, observed in 3% to 37% of patients [3]. Pericarditis is often accompanied by pleural effusion in 60% to 80% of cases [4]. Initial treatment typically involves nonsteroidal anti-inflammatory drugs, with corticosteroid therapy proving effective as a first-line approach. However, if corticosteroid therapy fails to yield improvement or to reduce reliance on cortisone, early administration of anakinra (an interleukin 1R antagonist) or tocilizumab may be considered. In cases with severe systemic manifestations, immediate consideration of biotherapies is warranted [5,6].

Cardiac MRI is the reference examination for characterizing myocardial lesions using specific sequences, with or without the use of contrast agents. Late gadolinium enhancement following contrast agent injection is a sensitive and effective method for detecting sub-epicardial or mid-wall areas of myocardial necrosis, frequently affecting the lateralwall[7]. Thallium-201 scintigraphy can also be used for diagnosis by demonstrating non-reversible hypofixation indicative of myocardial necrosis sequelae.

Follow-Up:

At the 3-month follow-up, the patient was asymptomatic, and the clinical examination revealed no abnormalities. Additionally, the electrocardiogram, transthoracic echocardiogram (TTE), and results from the maximal exercise test were all within normal limits.

Conclusion:

Cardiac involvement in adult-onset Still's disease (AOSD) is infrequent but can have severe consequences, including potentially fatal acute myocarditis or significant morbidity. The recommended treatment approach involves administering high doses of corticosteroids and promptly initiating biotherapies.

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