

Adult-Onset Still's Disease Revealed by Myopericarditis: A Case Report from Parakou, Benin

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Abstract

Introduction: Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology, characterized by marked clinical polymorphism and the absence of a specific diagnostic biomarker. Cardiac involvement, particularly myopericarditis, is uncommon and may represent a diagnostic challenge.

Case Presentation: We report the case of a 25-year-old male with no significant past medical history, admitted for febrile myopericarditis. The initial clinical presentation included high-grade fever (40.5°C), retrosternal chest pain, marked neutrophilic leukocytosis ($25.3 \times 10^9/L$ with 88.9% neutrophils), elevated inflammatory markers, and increased cardiac troponin I levels. Transthoracic echocardiography revealed a moderate circumferential pericardial effusion with preserved left ventricular systolic function (ejection fraction 60%). During hospitalization, the patient developed diffuse disabling arthralgia, odynophagia with an erythematous pharynx, and a transient evanescent rash. The diagnosis of AOSD was established according to the Yamaguchi classification criteria. High-dose corticosteroid therapy (prednisone 1 mg/kg/day) was initiated, leading to rapid clinical improvement, with complete resolution of fever and arthralgia within two weeks.

Conclusion: This case highlights an atypical presentation of AOSD revealed by myopericarditis and underscores the diagnostic challenges encountered in resource-limited settings. Early recognition and prompt corticosteroid therapy are associated with favorable outcomes.

Keywords: Adult-onset Still's disease; Myopericarditis; Fever; Neutrophilic leukocytosis; Benin

1. Introduction

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology, classically characterized by the triad of high spiking fever, arthralgia, and an evanescent salmon-pink rash [1]. Its estimated prevalence ranges from 0.16 to 0.4 cases per 100,000 inhabitants in Western countries [2]. Epidemiological data from sub-Saharan Africa remain scarce, making it difficult to assess the true burden of the disease in this region. Diagnosis relies primarily on classification criteria, with the Yamaguchi criteria being the most widely used in clinical practice [3]. Hyperferritinemia is considered a useful, although non-specific, laboratory marker [4]. Cardiac involvement has been reported in 15–35% of cases, but rarely constitutes the initial manifestation [5]. Myopericarditis as the presenting

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feature is exceptional and may delay diagnosis. We report the case of a young Beninese patient in whom AOSD was diagnosed following a presentation of febrile myopericarditis.

2. Case presentation

A 25-year-old male tailor residing in Parakou, Benin, with no significant past medical, surgical, or family history, presented with a three-week history of fever, fatigue, holocranial headache, stage II dyspnea according to the NYHA classification, and palpitations. He denied tobacco, alcohol, or illicit drug use and reported no recent travel.

He described retrosternal chest pain (intensity 4/5 on a verbal rating scale), radiating diffusely across the thorax and upper limbs, exacerbated by deep inspiration and relieved by sitting forward and analgesics. Gastroesophageal reflux symptoms were also reported.

On admission, he appeared in poor general condition (WHO performance status 3). Vital signs revealed: temperature 38°C, heart rate 106 bpm, respiratory rate 22/min, blood pressure 128/85 mmHg (right arm), oxygen saturation 97% on room air. BMI was 25.18 kg/m². Cardiovascular examination showed jugular venous distension without murmurs or additional heart sounds. Pulmonary examination revealed decreased vesicular breath sounds at the right lung base. Joint examination demonstrated tenderness of elbows, wrists, knees, and ankles bilaterally without overt arthritis. No rash was observed at admission. No lymphadenopathy was detected.

Initial laboratory tests showed:

- Leukocytosis: $19.5 \times 10^9/L$ (92% neutrophils), later peaking at $25.3 \times 10^9/L$ (88.9% neutrophils)
- Progressive anemia (hemoglobin decreased from 12.2 g/dL to 7.5 g/dL)
- Elevated CRP (48 mg/L)
- Mild transaminase elevation
- Severe hyponatremia (122 mEq/L) and hypokalemia (2.73 mEq/L)
- Markedly elevated high-sensitivity troponin I (20.7 ng/mL, then 8.8 ng/mL)
- Blood cultures were negative. Malaria testing, hepatitis B surface antigen, anti-HCV antibodies, and GeneXpert testing for tuberculosis were negative. Thyroid-stimulating hormone was normal.

Electrocardiography showed sinus tachycardia, prolonged QTc (534 ms), and inferior ST-segment elevation consistent with acute pericarditis.

Transthoracic echocardiography revealed:

- Moderate circumferential pericardial effusion (11 mm in diastole)
- Preserved left ventricular ejection fraction (60%, Simpson biplane method)
- No significant valvular abnormalities

Thoracoabdominal CT scan confirmed moderate pericardial effusion, non-necrotic mediastinal lymphadenopathy (≤ 10 mm), mild right posterior basal consolidation, bilateral atelectatic bands, and homogeneous hepatomegaly.

2.1. Diagnostic Reasoning and Management

Initial differential diagnoses included infectious myopericarditis, post-streptococcal rheumatic carditis (elevated ASLO), and hematologic malignancy. However, negative blood cultures, absence of immature cells on peripheral smear, and clinical evolution ruled out these conditions. Persistent high spiking fever (up to 40.5°C), disabling arthralgia, odynophagia with erythematous pharyngitis, and transient rash strongly suggested AOSD.

The patient fulfilled four major Yamaguchi criteria:

- Fever $\geq 39^\circ\text{C}$ for ≥ 1 week
- Arthralgia ≥ 2 weeks
- Leukocytosis $\geq 10 \times 10^9/L$ with $\geq 80\%$ neutrophils
- Typical rash

And two minor criteria:

- Pharyngitis
- Abnormal liver function tests

Infectious, neoplastic, and autoimmune causes were excluded (negative ANA and rheumatoid factor).

Given the severity (myopericarditis, persistent hyperpyrexia, disabling arthralgia), corticosteroid therapy (prednisone 1 mg/kg/day) was initiated. Clinical improvement was rapid, with complete resolution of fever and arthralgia within two weeks.

A gradual taper to 0.1 mg/kg/day over six weeks was planned with close clinical and laboratory monitoring.

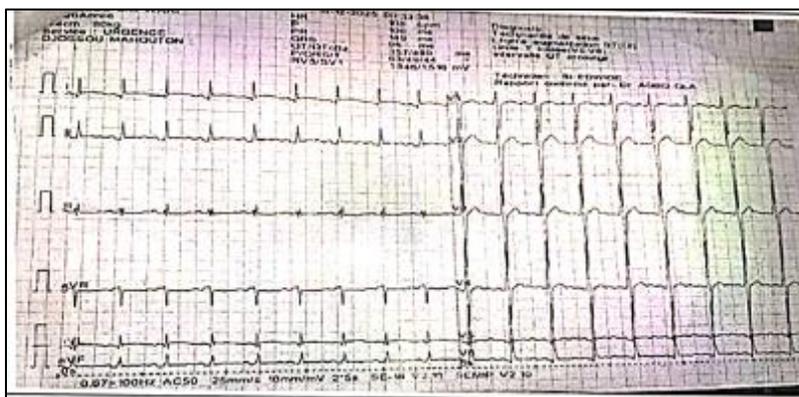


Figure 1 Electrocardiogram showing regular sinus tachycardia at 108 beats per minute, a normal cardiac axis, normal and constant PR interval, left atrial enlargement, and QTc prolongation to 534 ms, with ST-segment elevation in the inferior leads suggestive of acute pericarditis.



Figure 2 Transthoracic echocardiography showing a circumferential pericardial effusion measuring 11 mm in diastole (moderate in size) without hemodynamic compromise. Left ventricular systolic function is preserved, with a left ventricular ejection fraction of 60% by the biplane Simpson method

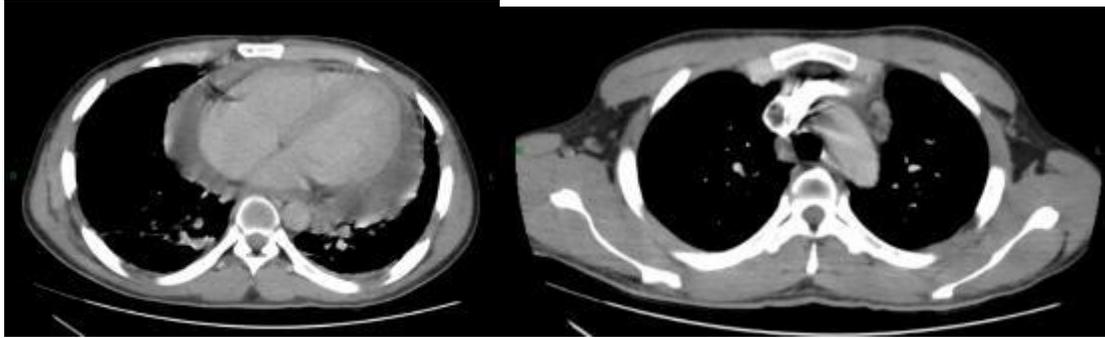


Figure 3 Thoracoabdominopelvic computed tomography showing a moderate pericardial effusion and non-necrotic mediastinal lymphadenopathy, the largest measuring 10 mm in short-axis diameter

3. Discussion

Adult-onset Still's disease is a rare condition, and documented cases in sub-Saharan Africa are exceptional. This report contributes to the epidemiological data on this disorder in our region, where the literature remains scarce. The paucity of African publications likely reflects an underestimation of the true prevalence, related to diagnostic challenges in resource-limited settings. The inaugural presentation as myopericarditis represents a remarkable feature of our observation. Cardiac involvement in adult-onset Still's disease is reported in 15–35% of cases and includes pericarditis (31%), myocarditis (10%), cardiac tamponade (4%), and endocarditis (2%) [5-6]. However, these cardiac manifestations rarely constitute the initial mode of presentation. Myopericarditis may mislead the initial diagnosis toward an infectious etiology (viral or bacterial) or post-streptococcal rheumatic carditis, as initially suspected in our patient [7]. The presentation with myopericarditis, marked leukocytosis, and elevated antistreptolysin O titers initially suggested post-streptococcal rheumatic carditis. However, the patient's age (25 years), absence of a history of acute rheumatic fever, negative blood cultures, and clinical course allowed this hypothesis to be excluded. The marked leukocytosis ($25.3 \times 10^9/L$) also initially raised suspicion of a hematologic malignancy, which was ruled out by the absence of immature cells on peripheral smear and clinical evolution [8]. Other differential diagnoses to consider in the setting of prolonged fever with leukocytosis include severe bacterial infections, malignancies (particularly lymphomas), systemic vasculitides, and systemic lupus erythematosus. Clinical evolution and response to therapy are often decisive in establishing the diagnosis [9]. The diagnosis of adult-onset Still's disease remains one of exclusion due to the absence of a specific biological marker. The most widely used classification criteria are those of Yamaguchi (sensitivity 96%, specificity 92%), which require the presence of at least five criteria, including two major criteria, after exclusion of infections, malignancies, and other systemic diseases [3]. Our patient fulfilled four major criteria (fever $\geq 39^\circ C$ for at least one week, arthralgia for at least two weeks, leukocytosis $\geq 10 \times 10^9/L$ with $\geq 80\%$ neutrophils, and typical rash) and two minor criteria (pharyngitis and elevated transaminases with hypoalbuminemia reflecting systemic inflammation), for a total of six criteria supporting the diagnosis with high reliability. Neutrophilic leukocytosis is a characteristic laboratory feature, present in 60–100% of patients [5]. In our case, the leukocyte count reached $25.3 \times 10^9/L$ with 88.9% neutrophils, values well above typical diagnostic thresholds. Hyperferritinemia, although non-specific, is an important laboratory marker in the diagnosis of Still's disease [4, 10]. Ferritin levels greater than five times the upper limit of normal are found in 70–80% of patients, and a glycosylated ferritin fraction below 20% has good diagnostic value [11]. A ferritin level exceeding 1000 ng/mL with a glycosylated fraction below 20% is highly suggestive of Still's disease. Unfortunately, ferritin testing could not be performed in our setting due to reagent unavailability, illustrating the diagnostic challenges encountered in resource-limited countries. Our observation highlights the diagnostic challenges encountered in a limited technical setting. The absence of ferritin measurement, inability to perform a complete immunological workup (antinuclear antibodies, rheumatoid factor, anti-CCP antibodies), and lack of bone marrow biopsy made diagnosis more difficult and necessitated reliance on rigorous clinical reasoning and application of the Yamaguchi criteria. These limitations are common in sub-Saharan Africa and likely contribute to underestimation of the true prevalence of Still's disease in this region. They underscore the importance of clinician training to recognize the classical manifestations of the disease and apply validated diagnostic criteria [12]. Management of adult-onset Still's disease includes nonsteroidal anti-inflammatory drugs as first-line therapy in moderate forms, corticosteroids in severe or resistant cases, and immunosuppressive agents (methotrexate) or biologic therapies (anti-IL-1, anti-IL-6, anti-TNF) in refractory disease [13]. Given the severity of our patient's presentation (myopericarditis, persistent high fever, disabling arthralgia, and marked inflammatory syndrome), high-dose corticosteroid therapy was initiated promptly. The excellent response observed after two weeks (complete defervescence and resolution of arthralgia) retrospectively confirms the diagnosis and highlights the favorable

prognosis when the disease is recognized and treated early [14]. Cases with cardiac involvement generally require high-dose corticosteroid therapy and close monitoring [6].

4. Conclusion

Adult-onset Still's disease is a rare systemic inflammatory disorder whose diagnosis relies on clinical and biological criteria in the absence of specific biomarkers. This case emphasizes that AOSD should be considered in any patient presenting with prolonged fever and marked neutrophilic leukocytosis, even when initial presentation is dominated by cardiac involvement such as myopericarditis. Early diagnosis and prompt initiation of corticosteroid therapy are associated with favorable outcomes, particularly in severe presentations.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

The patient's consent for the writing and publication of the article was obtained.

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