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Acute perimyocarditis as the initial manifestation of adult-onset Still's disease

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Abstract. Adult-onset Still's disease (AOSD) is a rare systemic autoinflammatory disorder characterized by quotidian high-spiking fever, inflammatory arthralgia, and an evanescent salmon-coloured rash. Cardiac involvement is uncommon but may include pericarditis and, less frequently, myocarditis. We report the case of an 18-year-old male presenting with persistent fever, myalgias, and arthralgia. Laboratory investigation demonstrated markedly elevated C-reactive protein (CRP 419 mg/L) and increased high-sensitivity cardiac troponin T (10511 ng/L). Electrocardiography showed sinus tachycardia with T-wave inversion in the inferolateral leads, and transthoracic echocardiography revealed a small posterior pericardial effusion without ventricular dysfunction. A diagnosis of non-complicated acute perimyocarditis was established, and anti-inflammatory therapy was initiated. During hospitalization, recurrent high-grade fever spikes were associated with progression of an evanescent maculopapular rash and dynamic troponin variation. Marked hyperferritinaemia (11495 µg/L) was subsequently documented. Extensive infectious and autoimmune evaluation was unremarkable. The constellation of clinical and laboratory findings fulfilled classification criteria for AOSD. High-dose intravenous methylprednisolone led to rapid clinical and biochemical improvement. Corticosteroids were gradually tapered, and adalimumab was introduced as a steroid-sparing strategy, with no recurrence during follow-up.

This case highlights acute perimyocarditis as a potential initial manifestation of AOSD and underscores the importance of considering systemic autoinflammatory disorders in patients presenting with inflammatory myopericardial syndromes and persistent systemic inflammation. Fig. 3, Tab 1, Ref. 11, on-line full text (Free, PDF), www.cardiologyletters.sk

Keywords: Adult-onset Still's disease – Perimyocarditis – Hyperferritinaemia – Autoinflammatory disease

Acute perimyocarditis is characterized by concomitant inflammation of the myocardium and pericardium and may result from infectious, autoimmune, or autoinflammatory mechanisms (1 – 3). Although viral infection remains the most frequent aetiology in young individuals, systemic inflammatory disorders should be considered when cardiac involvement occurs in association with persistent fever and marked systemic inflammatory activation (3).

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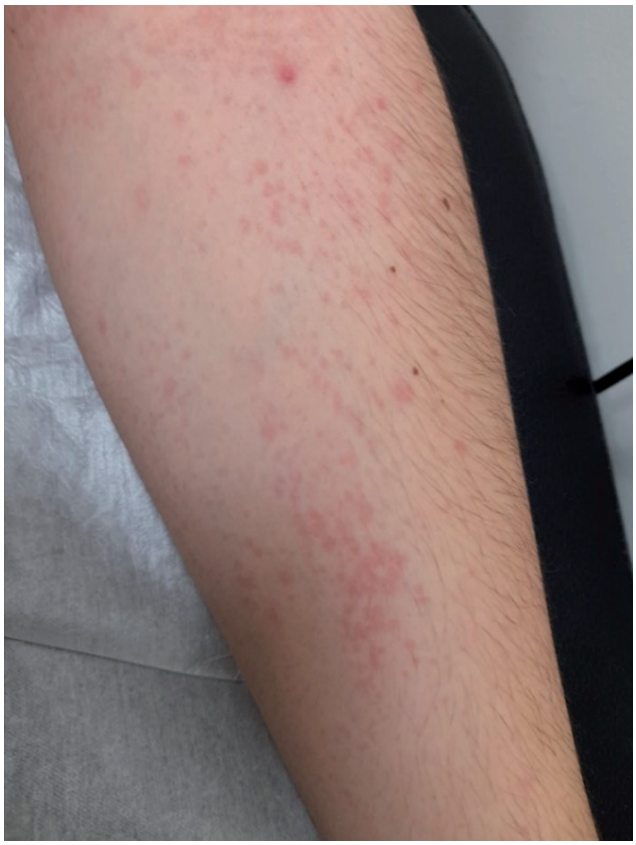

Figure 1 Maculopapular lesions over the left limb

Adult-onset Still's disease (AOSD) is a rare systemic autoinflammatory disorder characterized by quotidian high-spiking fever, evanescent salmon-coloured rash, inflammatory arthralgia or arthritis, neutrophilic leukocytosis, and marked hyperferritinaemia (4 – 6). Cardiac involvement is uncommon but may present as pericarditis and, less frequently, myocarditis (6 – 7). We report a case in which acute perimyocarditis represented the initial clinical manifestation of AOSD.

Case Presentation

An 18-year-old male was admitted with a 7-day history of high-grade fever, asthenia, anorexia, diffuse myalgias, and arthralgia, accompanied by mild odynophagia. His medical history was unremarkable, except for a transient pruritic rash previously treated with antihistamines.

On admission, the patient was febrile (38.1°C), tachycardic (105 beats/min), normotensive (120/72 mmHg), and had an oxygen saturation of 96% on room air. Dermatological examination identified scattered erythematous macular lesions over the limbs (**Figure 1**) and thorax, without lymphadenopathy. Cardiopulmonary examination was otherwise unremarkable.

Laboratory evaluation demonstrated leukocytosis (13.3×10^9/L) with neutrophilia (84%), markedly elevated C-reactive protein (CRP) 419 mg/L, and increased high-sensitivity cardiac troponin T (hs-cTnT) 10511 ng/L. Serum ferritin was markedly elevated at 11495 µg/L. Mild transaminase elevation was observed. Procalcitonin was 0.19 ng/mL, and serial blood cultures remained negative.

Electrocardiography showed sinus tachycardia with T-wave inversion in the inferolateral leads (**Figure 2**).

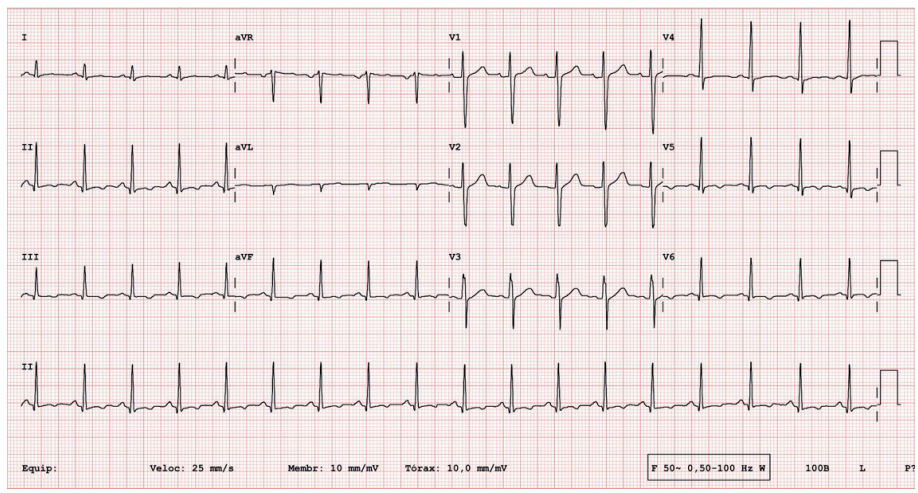

Figure 2 ECG showing sinus tachycardia with T wave inversion inferolaterally

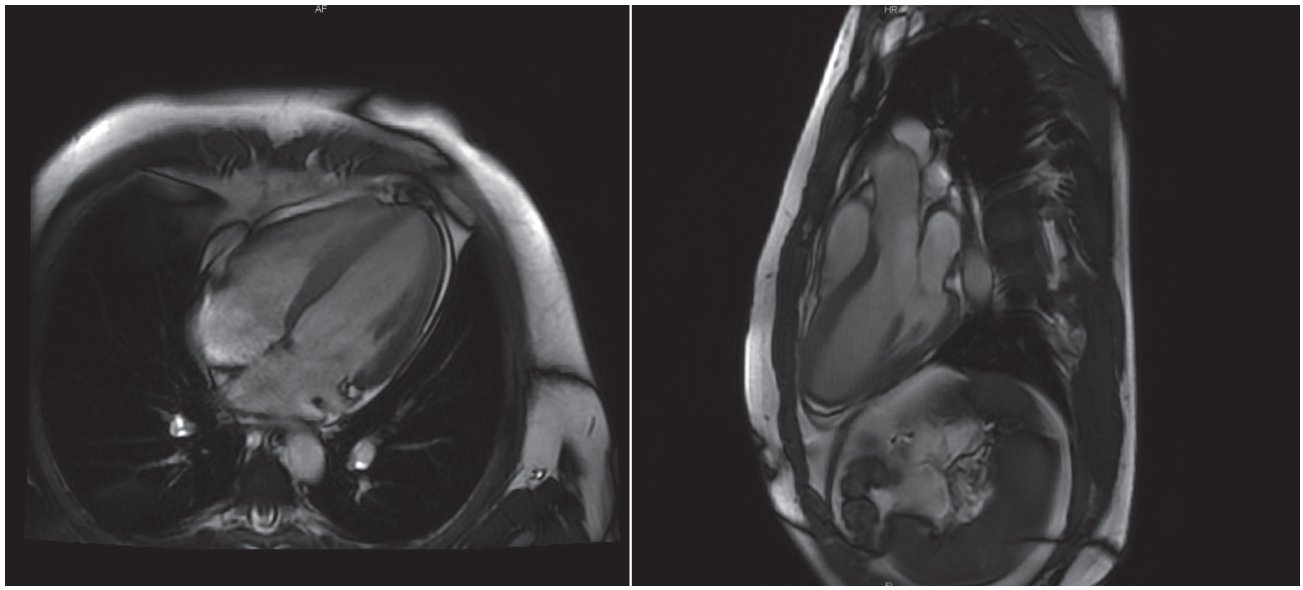

Figure 3 Cardiac magnetic resonance performed three weeks after presentation without late gadolinium enhancement or residual myocardial oedema

Transthoracic echocardiography identified a small posterior pericardial effusion without left ventricular systolic dysfunction.

In accordance with the 2025 guidelines of the European Society of Cardiology for the management of myocarditis and pericarditis, the clinical presentation and objective evidence of myocardial injury fulfilled criteria for suspected acute inflammatory myopericardial syndrome (8). The absence of haemodynamic instability, sustained ventricular arrhythmias, or ventricular dysfunction allowed classification as non-complicated acute myocarditis (8).

The patient was admitted for continuous rhythm monitoring and initiated on ibuprofen and colchicine due to concomitant pericardial involvement.

During hospitalization, recurrent quotidian fever spikes up to 40°C were documented, accompanied by progression of a salmon-coloured maculopapular rash accentuated during febrile episodes. Bilateral inflammatory arthralgia involving the metacarpophalangeal and interphalangeal joints subsequently developed.

Inflammatory markers further increased (peak CRP 900 mg/L), whereas serial hs-cTnT measurements showed dynamic variation without clinical deterioration. Continuous monitoring did not reveal arrhythmic events.

Comprehensive infectious evaluation, including viral serologies and interferon-γ release assay, was negative. Autoimmune screening, including antinuclear antibodies, rheumatoid factor, and antineutrophil cytoplasmic antibodies, was unremarkable. Cross-sectional imaging excluded lymphoproliferative disease.

After exclusion of infectious, malignant, and alternative autoimmune conditions, the combination of quotidian fever, evanescent rash, inflammatory arthralgia, neutrophilic leukocytosis, and marked hyperferritinaemia fulfilled Yamaguchi classification criteria for adult-onset Still's disease (**Table 1**) (9).

Table 1 Yamaguchi Diagnostic Criteria for Adult-Onset Still's Disease. Diagnostic requirement: At least 5 criteria must be present, including at least 2 major criteria, after exclusion of infections, malignancies, and other rheumatic diseases.

MAJOR CRITERIA
Fever ≥39°C lasting ≥1 week
Arthralgia lasting ≥2 weeks
Evanescent (salmon-pink) rash
Leukocytosis ≥10,000/mm^3 (≥80% neutrophils)
MINOR CRITERIA
Sore throat
Lymphadenopathy
Hepatomegaly and/or splenomegaly
Abnormal liver function tests
Negative antinuclear antibody and rheumatoid factor

Cardiac magnetic resonance imaging performed three weeks after presentation demonstrated preserved biventricular volumes and systolic function, without late gadolinium enhancement or residual myocardial oedema (**Figure 3**). No features of active myocardial inflammation were identified according to the updated Lake Louise criteria (10), consistent with interval resolution of the acute inflammatory process. In the absence of haemodynamic compromise, malignant ventricular arrhythmias, or progressive ventricular dysfunction, endomyocardial biopsy was not performed, in line with contemporary guideline recommendations (8).

High-dose intravenous methylprednisolone (1 g/day for three consecutive days) was administered, resulting in rapid defervescence and a marked decline in inflammatory markers. Cutaneous manifestations resolved promptly. Corticosteroids were gradually tapered, and adalimumab was introduced as a steroid-sparing strategy, in line with contemporary therapeutic approaches for systemic AOSD (5, 6).

At follow-up, the patient remained asymptomatic, with normalization of CRP and hs-cTnT levels. Repeat echocardiography confirmed preserved ventricular function and complete resolution of the pericardial effusion.

Discussion

Myocarditis is a clinically heterogeneous condition with a broad spectrum of presentation and prognosis (1 – 3). The 2025 ESC guidelines integrate myocarditis and pericarditis under the concept of inflammatory myopericardial syndrome and provide structured diagnostic and risk stratification criteria (8).

Although pericarditis represents the most frequent cardiac manifestation of AOSD, myocardial involvement, albeit less common, may lead to significant ventricular dysfunction if not promptly recognized (6, 7). Early identification of the underlying systemic inflammatory disorder is therefore essential, as timely immunosuppressive therapy may prevent adverse cardiac outcomes.

Marked hyperferritinaemia, particularly values exceeding 10000 µg/L, strongly supports the diagnosis of AOSD in the appropriate clinical context (4, 5). In young patients presenting with presumed viral perimyocarditis, the coexistence of persistent high-spiking fever, extreme hyperferritinaemia, neutrophilic leukocytosis, and an evanescent rash should prompt consideration of autoinflammatory disease (4 – 6).

Management in this case was aligned with ESC recommendations for non-complicated myocarditis, including in-hospital surveillance and temporary restriction from intense physical activity (8). Current position statements advise avoidance of competitive sports for at least 3–6 months following acute myocarditis, with comprehensive reassessment prior to return to play (11).

Conclusion

Acute perimyocarditis may represent the initial clinical manifestation of adult-onset Still's disease. In patients presenting with inflammatory myopericardial syndrome accompanied by persistent systemic inflammation, clinicians should maintain a high index of suspicion for autoinflammatory disorders. Guideline-directed evaluation and multidisciplinary management are essential to ensure timely diagnosis and favourable clinical outcomes.

Luís Cotrim and António Carvalho contributed equally to this case report

References

1. Caforio ALP, Pankuweit S, Arbustini E, Basso C, Gimeno-Blanes J, Felix SB, et al. Current state of knowledge on aetiology, diagnosis, management, and therapy of myocarditis. Eur Heart J 2013;34:2636-2648.
2. Tschöpe C, Ammirati E, Bozkurt B, Caforio ALP, Cooper LT, Felix SB, et al. Myocarditis and inflammatory cardiomyopathy: current evidence and future directions. Nat Rev Cardiol 2021;18:169-193.
3. Kindermann I, Barth C, Mahfoud F, Ukena C, Lenski M, Yilmaz A, et al. Update on myocarditis. J Am Coll Cardiol 2012;59:779-792.
4. Gerfaud-Valentin M, Jamilloux Y, Iwaz J, Sève P. Adult-onset Still's disease. Autoimmun. Rev. 2014;13:708-722.
5. Feist E, Mitrovic S, Fautrel B. Mechanisms, biomarkers and targets for adult-onset Still's disease. Nat Rev Rheumatol 2018;14:603-618.
6. Fautrel B. Adult-onset Still disease. Best Pract Res Clin Rheumatol 2008;22:773-792.
7. Bodard Q, Langlois V, Guillevin L, Papo T. Cardiac involvement in adult-onset Still's disease: manifestations and management. Rheumatology (Oxford) 2010;49:2159-2166.

8. Schulz-Menger J, Collini V, Gröschel J, Adler Y, Brucato A, Christian V, et al. 2025 ESC Guidelines for the management of myocarditis and pericarditis: Developed by the Task Force for the management of myocarditis and pericarditis of the European Society of Cardiology (ESC). Eur Heart J. 2025;46:3952–4041. doi:10.1093/eurheartj/ehaf192.
9. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, et al. Preliminary criteria for classification of adult Still's disease. J Rheumatol 1992;19:424-430.
10. Ferreira VM, Schulz-Menger J, Holmvang G, Kramer CM, Carbone I, Sechtem U, et al. Cardiovascular magnetic resonance in nonischaemic myocardial inflammation: expert recommendations. Eur Heart J 2018;39:2828-2839.
11. Pelliccia A, Solberg EE, Papadakis M, Adami PE, Biffi A, Caselli S, et al. Recommendations for participation in competitive sport in athletes with cardiovascular disease. Eur Heart J 2021;42:17-96.