Case study

A rare case of Adult-Onset Still's Disease revealed by acute myopericarditis.

Abstract

Background :

Still's disease in adults is a systemic inflammatory pathology of unknown aetiology, characterized by clinical manifestations associating feverish peaks, arthritis or arthtralgia, transient rashes and hyperferritinemia.

Currently, this disease remains a multisystemic disease with generally poor outcome, poorly described in the literature with very few studies unlike other rheumatic diseases, probably underdiagnosed due to its clinical polymorphism.

Cardiac forms are quite rare and among the manifestations described, pericarditis remains the most reported entity, myocardial involvement is exceptional.

the aim of our observation is to report an exceptional case of discovery of stille disease following a myo-pericarditis mimicking a coronary Sd initially

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INTRODUCTION

Adult Still's disease (ASD) is a systemic inflammatory disease of unknown aetiology, typically affecting young adults. characterized by clinical manifestations associating feverish peaks, arthritis or arthtralgia, fleeting rashes and hyperferritinemia [1,2]. Still's disease was first described in 1897, in 22 children with systemic onset juvenile idiopathic arthritis (by George Still [3]. Subsequently, in 1971, Eric Bywaters described a serie of 14 adult patients, presenting with rash, fever, polyarthritis, which strongly resembled pediatric Still's disease symptoms, thus defining a new clinical entity: Still's disease in adults [4].
Currently, this disease remains a rare multisystem disorder of unknown aetiology and usually poor outcomes, not well described in the literature with very few studies unlike other rheumatic diseases, probably underdiagnosed due to its clinical polymorphism[1–5]. Cardiac forms are quite rare and among the manifestations described, pericarditis remains the most reported entity, myocardial involvement is exceptional.

We here report the case of a young patient admitted foracute myocarditis revealing Still's disease with multisystem involvement.

Case report

A 26-year-old male patient presented to the emergency department with exertional dyspnoea that first started 2 months ago, and progressively worsened into Stage III according to the New York Heart Association (NYHA) classification, associated withleft-sided chest pain at rest radiating to the upper limbs.

He had history of recurrent fevers, migrating polyarthralgia affecting his knees and elbows, as well as an episode of intermittent ankle swelling in the past few months.

On physical examination, the patient had a blood pressure of 130 / 85mmHg, a heart rate of 120 beats/minute, oxygen saturation in ambient air was 88%, his body temperature was 38.8°C. His pulmonary auscultation found bilateral basal crackles bases and cardiac auscultation revealed a pronounced pulmonic component of the second heart soundin addition to a discrete apical systolic murmur (2/6).

The patient's electrocardiogram (ECG) showed a right bundle branch block with abnormal repolarization as well as a ST segment depression in the inferior leads (Figure 1).
Chest X-rayfound a non specific interstitial syndrome.
Blood tests assessed high cardiac markers: High sensitive troponin level at 4500 pg / ml, a biological inflammatory syndrome: C- reactive protein (CRP) at 140 mg / ml, a Sedimentation velocity (SV) of 110 mm, High white blood cell count with neutrophils at 8200/ mm3.

Thyroid hormones, procalcitonin, immunoglobulins and complement were normal. Rheumatoid factor, antinuclear antibodies, extractable nuclear antigen, anti-native DNA antibodies, cytoplasmic antineutrophils and antiphospholipid antibodies were also negative.

Following these findings, a Transthoracic echocardiography was performed in our patient and revealed a non dilated left ventricle with the inferior wall hypokinesia with a preserved ejection fraction of 55%, in addition to high filling pressures;moderate mitral regurgitation (MR), a moderate tricuspid regurgitationwith an estimated pulmonary artery pressure of 49 mmHg with no right chambers' dilatation nor right ventricular dysfunction. Finally, minimal circumferential pericardial effusion was found.

A coronary angiography was also performed considering the high troponin levels, the ST segment depression in the EGG and the regional wall motion abnormalities, which turned out to be normal (Figure 2).

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We then completed our investigations with a cardiac Magnetic Resonance Imaging(MRI)that showed an increased T2 signal ofthe infero-lateral and inferior basal segments indicative of myocardial edema. Moreover, delayed contrast-enhanced images showed diffuse subepicardial enhancement with no involvement of the subendocardium, in addition to a mild pericardial effusion regarding thelateral wall of the left ventricle (LV) (Figure 3), thus concluding to a diagnosis of acute myopericarditis.

In our patient, we retained the diagnosis of Adult form of Still's disease, considering the combination of clinical criteria (fever, arthralgia, arthritis) and biological ones (hyperferritinemia, and normal bacteriological, viral and immunological assessment). Medical treatment including corticosteroids, metothrexate and interleukin 1 inhibitors was started, with good clinical, biological and echocardiographic evolution.

DISCUSSION

Adult onset Still's disease (AOSD) is a rare systemic inflammatory disease, causing fever, rash, and arthritis. Biological findings show neutrophilic granulocytosis, high levels of ferritin, mildly disturbed liver enzymes, and negative results for rheumatoid factor and antinuclear antibodies [6-7]. The diagnosis is mostly clinical, often based on the criteria of Yamaguchi [7], which are the most used; and the positive diagnosis require 5 criteria including at least 2 major ones.
In the absence of pathognomonic features, the diagnosis of AOSD is usually considered after excluding other conditions, especially infections, neoplasms (especially lymphomas) and autoimmune disorders (particularly, vasculitis and polymyositis).
All these features are assessed in Table 1.

The most common sites of cardiopulmonary involvement are the pleural and pericardial cavities by inflammatory mechanism, thus, cardiac manifestations in AOSD usually present as pericarditis [1]. Its association with myocarditis is very rare and the literature on the subject remains scarce even in the largest series of cases although it is a severe manifestation of the disease [8-9].

Myocarditis is less common, accounting for approximately 3% of all cases of cardiac involvement reported [10-11] and may be complicated by complete atrioventricular block, tachyarrhythmia, heart failure, and even cardiogenic shock with a high risk of mortality. Endocardial involvement is even rarer and may present as non-infectious endocarditis.

Considering the frequency and severity of cardiac involvement in AOSD, all patients with this condition should undergo cardiovascular screening with clinical, electrical and echocardiographic assessmentto look for cardiac complications.

Treatment of acute myocarditis in AOSD is not different from acute myocarditis basic treatment in the general population, including symptomatic treatment such as loop

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diureticsin case of congestive signs, and heart failure treatment with betablockers and/or angiotensin-converting enzyme (ACE) inhibitors or Angiotensin receptor blockers (ARBs) in case of LV systolic dysfunction.
In addition to etiological treatment with corticosteroids as first-line treatment in patients with myocarditis, since they proved their effectiveness in this indication[12]. Their initiation should not be delayed if there is a high probability for the disease.
In case of relapse or resistance to treatment, intravenous immunoglobulins, methotrexate, and tumor necrosis factor (TNF)-alpha inhibitors such as etanercept can be used.
Non-steroidal anti-inflammatorydrugs (NSAIDs) are generally avoided because they can prevent myocardial healing.

Finally, these patients should undergo long-term follow-up with repeated echocardiography and should refrain from intense physical activity until full myocardial function recovery is documented by non-invasive imaging.

CONCLUSION:

To conclude, our patient’s case provides good evidence that adult Still's disease should be considered in the diagnosis work-up of patients with myopericarditis, especially in the presence of systemic involvement.It also underlines the importance to perform serial screening echocardiographic evaluations in patients with AOSD, for an early detection of cardiac involvement, which can progress to cardiac complications which can be irreversible and fatal.Patients usually improve undercorticosteroids and NSAIDs are avoided.

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Major criteria	• Fever >39°C, lasting 1 week or longer
	• Arthralgia or arthritis, lasting 2 weeks or longer
	• Typical rash
	• Leukocytosis >10 000/mm with >80% polymorphonuclear cells
Minor criteria	• Sore throat
	• Recent development of significant lymphadenopathy
	• Hepatomegaly or splenomegaly
	• Abnormal liver function tests
	• Negative tests for antinuclear antibody and rheumatoid factor (IgM)
Exclusion criteria	• Infections
	• Malignancies
	• Other rheumatic diseases
Five or more criteria are required with 2 or more being major criteria for diagnosis of AOSD	

Abbreviations: AOSD, adult-onset Still's disease; IgM, immunoglobulin M.

Table 1: Yamaguchi Criteria for Diagnosis of AOSD.

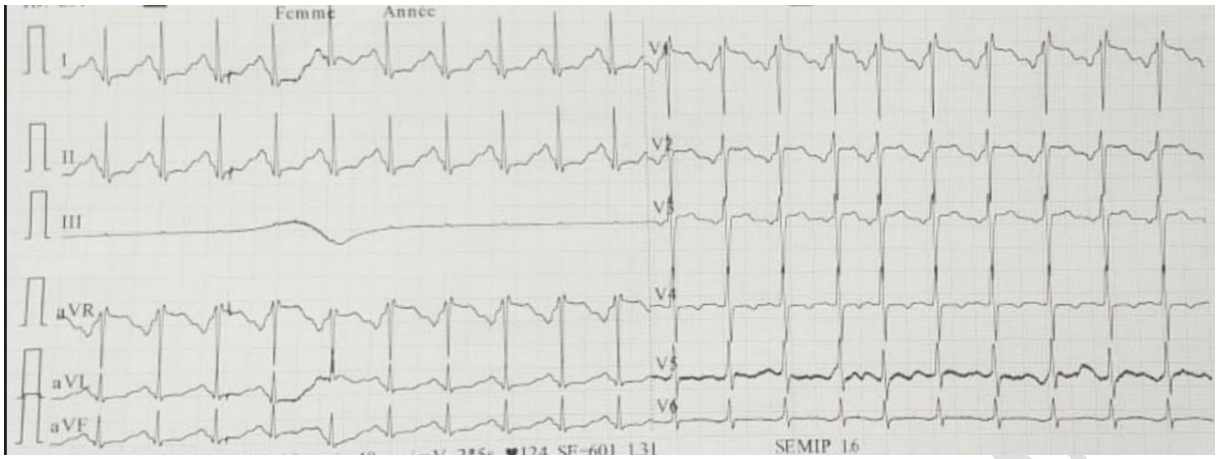

Figure 1 : Patient's ECG

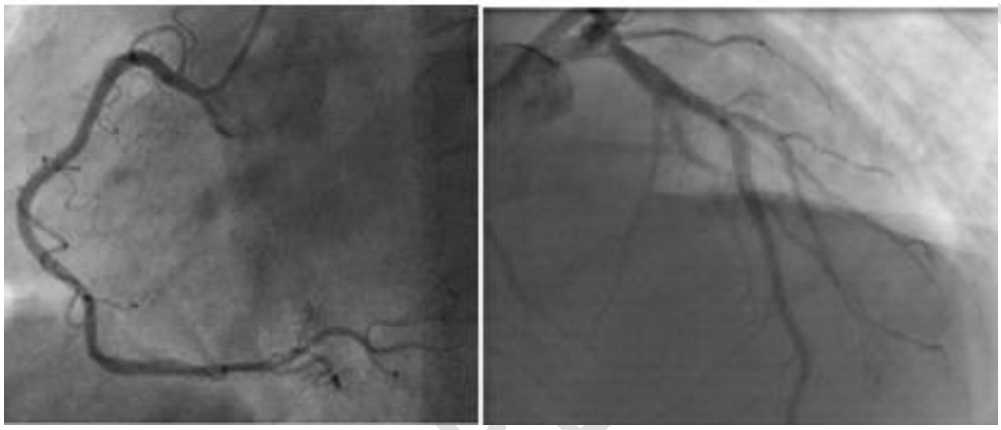

Figure 2 : patient's coronarography

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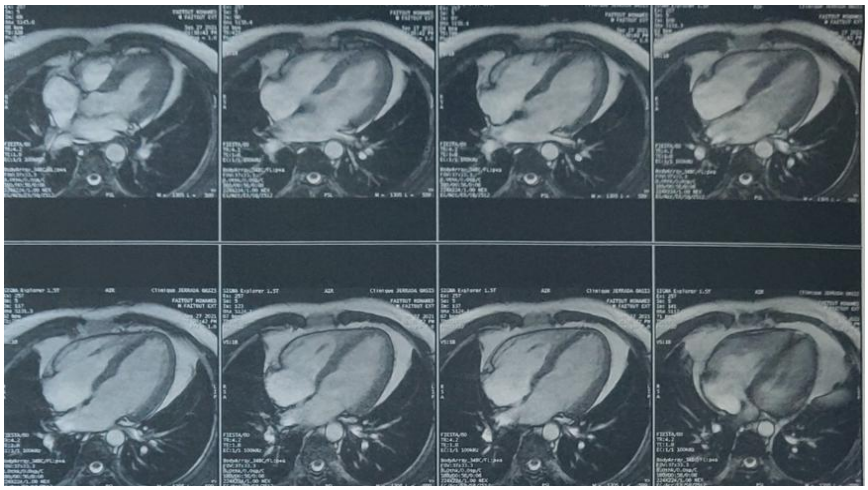

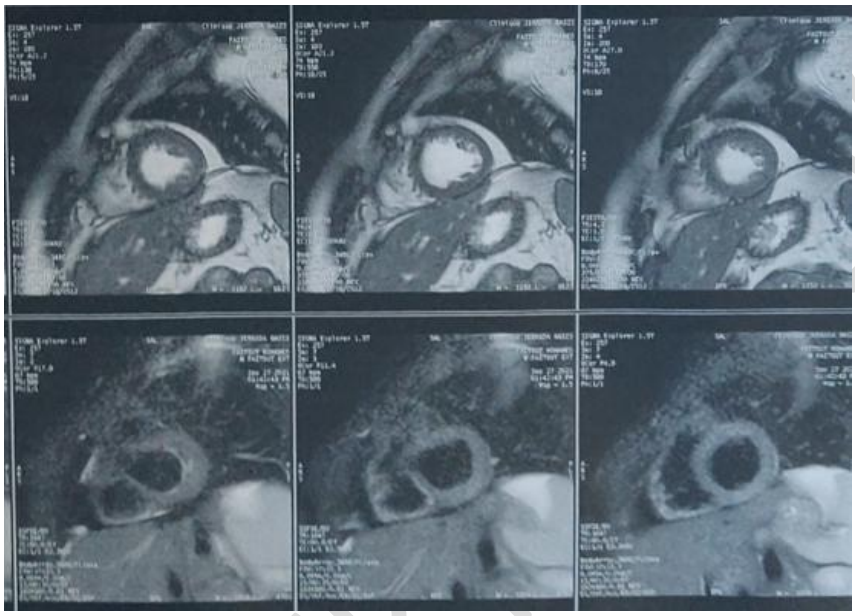

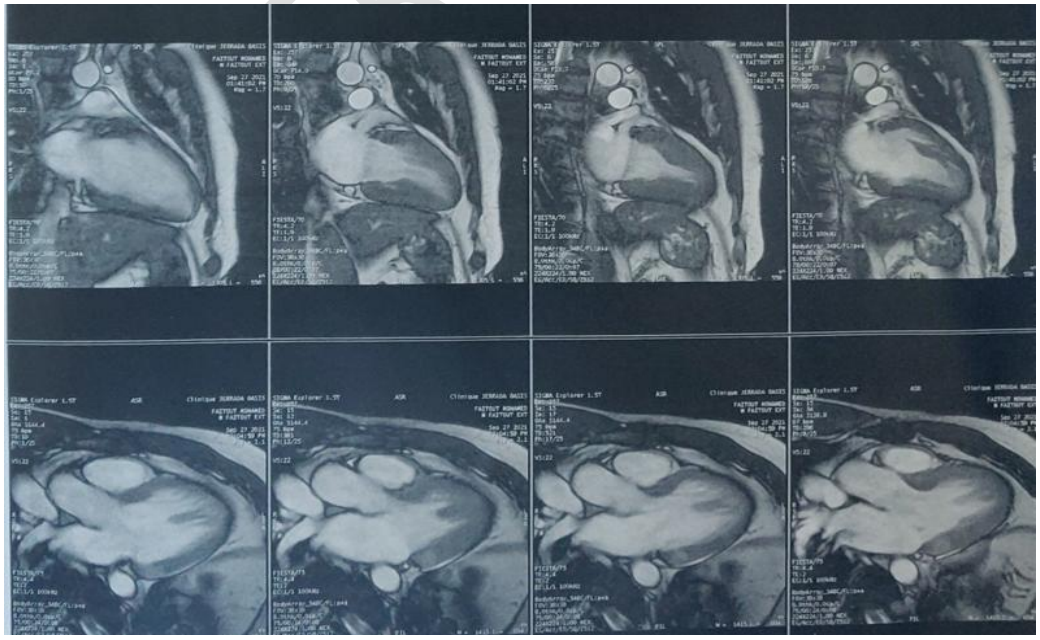

Figure 3 : patient's cardiac MI

Reference

[1] Roberto Giacomelli .A comprehensive review on adult onset Still's disease , Journal of Autoimmunity .2018

[2] M. Gerfaud-Valentin, Y. Jamilloux, J. Iwaz, P. Sève, Adult-onset Still's disease, Autoimmun. Rev. 13 (2014) 708–722.

[3] G.F. Still, On a form of chronic joint disease in children, Arch. Dis. Child. 16 (1941) 156–165.

[4] E.G. Bywaters, Still's disease in the adult, Ann. Rheum. Dis. 30 (1971) 121–133.

[5] Y. Cagatay, A. Gul, A. Cagatay, S. Kamali, A. Karadeniz, M. Inanc, et al., Adultonset Still's disease, Int. J. Clin. Pract. 63 (2009) 1050–1055.

[6] Efthimiou P, Paik PK. Diagnosis and management of adult onset Still's disease. Ann Rheum Dis [Internet] 2006;65:564–572.

[7] Yamaguchi M, Ohta A, TsunematsuT, Kasukawa R, Mizushima Y, Kashiwagi H, Kashiwazaki S, Tanimoto K, MatsumotoY, OtaT.Preliminary criteria for classification of adult Still's disease.JRheumatol 1992;19:424–430.

[8] adhav P, Nanayakkara N: Myocarditis in adult-onset Still's disease. Int J Rheum Dis. 2009, 12:272-274. 10.1111/j.1756-185X.2009.01423.x

[9] Gerfaud-Valentin M, Sève P, Iwaz J, et al.: Myocarditis in adult-onset Still's disease. Medicine. 2014, 93:280-289. 10.1097/MD.0000000000000112

[10] Moder K. G., Miller T. D., Allen G. L. Cardiac tamponade: an unusual feature of adult onset Still's disease. J Rheumatol. 1995;22(1):180–2. [PubMed] [Google Scholar]

[11] Drouot M. H., Hachulla E., Houvenagel E., Hatron P. Y., Flipo R. M., Goullard L. et al. Cardiac complications in adult onset Still's disease: from pericarditis to tamponade as manifestations [in French] Rev Med Interne. 1994;15(11):740–3. [PubMed] [Google Scholar]

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[12] Kumar M, Tandon V, Lopetegui Lia N, et al. (June 14, 2019) Still's Disease and Myopericarditis. Cureus 11(6): e4900. doi:10.7759/cureus.4900

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