Adult-onset Still’s disease: A diagnosis to remember

Ana Rita Fernandes Matos, Elisabete Coelho, Sofia Caridade

ABSTRACT

Introduction: Adult-onset Still’s disease (AOSD) is a rare systemic inflammatory disorder that occurs mainly in young people. It characteristically presents with the triad of high-grade fever, arthralgia, and salmon-colored evanescent rash, and is a diagnosis of exclusion. Case Report: We present the case of a 33-year-old man admitted for a second time to the hospital with clinical symptoms compatible with this disease. After exclusion of differential diagnosis, it was concluded that the patient had AOSD and started proper treatment with good response. Conclusion: This case highlights the importance of thinking in this diagnosis in order to make a timely diagnosis.

Keywords: Adult-onset Still’s disease, Autoinflammatory disease, Fever, Skin rash

INTRODUCTION

Adult-onset Still’s disease is a rare nonfamiliar multisystemic inflammatory disorder [1–4]. The difficulty in the diagnosis of AOSD is related to the heterogeneity and nonspecificity of clinical symptoms [2]. In the absence of pathognomonic features or diagnostic tests, this diagnosis implies a high clinical suspicion [1]. The range of differential diagnosis is extensive—for example, AOSD can mimic rheumatic diseases, malignancy, or infectious diseases/sepsis [1]. The prognosis is frequently good [4] but some patients can develop fatal complications [1, 4].

CASE REPORT

A 33-year-old man with no known chronic diseases was admitted to the hospital with a three-week history of sore throat, high-grade fever, myalgia, polyarthralgia involving predominantly the wrists and ankles and an evanescent erythematous and nonpruritic skin rash. The rash involved mainly the trunk and neck.

Further history revealed that he had been hospitalized about six months before for a similar clinical condition that lasted for about three weeks. Despite the absence of a definitive diagnosis, he became asymptomatic with anti-inflammatory therapy only and was discharged.

At admission, the physical examination revealed high-grade fever, salmon-colored evanescent rash on the trunk (Figure 1), and mildly congested throat but overall was, otherwise, unremarkable.

Blood tests showed leukocytosis (21.2 × 10^9/L), thrombocytosis (748 × 10^9/L), elevated acute phase reactants (C-reactive protein: 220 mg/L, erythrocyte sedimentation rate: 84 mm/h) and elevated liver enzymes (aspartate transaminase: 200 U/L; alanine transaminase: 175 U/L; lactate dehydrogenase concentration: 627 U/L). It was also observed a markedly elevation of serum ferritin levels (16,626 ng/mL) and hypoalbuminemia (2.3 g/dL). The remaining analytical investigations (immunological and infectious studies) were normal. Immunological panel included antinuclear
antibody, rheumatoid factor, antineutrophil cytoplasmic autoantibody, and complement. Infectious work-up included, among others, cultural tests and serologies for legionella, Mycoplasma pneumoniae, Epstein–Barr virus, respiratory viral panel, human immunodeficiency virus, cytomegalovirus, parvovirus B19, herpes virus I and II, hepatitis B and C, and Lyme disease. A computed axial tomography demonstrated hilar and mediastinal lymph nodes. A transendoscopic ultrasound-guided biopsy of the mediastinal lymph node was performed and showed nonspecific reactive lymphadenitis, excluding granulomatous or malignant lesions.

Based on his clinical presentation, laboratory results, and exclusion of differential diagnosis, the patient was diagnosed to have AOSD, using the Yamaguchi criteria.

He started anti-inflammatory therapy with indomethacin, with an initial good clinical and analytical response. However, as it was then verified a recrudescence of the fever and a re-elevation of the acute phase reactants, it was decided to change to corticosteroids therapy. With prednisolone (1 mg/kg/day) he became almost completely asymptomatic, with the exception of a persisting mild polyarthralgia. The blood tests, also, presented a good evolution (Table 1).

During the follow-up, the corticosteroids were tapered off after one month of initiation and it was started methotrexate, as a steroid sparing agent, with excellent result.

### DISCUSSION

Adult-onset Still’s disease has a prevalence of less than 1/100,000 [1] and occurs mainly in young people [1]. The etiology of AOSD is not completely understood [1, 2, 4] but it is accepted that this pathology is part of the spectrum of autoinflammatory diseases [5], which originate in a dysfunction of the innate immune system [5]. Proinflammatory cytokines have been implicated since serum and tissue levels of interleukin (IL)-1β, IL-6, and tumor necrosis factor (TNF)-α are elevated in those patients [2–4].

A clinical triad of high-grade spiking fever, arthralgia, and erythematous rash is typical of this disease [3, 4]. The rash is frequently described as a salmon-colored evanescent maculopapular skin eruption and skin biopsy is not obligatory [3]. However, it may manifest also with sore throat, myalgia, arthritis, serositis, hepatosplenomegaly, lymphadenopathy, leukocytosis, hepatic cytolysis, and hyperferritinemia [1, 2]. Regarding its temporal evolution, AOSD may present with a monocyclic or polycyclic pattern and/or it can be complicated by a chronic erosive polyarthritis [1].

The differential diagnosis is extensive [1] and there are no specific pathological finding or diagnostic test.

![Figure 1: Salmon-colored evanescent rash on the patient’s trunk.](image)

<table>
<thead>
<tr>
<th>Table 1: Evolution of analytical results of the patient</th>
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<tbody>
<tr>
<td><strong>Hospital admission</strong></td>
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<td>------------------------</td>
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<td>Leukocytes</td>
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<td>Platelets</td>
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<td>C-reactive protein</td>
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<td>Erythrocyte sedimentation rate</td>
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<td>Aspartate transaminase</td>
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Although rare, the disease can present fatal complications. An early diagnosis is of utmost importance because, a revision of previous episodes suggestive of this disease. Indeed, in some cases, the diagnosis is only evident after second clinical episode it was hypothesized to be AOSD. and diagnose this disease. In this case, only after the differential diagnosis. Therefore, it is not easy to identify heterogeneity of clinical symptoms and has a broad of

CONCLUSION

Adult-onset Still’s disease may present with heterogeneity of clinical symptoms and has a broad of differential diagnosis. Therefore, it is not easy to identify and diagnose this disease. In this case, only after the second clinical episode it was hypothesized to be AOSD. Indeed, in some cases, the diagnosis is only evident after a revision of previous episodes suggestive of this disease. An early diagnosis is of utmost importance because, although rare, the disease can present fatal complications.

REFERENCES


Author Contributions

Ana Rita Fernandes Matos – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Elisabete Coelho – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.
Data Availability
All relevant data are within the paper and its Supporting Information files.

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