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Adult still's disease with atypical skin manifestations: A case report

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ABSTRACT

Adult Still's Disease (ASD) is a systemic inflammatory disorder of unknown etiology, thought to be triggered by infectious agents in genetically susceptible individuals, resulting in aberrant cytokine production including interleukin (IL) −1, IL-6, AND IL-18. The disease severity varies and may include macrophage activation syndrome (mas), a potentially life-threatening complication. It is associated with seronegative chronic polyarthritis, predominantly affecting young individuals. It is a rare condition with a complex diagnosis due to its overlapping clinical features with more common diseases.

Introduction

Adult Still's Disease (ASD) is characterized by a prolonged course of symptoms, including severe arthralgia affecting one or more joints, arthritis, high spiking fever (> 39°C) with a quotidian pattern, odynophagia, splenomegaly, lymphadenopathy, hepatic involvement, and a characteristic macular evanescent rash of salmon-pink hue, often pruritic, which affects the trunk, arms, legs and pressure areas, typically occurring during febrile episodes. Atypical cutaneous manifestations, including alopecia, urticaria, angioedema, acneiform lesions, and persistent generalized erythema, have also been reported.[1,2,3] Laboratory findings often reveal leukocytosis with neutrophilia, elevated erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP), along with hyperferritinemia (> 1000 ng/mL), particularly the non-glycosylated isoform. The immunological profile is typically negative for antinuclear antibodies (ANA) and rheumatoid factor (RF).[4] Yamaguchi's criteria (Table 1) are frequently employed for ASD classification, as they provide the highest sensitivity among available classification systems, despite limitations in specificity. ASD is a rare disease with an incidence of 0.16 cases per 100,000 inhabitants, as estimated in a French study,[5] fewer than a few hundred cases have been reported in the medical literature. Atypical clinical and cutaneous manifestations further underscore the importance of considering ASD in compatible presentations.

Case report

We report the case of a 26-year-old previously healthy female who presented to the Dermatology Department with sudden-onset urticarial, erythematous, and pruritic skin lesions accompanied by secondary excoriations. These lesions were localized to the periumbilical region, groin, axillae, and interdigitally. The patient also reported generalized myalgia and mechanical-type arthralgia. Initially diagnosed with scabies, she was treated with ivermectin, permethrin, and ketoprofen, without clinical improvement. Symptoms progressed over a three-week period, during which the patient developed fever and odynophagia, along with dactylitis in the second and third fingers of the left hand and arthritis in the proximal and distal interphalangeal joints, metacarpophalangeal joint of the second finger, and knees. Her skin lesions worsened, becoming widespread (Figs. 1 and 2) and accompanied by angioedema.

The patient was referred to Rheumatology, where an initial hypothesis of arthritis and reactive erythema to an infectious process was made, and antibiotic therapy was initiated without improvement. As the disease progressed, she was referred to Infectious Diseases, where scarlet fever was considered and treated, again without success. Laboratory investigations revealed leukocytosis with neutrophilia, elevated CRP and ESR, hyperferritinemia (> 2000 ng/mL), and negative blood cultures.

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Table 1
Yamaguchi Diagnostic Criteria (5 positive criteria required, including at least 2 major).

Major Criteria	Minor Criteria
Fever > 39°C for > 1 week	Odynophagia
Arthralgia/arthritis > 2 weeks	Lymphadenopathy
Evanescent rash	Altered aminotransferasis
Leukocytes > 10,000/μL	Rheumatoid factor (-)
Neutrophils > 80 %	Antinuclear antibodies (-)

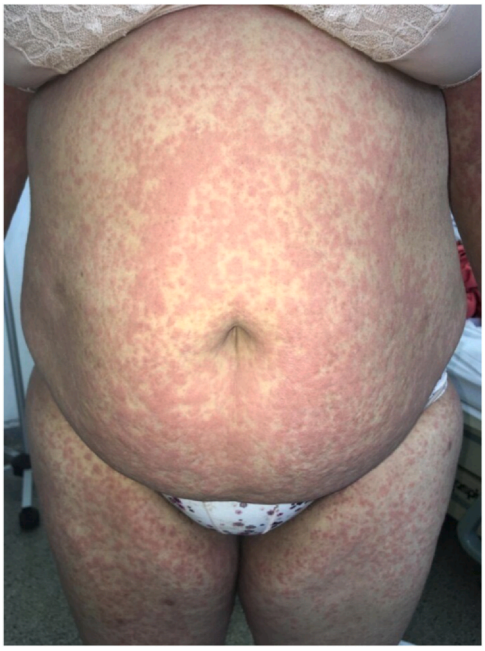

Fig. 1. Urticarial erythematous plaques distributed in the abdominal and inguinal regions.

Laboratory investigations revealed

- Leukocytosis (17,000/μl) with 85 % neutrophils
- Elevated CRP (148 mg/l)
- ESR(95 mm/hr)
- Hyperferritinemia (> 2000 ng/mL)
- Negative blood cultures
- Negative RF and ANA

Infectious causes were excluded.[6] The patient reported a similar but milder episode four months prior, which had improved with prednisone therapy. Based on clinical findings and fulfillment of Yamaguchi criteria, ASD was hypothesized, and the patient was referred back to Rheumatology. Immunological testing was negative for RF and ANA, and abdominal CT imaging revealed splenomegaly and lymphadenopathy.[7] Skin biopsy was not performed, as literature indicates its limited utility in ASD diagnosis.[8,9,10] Clinical presentation and laboratory results were deemed sufficient for diagnosis.

Treatment with prednisone was initiated, and methotrexate was added due to persistent symptoms. The patient showed clinical improvement. Methotrexate was chosen as a steroid-sparing agent based on evidence of efficacy in controlling arthritis and systemic symptoms in ASD.[11, 12]

Discussion

Understanding the cutaneous manifestations of ASD is essential for early diagnosis, which is critical for appropriate management and favorable patient outcomes, while also reducing unnecessary diagnostic and therapeutic costs. This case illustrates an atypical cutaneous presentation of ASD, where the patient exhibited widespread urticarial plaques and angioedema without febrile peaks.

A comparison with similar cases in the literature[13,14] shows that urticaria-like lesions and angioedema, though rare, may be present in ASD. This underscores the need to include asd in differential diagnosis even in absence of classical rash.

Although skin biopsy and immunofluorescence are not required for the diagnosis of asd, they may be performed in uncertain cases. In this instance, the typical clinical course, characteristic laboratory findings, and history of steroid responsiveness sufficiently supported the diagnosis.

A discussion on the Yamaguchi criteria is necessary, because although they are widely used to classify ASD, they are not specific for ASD and have limitations. They can be observed in other conditions, such as infections (for example, infectious mononucleosis, tuberculosis), neoplasms (such as lymphomas) and other rheumatological diseases (such as systemic lupus erythematosus and vasculitis).[11,12] However, its sensitivity and simplicity make it the preferred tool in clinical practice.[10] It is essential to perform a comprehensive evaluation to exclude these conditions before confirming the diagnosis of ASD.

The differential diagnosis should consider infectious, granulomatous, neoplastic diseases, and connective tissue disorders. Patients with atypical manifestations often undergo multiple specialty consultations before the correct diagnosis is reached. Thus, ASD should be considered in the presence of atypical or absent cutaneous manifestations, accompanied by compatible clinical and laboratory findings.[8, 9, 10, 13]

The choice of treatment with prednisone followed by methotrexate was based on clinical response and therapeutic patterns described in the literature. In refractory cases, biologic therapies including interleukin-1 (IL-1) (anakinra) and IL-6 (tocilizumab) inhibitors may be considered. These agents have been shown to be effective in controlling systemic inflammation and preventing relapses.[15, 16]

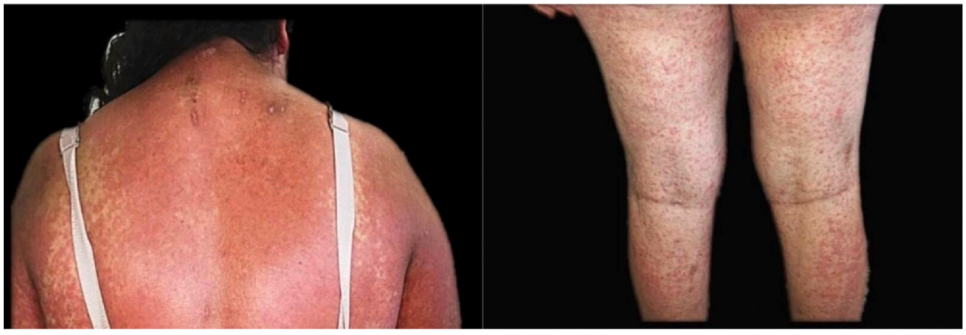

Fig. 2. Erythematous plaques and macules located on the back and posterior regions of the lower limbs.

Understanding the cutaneous presentation of ASD is crucial for early diagnosis, which is essential for proper management and favorable patient outcomes, as well as reducing unnecessary treatment and examination costs.

Conclusion

This case report demonstrates an atypical cutaneous presentation of ASD, with urticarial, erythematous, pruritic plaques not associated with fever peaks. ASD suspicion should be considered in the presence of atypical skin manifestations and compatible laboratory findings. Early recognition and appropriate treatment are essential to improving patient quality of life.

CRediT authorship contribution statement

Giovana Viotto Cagnon: Writing – review & editing, Writing – original draft, Methodology, Investigation, Formal analysis, Conceptualization. **Giovanna Beatrice de Souza:** Writing – original draft, Investigation, Conceptualization. **Elizandra Gomes Pereira:** Writing – original draft, Investigation, Formal analysis, Conceptualization. **Gabriela Roncada Haddad:** Writing – review & editing, Supervision, Project administration, Formal analysis, Conceptualization.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper. I certify that the work presented did not receive any financial support from the pharmaceutical industry or other commercial source other than those described below, and neither I nor any first-degree relative have financial interest in the subject covered in the manuscript.

Ethics Statement

All experiments and procedures were approved by the Botucatu Medical School and the Brazilian Ethics Committee. The publication of a single clinical case is permitted by the Ethics Committee of the Botucatu Medical School (CEP-FMB-UNESP) without the need for additional submission to Plataforma Brasil. All procedures used were ethically reviewed and approved by the Ethics Committee of the Botucatu Medical School (CEP-FMB-UNESP). The authors complied with the relevant ethical standards for publication and obtained informed consent from the patient.

Data availability

The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

Appendix A. Supporting information

Supplementary data associated with this article can be found in the online version at doi:10.1016/j.jdsct.2025.100088.

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