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CASE REPORT

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Flagellate dermatitis: An atypical skin finding in adult-onset Still's disease

Sana Toujani | **Amira El Ouni** | **Asma Belhassen** | **Kamel Bouslama**

Internal Medicine Department, University Hospital Center Mongi Slim, La Marsa, Tunisia

Correspondence
Sana Toujani, Internal Medicine Department, University Hospital Center Mongi Slim, La Marsa, Tunisia.
Email: sana.toujani@fmt.utm.tn

Abstract

A 23-year-old woman followed for adult-onset Still's disease (AOSD) presented fever and chest pain. Clinical examination showed erythematous papules suggestive of flagellate dermatitis. Laboratory findings showed increased Cardiac troponin. Myocarditis due to AOSD was therefore suspected. The patient was treated with prednisone and methotrexate with significant clinical improvement.

KEYWORDS

adult-onset Still's disease, flagellate dermatitis, myocarditis, prognosis

1 | INTRODUCTION

Flagellate dermatitis is a nonspecific inflammatory skin manifestation characterized by linear or curvilinear streaks and plaques, occurring mainly on the trunk. It is a rare skin finding that is most often caused by Bleomycin or the intake of shiitake mushrooms. The association with adult-onset Still's disease (AOSD) is rare. Patients with atypical skin manifestations of AOSD like flagellate dermatitis have more severe and persistent disease with systemic complications. We report in this regard a case of flagellate dermatitis associated with suspected myocarditis in AOSD.

2 | CASE REPORT

A 23-year-old woman without any relevant history was referred to our internal medicine department in Mongi Slim Hospital in Tunis with a 3-week history of fever, asthenia, and joint pain affecting her elbows, wrists, hands, knees, and ankles. She also had a history of odynophagia, loss of appetite, and asthenia. However, there was no photosensitivity, malar rash, muscle weakness, chest pain, or palpitations. The patient had no documented allergies or cardiovascular risk factors.

On physical examination, her weight was 62 kg; she had fever (39.4°C), tenderness, and mild swelling of wrists and interphalangeal joints of hands. There was diffuse macular erythematous eruption that appeared with the spikes of fever mainly on the neck, trunk, and upper limbs. Electrocardiogram was without abnormalities. Laboratory studies showed leukocytosis (30,350/μl, 95% neutrophils), anemia (9.3 g/dl), increased levels of C-reactive protein (286 mg/L), high erythrocyte sedimentation rate (150 mm), and a very high serum ferritin level (18,000 ng/ml). The results of liver function tests were within normal limits. Tests for antinuclear antibody, rheumatoid factor, and anti-cyclic citrullinated peptide antibody were negative. Parvovirus B19 immunoglobulin M (IgM) titers were normal. Tests for cytomegalovirus, human immunodeficiency virus, and hepatitis B or C virus were negative. Urine cytobacteriological examination was normal as well as Chest X-ray, abdominal ultrasound, and echocardiography.

The diagnosis of AOSD was considered in view of the clinical presentation: fever and polyarthritis in the absence

of evidence of infection, autoimmune disease, or malignancy. Her clinical and laboratory parameters fulfilled four major and two minor Yamaguchi's diagnostic criteria. The patient was treated with oral prednisone 60 mg once daily with resolution of all symptoms.

Two months later, while she was off treatment, the patient experienced the recurrence of fever, arthralgia associated with odynophagia and a 3-day history of persistent chest pain and palpitations. On physical examination, she had spiking fever (39.4°C), tenderness of wrists, hands, knees, and ankles without synovitis. Her blood pressure was 120/70 mmHg, and heart rate was 96 beats/min. Cardiopulmonary auscultation revealed no abnormalities. There was no organomegaly or lymphadenopathy. Skin examination showed macular erythematous eruption appearing with the spikes of fever associated with fixed linear erythematous streaks on the trunk, lower back, and stomach with a flagellated arrangement compatible with flagellate erythema (Figures 1 and 2). Electrocardiogram showed sinus rhythm, heart rate of 90 beats/min, and complete right bundle branch block.

Laboratory findings showed leukocytosis: 20,260/µl, neutrophils 19,900/µl, increased levels of C-reactive protein (480 mg/L), and thrombocytosis (Plq = 1,328,000/µL). High-sensitivity troponin I concentration was raised (849 nmol/ml). Electrocardiography and cardiac MRI were normal. Chest computed angio-tomography demonstrated only minimal bilateral pleural effusion, without pulmonary embolism. An infectious disease was ruled out. Coronary angiography was without abnormalities. Serology test results for parvovirus B19 immunoglobulin M, Epstein–Barr virus, and cytomegalovirus were negative. Myocarditis due to AOSD was suspected. The patient was treated with Bisoprolol, prednisone 1 mg/kg daily, and methotrexate 10 mg/week. The patient became afebrile with significant clinical improvement in rash and flagellate dermatitis, fever, and chest pain. There was also normalization of all biological parameters including cardiac troponin. After 4 years of follow-up, the progression was favorable despite tapering steroids dosage.

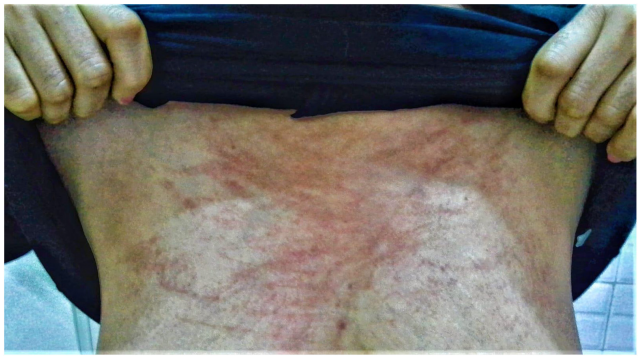

FIGURE 1 Fixed linear erythematous streaks on the trunk

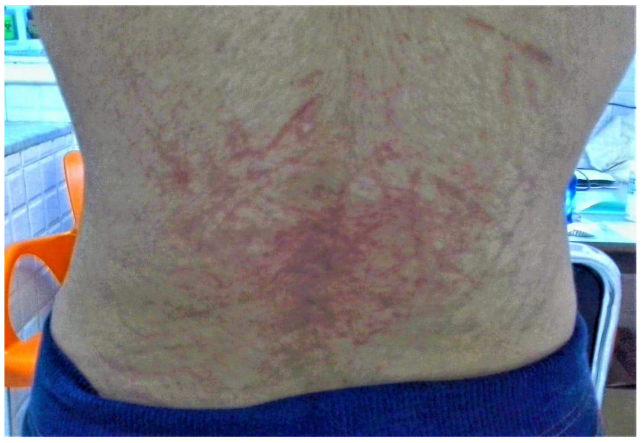

FIGURE 2 Erythematous papules grouped in lines with a flagellated arrangement on the lower back

3 | DISCUSSION

Adult-onset Still's disease is a systemic inflammatory disorder of unknown etiology and pathogenesis that usually affects young adults with an estimated prevalence of 1.5 cases per 100,000–1,000,000 people.[1]

Clinical manifestations include spiking fever, arthralgia or arthritis, serositis, transient cutaneous manifestations, lymphadenopathy, and hepatosplenomegaly. The main differential diagnoses are infections, neoplasms, and autoimmune disorders.[2]

The typical skin feature is an evanescent, non-pruritic salmon-pink maculopapular eruption that appears concomitantly with fever spikes and subsides when fever resolves. It occurs in 60%–80% of patients and is predominantly found on the proximal limbs and trunk with rare involvement of the face and distal limbs.[3]

In recent years, atypical cutaneous manifestations of AOSD have been reported often in addition to the typical evanescent rash in 14% of cases but may be the only skin manifestation.[4] Flagellate dermatoses are uncommon figurate dermatoses characterized by parallel linear or curvilinear arrangement simulating the marks of whiplashes. Persistent pruritic papules and plaques with a flagellate appearance resembling those in our patient are a well-described feature of chikungunya, bleomycin-intake, dermatomyositis, and shiitake mushroom.[5] It is a rare cutaneous feature in AOSD.

Other commonly reported cutaneous manifestations are non-pruritic persistent erythema, urticarial or lichenoid papules, pigmented plaques, and prurigo pigmentosa–like.[6,7] Atypical skin findings may appear at any time over the course of the disease but in the great majority of cases at the time of disease onset concurrently with systemic symptoms, or shortly afterward.[4]

It was reported that the majority of patients with atypical skin manifestations of AOSD had more severe and persistent disease. These patients developed complications like pericarditis, myocarditis, serositis, lung involvement, neurological involvement, and reactive hemophagocytic syndrome.[8] The same findings were observed in our case since flagellate dermatitis was associated with probable myocarditis. Cardiac involvement in AOSD usually manifests itself as a pericardial disease (up to 37% of cases) while myocarditis is uncommon with a prevalence of 7%.[9]

Patients with atypical skin manifestations have also a worse prognosis with a mortality rate of around 8%.[10] Most of the patients with atypical manifestations of ASOD require moderate to higher doses of glucocorticoids. Around 40% of these patients require immunosuppressant therapy like methotrexate, azathioprine, cyclosporin, and hydroxychloroquine.[11] Our patient was treated with prednisone and methotrexate because of suspected myocarditis with significant clinical improvement of chest pain and flagellate dermatosis.

4 | CONCLUSION

Our case illustrates a rare presentation of AOSD with flagellate dermatitis. Atypical skin findings such as flagellate dermatosis have been reported in AOSD and may be associated like other atypical skin manifestations with systemic complications such as myocarditis and worse prognosis. Awareness of atypical skin findings in AOSD may lead to earlier diagnosis of systemic complications and better management.

AUTHOR CONTRIBUTIONS

Sana Toujani wrote the manuscript with support of Asma Belhassen. Amira El Ouni and Kamel Bouslama reviewed and edited the manuscript.

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CONFLICT OF INTEREST

None.

DATA AVAILABILITY STATEMENT

None.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

Sana Toujani https://orcid.org/0000-0003-4943-3416

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