



Persistent Pruritic Plaque Like Rash in Adult-Onset Still's Disease: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Authors MAA and AS encountered the case and wrote the first draft of the manuscript. Author MA managed the literature searches. All authors read and approved the final manuscript.

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Case Study

ABSTRACT

Adult-onset Still's disease (AOSD) is a multi-system, inflammatory disease that presents with acute fevers, arthritis or arthralgia, and a evanescent maculopapular rash, mostly occurring with myalgia, sore throat, splenomegaly, lymphadenopathy, raised C-reactive protein (CRP), serum ferritin, and neutrophilic leukocytosis. This is a case report of a 51-year-old female who came with pruritic plaques on her neck, chest, shoulders, upper arms, thighs, and lower back along with fever and arthritis. Her initial lab work showed borderline low C4, positive antinuclear antibody (ANA), and a CRP of 14 mg/dl (that was later increased to 281 mg/dl), microscopic hematuria with +1 protein on urinalysis with a creatinine of 1.3 mg/dl, WBC of 23.5x10⁹/L mildly elevated liver function tests, brain natriuretic peptide (BNP) of 480 mg/dl, and high troponin of 0.130. In this case, we are highlighting the atypical presentation of rash in Adult-onset Still's disease.

Keywords: Pruritic plaque rash; arthritis; fever.

1. INTRODUCTION

Adult-onset Still's disease (AOSD) is a multi-system, inflammatory disease that presents with

acute fevers, arthritis or arthralgia, and a maculopapular rash, mostly occurring with myalgia, sore throat, splenomegaly, lymphadenopathy, raised CRP, serum ferritin

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and neutrophilic leukocytosis [1]. Usually, Yamaguchi et al. criterion is used. According to Yamaguchi criteria, major criteria includes spiking fever $\geq 39^{\circ}\text{C}$, arthralgia/arthritis lasting two weeks or more, typical (evanescent) maculopapular rash, and leukocytosis $>10 \times 10^9/\text{L}$. Minor criteria include sore throat, recent lymphadenopathy development, hepatic or splenomegaly, abnormal liver function, negative anti-nuclear antibody, and rheumatoid factor [2]. A minimum of two major criteria is needed for diagnosis. The treatment of AOSD involves NSAIDs, corticosteroids, steroid-sparing agents such as methotrexate, and other biological agents. The treatment of AOSD remains life long, and prognosis tends to be variable [3]. The objective of this case report is to present a fairly rare presentation of rash in AOSD.

2. CASE DISCUSSION

We present a case of Adult-onset Still's disease in a 51-year-old female who came to the University of Alberta Hospital, Edmonton, Canada. She presented with a history of pruritic rash on her chest and neck region. She was given topical steroids, which provided minimal improvement. Her rash progressed down to her shoulders, upper arms, thighs, and lower back. Her initial lab work showed ferritin of 2101 ng/ml, borderline low C4, positive antinuclear antibody (ANA), negative extractable nuclear antigen (ENA), and a C-reactive protein (CRP) of 14 mg/dl that was later increased to 281 mg/dl. One month later, she developed bilateral knee pain followed by bilateral wrist and ankle pain with some intermittent swelling in bilateral wrists and knees. She initially controlled her pain with Ibuprofen, but for the last few days, she was unable to ambulate. She also had a fever of 38°C , which was on and off along with chills. She also had an unintentional weight loss of about 30 pounds in the last two months. She denies any visual changes, nausea, vomiting, diarrhea, abdominal pain, and dyspnea. She reported orthopnea and paroxysmal nocturnal dyspnea for the last few days. She also stated that she noticed tea-colored urine, but denies any increased urinary frequency, dysuria, or hemoptysis. She was started on prednisolone and doxycycline, which improved her joint pain and resolved her tea-colored urine. On examination, she was found to be alert and oriented. Cardiovascular examination showed a 2/6 systolic ejection murmur at the upper sternal border, with no extra heart sounds. Pulmonary examination showed equal air entry bilaterally,

fine crackles to bilateral bases with no wheezes. Abdominal examination showed no hepatosplenomegaly; the abdomen was found to be soft, not tender, and not distended. Musculoskeletal examination showed bilateral small effusions to knees and ankles with a painful range of motion. Skin examination showed red/brown maculopapular rashes on back, arms, chest, with some eczematous plaques (Fig. 1).



Fig. 1. Maculopapular pruritic plaque like rash on left arm

Differential diagnosis included autoimmune disease (Still's disease, vasculitis, unlikely SLE), infection (EBV/CMV/?TB), or malignancy (lymphoma). Dermatology took a punch biopsy of the rash from the left upper arm and mid-back. Differential diagnosis of subacute/chronic dermatitis versus lichen simplex chronicus was considered and a trial of betamethasone valuate 0.1% ointment to the affected regions was started. The patient was discharged from the hospital after a week on a tapering dose of prednisone, and naproxen 500 mg, twice daily.

Two months after discharge, the patient presented with recurrence of her rash to the chest and neck along with fatigue, arthralgia of wrist, knees, and ankles. She also had new proteinuria with an acute kidney injury (AKI). She endorsed 50-pound weight loss over the past three months. The patient initially responded to prednisone and NSAIDs but flared after tapering off prednisone. Examination showed tender right and left wrist, tender right knee, and bilateral ankles. Mild soft tissue swelling was noted around the ankles. The right knee had some peri-

articular swelling. An eczematous rash was over the trapezius and around the clavicles. Rheumatology admitted this patient for a work-up of worsening Still's disease. The patient was restarted on prednisone 40 mg, daily along with colchicine 0.6 mg, three times daily. Colchicine discontinued later due to diarrhea. The patient was started on methotrexate 20 mg, subcutaneously, once a week. The patient was discharged from the hospital a week later with a two-month prednisone tapering dose while waiting for methotrexate to take effect.

3. DISCUSSION

There are few cases of AOSD reported in past with rare presentations, but they had different rarity. As in one case a 46-year-old male Indian patient who had intermittent high-grade fever, evanescent rash and polyarthritis for one month [4]. In another case, a 45-year-old male presented with multiple joint pains associated with unresolved fever associated with severe sore throat for the past two weeks. This was preceded by an evanescent, non-pruritic macular rash mainly over the trunk and extremities [5]. There is a case of a 24-year-old female who presented with sore throat, fever, raised liver enzymes and cervical lymphadenopathy with developed evanescent skin rash and polyarthritis having AOSD [6]. In a 61-year-old woman who had a history of flu-like illness for a month along with laboratory studies (elevated LFTS, ferritin and inflammatory markers), polyarthralgia, mouth ulcers and maculopapular rash which lead to diagnosis of AOSD [7]. There is a case of a young female with fever on unknown origin for two months with systemic symptoms (weakness, polyarthralgia) that led to diagnosis of AOSD based on lab investigations and treatment response [8]. There has been a case reported recently in which a AOSD was diagnosed after three years of initial presentation with classic symptoms (fever, myalgia, rash) [9]. Complications from delayed diagnosis of AOSD include: fulminant hepatitis, myocarditis, acute respiratory distress syndrome, serositis, macrophage activation syndrome, disseminated intravascular coagulopathy, septic shock, pulmonary artery hypertension, diffuse alveolar hemorrhage, and thrombotic thrombocytopenia purpura [10].

In the afore-mentioned cases it is seen that evanescent rash is presenting with fever and arthralgia/arthritis, unlike in our case where the rash is pruritic and persisted beyond fever. By

presenting this case we want to highlight a dermatologic variation of AOSD which should be kept in mind in future. We hope this assist in early diagnosis and better recovery of similar patients in future. Early diagnosis and treatment may lead to a positive prognosis and avoidance of serious complications.

4. CONCLUSION

Like in other studies, our patient with Adult-onset Still's disease also presented with a typical triad of fever, rash, arthralgia, along with leukocytosis and increased ferritin. In majority cases, Still's disease was preceded by sore throat, unlike in our case. In our case the rash was not typical (evanescent) maculopapular, but it was persistent pruritic plaque like at usual presentation sites. This dermatology variation is an important aspect to keep in mind when considering AOSD in future.

CONSENT

As per international standard or university standard, participant's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard, written approval of Ethics committee has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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