CASE REPORT

Flagellate Dermatosis -An Unusual Dermatological Finding in a Case of Systemic Lupus Erythematosus

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Abstract

A 28-year-old female presented with a history of 'on and off' low-grade fever for last 2 months, asymmetrical small and large joint involvement with oral ulcers and flagellate dermatosis and pigmentation over arms. Peripheral smear revealed anaemia and leukopenia and urine examination revealed proteinuria and haematuria. ANA profile was positive for anti-ds DNA, anti-Histone, and anti-Nucleosome antibody suggestive of active Systemic lupus erythematosus (SLE). Patient was treated with injectable steroids following which the rashes and symptoms subsided. We report here probably the second case report of flagellate dermatosis as one of the rare initial dermatological manifestations of SLE.

Keywords

Flagellate Dermatoses, SLE, Flagellate Erythema

Introduction

Flagellate dermatoses are unusual linear dermatoses distinguished by parallel, longitudinal, or curved formations resembling whiplash marks, first coined to describe bleomycin-associated dermatitis. [1] A few diseases and conditions like intake of shiitake mushrooms, chemotherapeutic drugs like docetaxel and bendamustine, hypereosinophilic syndrome, chikungunya fever, and phytophotodermatitis have been variably associated with this pattern of pigmentation. [2] Rheumatological conditions like dermatomyositis, systemic sclerosis and adult-onset Still's disease have been occasionally described with this pattern of erythema and pigmentation. Flagellate dermatoses in Systemic Lupus Erythematosus (SLE) have been described only once previously in the available literature to the best of our knowledge. [3] Recognition of such non-pruritic linear streaks with erythema in a setting of on-and-off fever with joint pain would make clinicians aware of the possible association with a rheumatological condition like SLE.

Case Report

A 28-year young female presented to the outpatient department with complaints of multiple, asymmetrical, non-specific joint pain, low-grade on-and-off fever for the last 2 months and oral ulcer for the last 10 days. She initially developed mild left first metacarpal joint pain followed by pain and tenderness of left wrist, elbow, knee, and ankle joints which were insidious in onset, dull aching type, associated with myalgia, non-radiating, exacerbated by movement and relieved with over-the-counter non-steroidal anti-inflammatory drugs (NSAIDs). This pain was often associated with low-grade intermittent fever (maximum recorded temperature - 101° F), not associated

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Manuscript Received: 18.08.2022; **Revision Accepted**: 1.11.2022;
Published Online First: 10 July 2023
Open Access at: https://journal.jkscience.org

Cite this article as: Dalai SP, Krishna AV, Das C, Sahu S. Flagellate Dermatosis -An Unusual Dermatological Finding in a Case of Systemic Lupus Erythematosus. JK Science 2023;25(3):191-93.

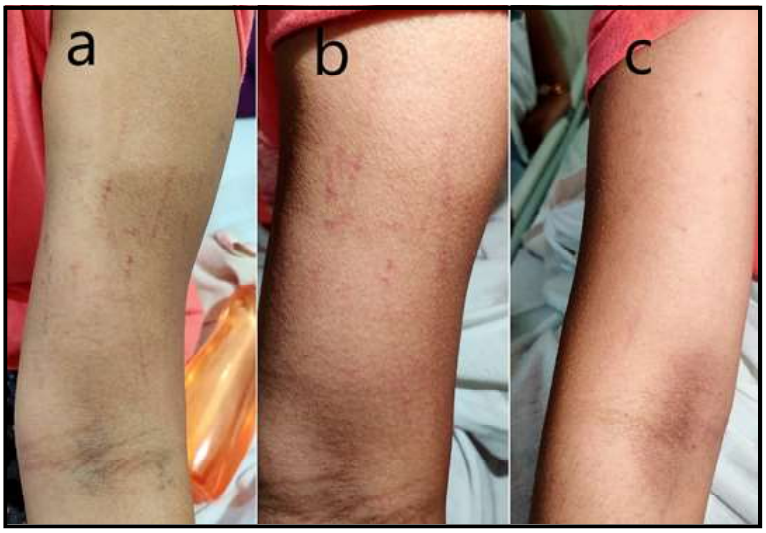

Fig 1:. Flagellate dermatosis over left arm at a. on day 1 (at presentation), b. on day 5 (after 3 days on steroids), c. on day 9 (after 7 days on steroids)

with chills or rigor, without any diurnal variation and subsided with or without acetaminophen. There was no history of weight loss, cough, or abdominal pain during the illness. She developed oral ulcers for the last 10 days before admission, which were associated with pain and difficulty taking food. On the day of hospitalisation, she noticed non-pruritic horizontal, parallel linear streaks with mild erythema over her left arm, above the left cubital fossa (*Fig 1*). There was no dermatographia. She was a known case of primary hypothyroidism and had been taking thyroxine 75 micrograms for the last 5 years. There was no significant past or family history of rheumatological diseases, malignancy, and tuberculosis. She did not give any history of recent intake of shiitake mushrooms, jellyfish stings, insect bites, trauma, torture, or abuse. There was no malar rash or photosensitivity, or alopecia. The per-abdominal examination did not reveal hepatosplenomegaly. Cardiovascular and respiratory system examinations were unremarkable.

Investigations:

A complete blood count (CBC) revealed anaemia and leukopenia with a Haemoglobin (Hb) of 8.0 gm/dl and a total leucocyte count (TLC) of 3670 cells/µl. Liver enzymes were mildly raised with SGOT 107 IU/L and SGPT 98.2 IU/L. Erythrocyte sedimentation rate (ESR) was 60mm/1hr, and C-reactive protein (CRP) was 0.63. Renal Function tests (RFT), lipid profile and electrolyte parameters were in the normal range. Serum lactate dehydrogenase (LDH) was mildly elevated at 338 U/L, and serum ferritin levels were significantly raised to 748.5 ng/ml. Urine microscopy revealed 2+ proteinuria. Thyroid profile showed mildly elevated TSH with levels of 5.48 µIU/ml. Prothrombin time (PT) was 10.1 sec, and PT-INR was 0.95. Blood panels for tropical infectious diseases like MP-QBC for malaria, Ig M salmonella, Ig M scrub typhus, and Ig M Chikungunya were negative. CT scan of abdomen revealed hepatomegaly. Blood and urine culture samples were unremarkable. X-ray of left hand and wrist was normal without any evident erosion or deformity. Repeat ferritin and LDH were not in the rising trend, with values of 782.7 ng/ml and 369 U/L, respectively. Serum Creatinine phosphokinase (CPK) levels were 97 U/L which was in the normal range.

Considering the clinical picture and blood parameters, the possibility of an autoimmune disease was suspected, and an Anti-nuclear antibody (ANA) titre and ANA profile were ordered. ANA titre was (4+) positive and homogenous pattern, and profile showed significant titres of anti-dsDNA, anti-Nucleosomes (NUC), and anti-Histone (HI) antibody positivity. Anti Jo-1 and other antibodies were negative. Serum complement levels C3c and C4c were significantly decreased with values of 0.26 g/L and 0.08 g/L, respectively. With the above lab investigations, a diagnosis of "definite SLE" was made according to both ACR and SLICC criteria. [4,5]

Oral prednisolone 40 mg/day was started and continued. Hydroxychloroquine 300 mg once daily was also initiated. The flagellate dermatoses and erythema decreased slowly

and vanished completely over the next 7 days. The fever and joint pain also subsided over the period of 10 days, and the patient was symptomatically better, hence discharged with advice to follow up after 1 month.

Discussion

Flagellate erythema is a rare, patterned rash that appears in linear, parallel streaks and is classically described after chemotherapy with bleomycin or docetaxel. They may start as a diffuse, erythematous pruritic plaques or flagellate lesions over the trunk and extremities generally described as scratch dermatitis. The rash usually heals with persistent flagellate brownish hyperpigmentation. [1] Such pigmentation is usually not associated with itching or pruritus. They usually clear after months of discontinuation, and no approved treatment exists. Flagellate dermatitis has also been described after 48 hours of consumption of raw or undercooked Shiitake mushrooms (Lentinus Edodes) where they usually appear as small, extremely pruritic papules with erythematous lesions and are referred as toxicoderma. [6] Flagellate lesions can also be triggered by Phyto-photodermatitis and poison ivy dermatitis where the leaves have rubbed against the skin. [7] Hypereosinophilic syndrome associated with HIV infection have been reported with development of flagellate dermatitis. [8] Cases of Chikungunya fever have been reported with flagellate patterns of pigmentation associated with melanin retention. [9] Sadomasochistic behaviour or abuse can also give rise to whiplash marks mimicking flagellate dermatitis. [10] Rheumatological disorders like Dermatomyositis [11], Systemic sclerosis [12], and adult-onset Still's disease [13] have been described rarely to be associated with centripetal flagellate erythema, linear streaks, and striped erythema. Such lesions are described as erythematous, pruritic lesions associated with scratching with minimal pigmentation. Histopathological examination shows mild perivascular inflammation with mononuclear cells, neutrophils, and perinuclear vacuolisation of the arrector pili muscle. [14] The rash usually disappears with corticosteroid therapy. Only 1 case of flagellate dermatosis with SLE has previously been reported from Japan. [3] The early appearance of such non-pruritic linear streaks of erythema and pigmentation in a setting of joint pain, unremitting fever should alert the physician about probable rheumatological disease. A careful history taking, and intelligent investigation might help in narrowing down the differential diagnosis.

Conclusion

The presence of such linear streaks and flagellate dermatosis, along with joint pain and recurrent fever with or without rashes, should alert the physician about the possibility of SLE or other associated rheumatological diseases and should prompt the treating physician for appropriate investigations to reach the correct diagnosis and initiate proper treatment at the earliest.

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