# Flagellate Rash in Adult-onset Still's Disease

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Sir,

A 34-year-old female was in care of an internist for high-grade fever with chills, generalized body ache, sore throat, and pain in multiple joints for 1 week. After 3 days she developed a rash over her skin for which she was referred to us. On examination, she was febrile (103 °F), wrist and ankle joints were extremely tender with minimal swelling. A few cervical lymph nodes were enlarged. Skin examination showed lichenoid to hyperpigmented, mildly pruritic plaques over the limbs and trunk in a flagellate pattern [Figures 1 and 2]. There was no mucosal involvement.

Her investigations showed a TLC count of 22,400/mm$^3$with 86.1% neutrophils and an increase in erythrocyte sedimentation rate (71 mm/h) and CRP (273.70 mg/L). *S. ferritin* level was very high (>10,000 ng/ml). She was tested negative for rheumatoid arthritis factor, antinuclear antibodies, as well as p-ANCA and c-ANCA. Liver function tests and renal function tests were normal. Chest X-ray and abdominal ultrasonography were normal. ELISA test for HIV was negative too. No microorganisms grew on throat swab culture. No evidence of any infection such as malaria/dengue/typhoid was found on blood test. She was empirically treated by the internist with doxycycline and anti-malarials (quinine and primaquine), ranitidine and ebastine but no improvement was seen. None of the drugs which she was taking, have been reported in the literature to cause flagellate rash [Table 1].[1] This prompted us to think of adult onset Still's disease (AOSD) as the diagnosis. Patient also fulfiled Yamagushi's criteria for the diagnosis.[2]

Skin biopsy showed mild ortho-keratosis, acanthosis with focal basal call vacuolation [Figure 3] and numerous eosinophils in epidermis and papillary dermis [Figure 4]. She was then referred to

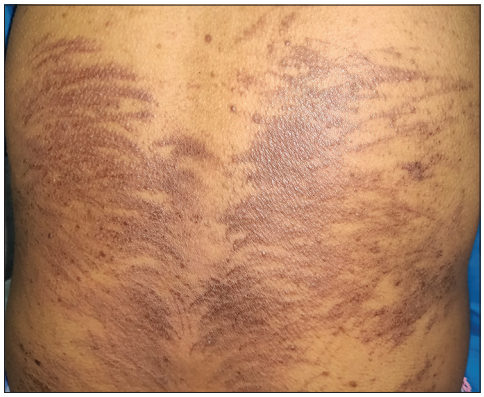

**Figure 1: Symmetrical lichenoid to hyperpigmented flagellate rash over back**

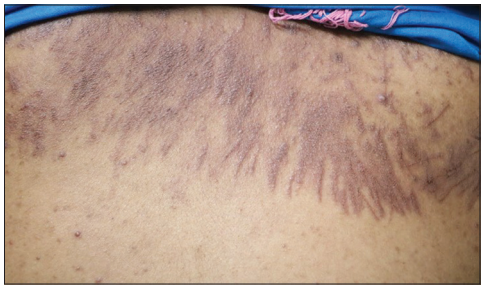

**Figure 2: Flagellate rash extending from chest to abdomen**

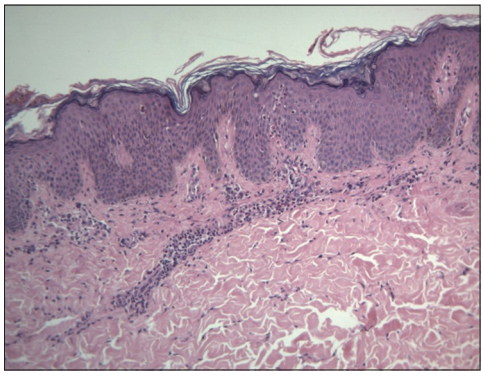

**Figure 3: Skin biopsy showing mild ortho-keratosis and acanthosis with focal basal call vacuolation and exocytosis of scattered eosinophils. The dermis exhibits mild to moderate peri-vascular inflammation [H&E × 100]**

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**Table 1: Causes of Flagellate dermatoses**

| |
|---|
| True Flagellation/mechanical |
| Religious punishment |
| Torture |
| Sexual pleasure (masochism) |
| Child/partner abuse |
| Dermatitis artefacta |
| Chemotherapy induced Flagellate dermatitis and pigmentation |
| Bleomycin |
| Peplomycin |
| Docetaxal |
| Bendamustin |
| Rheumatological disorders |
| Dermatomyositis |
| Adult-onset Still's Disease |
| Toxin-induced |
| Shiitake mushroom ingestion (raw or undercooked) |
| Cnidarian (Portuguese man-of-war and jellyfish) stings |
| Paederus and other insects |
| Other pruritic dermatoses |
| Dermographism |
| Excoriation by pruritic conditions |
| Phytophotodermatitis |
| Poison ivy dermatitis |
| Hypereosinophilic syndrome |
| Chikungunya induced flagellate pigmentation |
| Idiopathic Flagellate pigmentation |

rheumatologist who concurred with the diagnosis of AOSD after excluding infectious, malignant or autoimmune causes. She was treated with high-dose steroids after which her fever subsided, general conditions improved and the rash resolved. She was discharged after a week on oral steroids.

AOSD is a multisystem disorder of unknown etiology characterized by high spiking fever, typical evanescent maculo-papular skin rash, arthralgias, neutrophilic leukocytosis, negative rheumatoid factor, and antinuclear antibodies and marked hyperferritinemia.[3] The diagnosis requires exclusion of infectious, malignant and connective tissue diseases. In recent years, atypical cutaneous manifestations are increasingly being reported.[4,5] In our case, atypical lesions were present in the form of linear persistent pruritic plaques in a flagellate pattern which has been described previously in other case reports also.[4,5]

AOSD is categorised as a multigenic autoinflammatory disease.[3] It is defined a disorder at the "crossroads" of autoinflammatory and autoimmune diseases, because of the involvement of both arms of immune system, innate and adaptive ones.[3] The presence of eosinophils and absence of dyskeratotic cells on histopathology in our case differs somewhat from previously described cases.[4,5] As more cases are being reported, the clinicopathological spectrum of Still's disease will continue to be expanded and redefined.[6] We are reporting this case as accumulation of such cases are needed to sensitize the scientific community about the atypical skin rashes in AOSD. Recognition of such dermatological signs can also aid in the diagnosis of this uncommon entity.

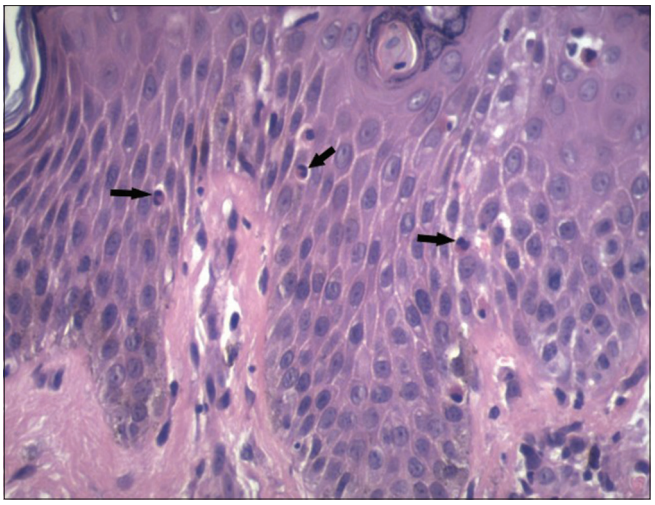

**Figure 4: High power view showing exocytosis of eosinophils [H&E 400x]**

## *Declaration of patient consent*

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## *Conflicts of interest*

There are no conflicts of interest.

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