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IP Indian Journal of Clinical and Experimental Dermatology

Journal homepage: www.ijced.org/

Case Report

# Skin lesions as the dominant clinical sign in hyper eosinophilic syndrome: A case report

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ARTICLE INFO

*Article history:*
Received 25-06-2023
Accepted 09-09-2023
Available online 17-10-2023

*Keywords:*
Hyper eosinophilic syndrome
HES
eosinophilia

ABSTRACT

Hyper eosinophilic syndrome (HES) is a leukoproliferative disorder with persistently elevated eosinophils (>1500/$\mu$L) for more than six months. Most commonly affect skin, lungs, intestine, heart, kidneys, eyes, and peripheral nervous system. Dermatological manifestation is one of the significant clinical sign indicating the disease activity in HES and may be the only manifestation of hypereosinophilic syndrome.Therefore, early identification of the cutaneous lesions may help the clinicians to approach a definitive diagnosis and treatment.



## 1. Introduction

Hardy and Anderson described hyper eosinophilic syndrome (HES) in 1965.[1] Idiopathic hyper eosinophilic syndrome is a rare disorder marked by elevated eosinophil count without a clear etiology. Common skin lesions associated with this disorder are itchy swollen erythematous papules, plaques and nodules that are usually located over the extremities. Extensive diagnostic workup is essential to rule out other causes of eosinophilia and to eliminate any multiorgan involvement. Majority of the patients respond to immunosuppressive agents but a regular follow up is needed to achieve a complete clinical remission and to detect early lymphoma.

This case report is about a patient who presented to our clinic with multiple symptomatic cutaneous lesions with no other systemic illness who was eventually diagnosed with idiopathic hyper eosinophilic syndrome based on her clinical presentation, extensive diagnostic work-up after excluding all known basic causes of hyper eosinophilia like infections, malignancy, hematological disorders etc.

## 2. Case Report

A 54-year-old female, who is a known case of type 2 diabetes mellitus presented with bilateral upper and lower limb swelling with associated intense pruritus at the same site for 8 months. She also noticed itchy skin lesions restricted to the exposed extremities since 3 months.On general examination vitals were stable, bilateral upper and lower limb showed pitting edema as well as local rise of temperature and redness. On cutaneous examination, multiple, discrete hyperpigmented papules and plaques with lichenification noted over bilateral dorsum of upper and lower limb with few lesions showing erosions as well as some of the lesions healed with depigmentation. Initial blood investigations showed eosinophilic leukocytosis without any atypical cells in peripheral smear, Absolute eosinophil count 3200/dl, mildly raised inflammatory markers, elevated serum lactate dehydrogenase and serum IgE level was 96 IU/ml. Skin Biopsy done from a papule revealed acanthosis, spongiosis with dermal edema

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https://doi.org/10.18231/j.ijced.2023.032

and eosinophilic infiltrate. Bone marrow biopsy was done which showed normocellular marrow with trilineage maturation with increased eosinophils and megakaryocytes. Lymphoscintigraphy revealed early lymphatic dysfunction of bilateral upper limb with normal lymphatic flow in bilateral lower limb. Doppler was done to rule out DVT. Blood culture and sensitivity-No growth. Other systemic examinations and investigations (ECG, ECHO, CT chest, Antinuclear antibody, rheumatoid factor, Urine and stool analysis, Infectious workup, liver enzymes, renal function test) were within normal limit. The patient was not willing to undergo genetic analysis.

Thus, with the above clinical evaluation we came to the diagnosis of hyper eosinophilic syndrome with skin lesions as the only indicator and started our patient on oral steroids and antihistamines as well as topical emollients and steroids with significant improvement of skin lesions and swelling of upper and lower limb with in a period of 2 weeks. However, this patient was lost to follow-up.

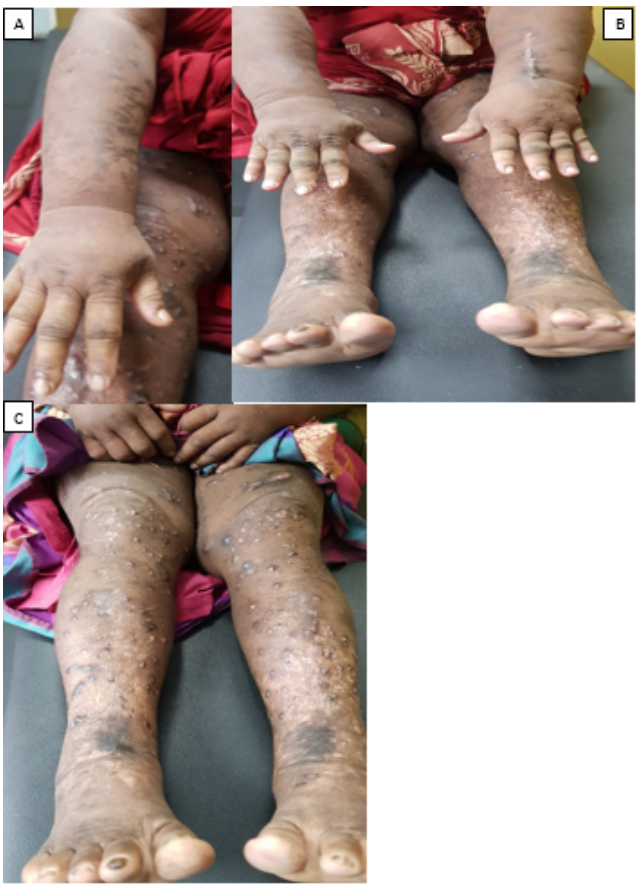


**Fig. 1:** **A** & **B:** Bilateral upper and lower limb swelling; **C:** Hyperpigmented papules and plaques with lichenification over bilateral lowerlimb

## 3. Discussion

Hyper eosinophilic syndrome is rare and lethal disorder which sometimes presents with cutaneous manifestations as the only indicator of the disease.[2]

It is defined by elevated eosinophilic counts >1.5×109 /l for more than 6 months devoid of any basic etiology and presence of any symptoms related to organ dysfunction which may be attributable to infiltration of eosinophils.[3] It is often seen among male patients in the age group of 20 to 50 years.[4]

Always other causes related to elevated eosinophils should be ruled out before concluding with a diagnosis of HES, such as helminthic parasitic infestation, atopic disease, drug allergy and so on. Certain common skin diseases associated with elevated blood eosinophilia consist of atopic eczema, bullous pemphigoid, cutaneous t cell lymphoma, eosinophilic cellulitis, panniculitis, vasculitis and many more.[3,5]

HES can affect any organs like heart, vascular system, skin, central and peripheral nervous system, gastrointestinal tract, and eyes.[6] The various mucocutaneous manifestation related to HES include vesicles, petechiae, papules and nodules, angio-oedema, livedo reticularis, erythematous lesions, necrosis, gangrene, Raynaud's phenomenon, eosinophilic cellulitis, and vasculitis, urticaria, mucosal ulcerations and pruritus.[7]

The histopathology of skin lesions is mostly non-specific with variable eosinophilic infiltration in HES.

It is hypothesized that an elevated IL-5 as well as involvement of myeloid cells with interstitial deletion of chromosomes on band 4q12 which may lead to the formation of a FIP1L1-PDGFRA fusion gene may be responsible for eosinophilia.[8]

Majority of the patients respond very well to high dose oral corticosteroid therapy with significant clearance of general malaise, skin changes as well as blood eosinophil levels. Other treatments suggested include PUVA therapy, Hydroxy urea and recently imatinib mesylate is also tried. Patients who are not symptomatic need no treatment usually.[9]

To conclude, clinicians should be aware of the rare possibility of hyper eosinophilic syndrome in which the dominating symptoms may be various mucocutaneous manifestations[2,10] and any persistently elevated blood eosinophilia for a chronic period should be investigated.

## 4. Conflict of Interest

Author has no conflict of interest to declare.

## 5. Source of Funding

No funding source.

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**Cite this article:** Kallan F, Pillai RT. Skin lesions as the dominant clinical sign in hyper eosinophilic syndrome: A case report. *IP Indian J Clin Exp Dermatol* 2023;9(3):166-168.