**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Dermatology**

# Adult Onset Urticaria Pigmentosa without Systemic Involvement: A Case Report

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**DOI:** https://doi.org/10.36347/sjmcr.2025.v13i04.027 | **Received:** 12.03.2025 | **Accepted:** 19.04.2025 | **Published:** 22.04.2025

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**Abstract** | **Case Report**

Cutaneous mastocytosis is a rare condition characterized by abnormal accumulation of mast cells in the skin. The most common clinical presentation, urticaria pigmentosa, manifests as itchy, brownish macules or plaques predominantly on the trunk and proximal extremities. In adults, it is generally associated with systemic involvement, making isolated cutaneous presentations highly unusual. We report a rare case of isolated urticaria pigmentosa in a 28 year-old adult male, presenting without systemic involvement confirmed through exhaustive investigations. Treatment with antihistamines and topical corticosteroids resulted in rapid clinical improvement. This case highlights the necessity for comprehensive evaluation to exclude systemic mastocytosis in adults.

**Keywords:** Cutaneous mastocytosis, Urticaria pigmentosa, Systemic involvement, CD117, Mast cells.



## INTRODUCTION

Cutaneous mastocytosis is a rare disorder defined by abnormal mast cell accumulation in the skin. Urticaria pigmentosa, the most prevalent clinical form, presents as itchy, brownish macules or plaques, predominantly affecting the trunk and proximal limbs. In adults, this presentation commonly indicates systemic involvement affecting the bone marrow or other internal organs. However, isolated skin involvement without systemic disease, as presented in our case, remains exceptional.

## CASE REPORT

A 28-year-old male presented with skin lesions evolving over 18 months, initially localized on the back and progressively extending to the trunk and proximal limbs. He experienced severe pruritus and intermittent flushing episodes. Clinical examination revealed confluent erythematous and brownish plaques with polycyclic borders, a negative Darier sign with negative dermatographism. Dermoscopy showed a brownish background with a pigmented network (**Figure 1**).

Histopathological analysis, supported by immunohistochemical staining, confirmed a diagnosis of urticaria pigmentosa by demonstrating significant dermal mast cell infiltration with positive CD117 expression. Biological and imaging investigations to detect systemic involvement revealed normal serum tryptase levels (5.9 ng/ml) and no hematological, osseous, or visceral abnormalities.

The patient was successfully managed with H1-antihistamines and topical corticosteroids, achieving significant clinical improvement and reduced pruritus within two weeks (**Figure 2**).

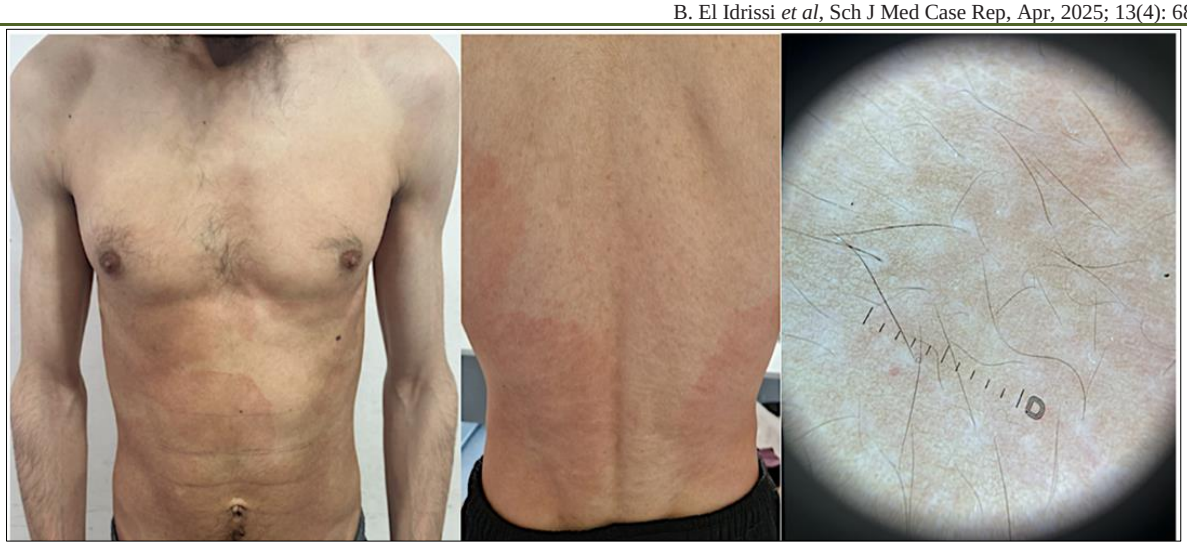

**Figure 1: Pigmented erythematous plaques on the trunk and back. Dermoscopic image showing a pigmented network and erythematous background**

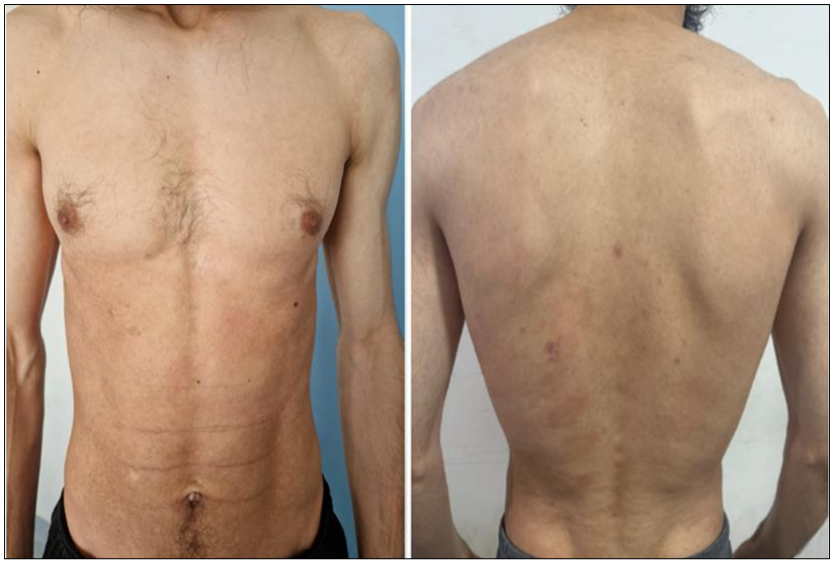

**Figure 2: Significant improvement after 2 weeks of treatment**

## DISCUSSION

Cutaneous mastocytosis in the form of pigmented urticaria is a common type of mastocytosis, but it is rarely isolated in adults, where it is often associated with systemic involvement [1]. The case presented is therefore unusual, highlighting the importance of a thorough assessment to rule out extra-cutaneous involvement.

The diagnosis is based on characteristic clinical elements, such as infiltrated brownish plaques and the Darier sign. Although the latter was negative, histopathology and immunohistochemistry confirmed the presence of a significant mast cell infiltrate with CD117 expression, a key criterion in diagnosing this condition [2]. The absence of elevated serum tryptase and signs of systemic involvement in imaging and biological assessments helped confirm that the condition was strictly cutaneous [3].

The treatment consisted of H1 antihistamines and topical corticosteroids, leading to a rapid improvement in lesions and itching. This approach is the standard treatment for skin forms, although more refractory cases may require alternatives such as leukotriene inhibitors or immunomodulatory therapies [4]. Finally, patient education remains essential to avoid common triggers, such as temperature fluctuations or histamine-releasing foods and medications [5].

## CONCLUSION

This report highlights a rare isolated case of cutaneous mastocytosis in an adult, underscoring the importance of long-term follow-up to monitor potential progression to systemic involvement.

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