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Adolescent-Onset Urticaria Pigmentosa: Clinical, Dermoscopic, and Histopathological Insights

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Abstract

Urticaria pigmentosa, the most common form of cutaneous mastocytosis, results from dermal mast cell proliferation and typically presents in childhood. We report a 14-year-old male with multiple hyperpigmented maculopapular lesions over the trunk and extremities, associated with intermittent pruritus. The patient had been previously misdiagnosed as papular urticaria, with poor response to treatment. Clinical examination revealed a positive Darier's sign, with no features suggestive of systemic involvement. Dermoscopy showed a reticular pigment network on a light-brown background. Histopathological examination demonstrated dense dermal mast cell infiltrates, predominantly perivascular. Serum tryptase levels were within normal limits.

A diagnosis of urticaria pigmentosa was established, and the patient was treated with oral antihistamines, mast cell stabilizers, and avoidance of triggers, resulting in improvement.

This case highlights the potential for misdiagnosis of urticaria pigmentosa as papular urticaria and other pruritic dermatoses. Important differential diagnoses include papular urticaria, lichen planus pigmentosus, post-inflammatory hyperpigmentation, and early prurigo. Recognition of key clinical signs such as Darier's sign, supported by dermoscopy and histopathology, is essential for accurate diagnosis. Early identification facilitates appropriate management and evaluation to exclude systemic involvement.

Keywords: Urticaria pigmentosa, Cutaneous mastocytosis, Darier's sign, Mast cells

Introduction

Mastocytosis is a heterogeneous group of disorders characterized by abnormal accumulation of mast cells in one or more organ systems. According to the World Health Organization (WHO), it is classified into cutaneous mastocytosis, systemic mastocytosis, and mast cell sarcoma. Cutaneous mastocytosis includes maculopapular cutaneous mastocytosis (urticaria pigmentosa), solitary mastocytoma, and diffuse cutaneous mastocytosis, with additional variants such as telangiectatic forms described.[1]

Urticaria pigmentosa, the most common variant, typically presents with multiple hyperpigmented macules and papules and may demonstrate a positive Darier's sign. Dermoscopy is a useful non-invasive adjunct that often reveals a reticular pigment network on a yellowish-brown background, aiding in differentiation from other pigmented dermatoses.[2,3]

Case report

A 14-year-old male presented with multiple tan- to brown-colored maculopapular lesions over the trunk and bilateral upper limbs for the past 2 years, associated with pruritus that had worsened over the preceding 15 days. The scalp, face, palms, and soles were spared. There was no history of blistering or systemic symptoms such as flushing, abdominal pain, diarrhea, dizziness, palpitations, syncope, fever, weight loss, or bone pain. There was no family history of similar complaints, and the patient's general health was unremarkable.

Cutaneous examination revealed multiple well-defined, discrete hyperpigmented macules and papules over the trunk and upper limbs. Stroking of lesions elicited urtication and erythema (positive Darier's sign) (Figure 1a–c). There was no hepatosplenomegaly on ultrasonography of the abdomen and pelvis.

Dermoscopy demonstrated a reticular pigment network on a light-brown background (Figure 2). Histopathological examination showed dense infiltrates of mast cells in the superficial and upper dermis, predominantly in a perivascular distribution. The mast cells were round to spindle-shaped with abundant granular cytoplasm and eccentrically placed nuclei. Scattered eosinophils, lymphocytes, and histiocytes were also noted. The overlying epidermis showed basal layer hyperpigmentation. Special stains such as Giemsa and toluidine blue demonstrated metachromatic granules within mast cells, confirming their identity (Figure 3a, figure 3b) These histopathological findings were consistent with and confirmed the diagnosis of urticaria pigmentosa. Laboratory investigations, including complete blood count, liver and renal function tests, urinalysis, and chest radiography, were within normal limits. Serum tryptase levels were normal.

The patient was managed with oral antihistamines, oral ketotifen (1 mg twice daily), and topical tacrolimus, along with advice to avoid known mast cell degranulating triggers. Significant symptomatic improvement was noted over 8 weeks.

Discussion

Urticaria pigmentosa is characterized by dermal infiltration of mast cells and release of mediators such as histamine, resulting in its clinical manifestations. It commonly presents as multiple yellowish-brown to reddish-brown macules and papules over the trunk and proximal extremities, often associated with pruritus. Darier's sign is a characteristic feature due to mast cell degranulation. In some patients, systemic symptoms such

as flushing, gastrointestinal complaints, or, rarely, anaphylaxis may occur.[1,4]

The pathogenesis is primarily linked to activating mutations in the KIT proto-oncogene, most commonly D816V, leading to constitutive mast cell proliferation and survival. Mast cell–derived mediators, including histamine, prostaglandins, leukotrienes, and cytokines, contribute to pruritus, vasodilation, and other clinical features.[4] In our case, the patient was initially misdiagnosed as papular urticaria, highlighting the clinical overlap with other pruritic dermatoses. Dermoscopy revealed a reticular pigment network without background erythema, corresponding to increased melanogenesis and basal layer hyperpigmentation induced by mast cell–derived growth factors. Similar findings have been described in previous studies, although additional features such as a reddish background may be variably present.[2,3]

Histopathological examination confirmed the diagnosis by demonstrating dense mast cell infiltrates in the papillary and upper reticular dermis, predominantly in a perivascular distribution. The mast cells showed round to oval morphology with granular cytoplasm and were highlighted by special stains such as Giemsa and toluidine blue, along with CD117 positivity.[5]

Evaluation for systemic involvement is essential in all patients. In our case, there were no clinical features suggestive of systemic disease, and serum tryptase levels were within normal limits, making systemic mastocytosis unlikely. Serum tryptase serves as a useful screening marker, and elevated levels warrant further evaluation, including bone marrow examination in selected cases.[1,6]

Management is primarily symptomatic and aimed at reducing mast cell mediator release and avoiding triggers. Patients should be counseled to avoid precipitating factors such as friction, heat, alcohol, and certain medications. First-line therapy includes oral H1 antihistamines, with H2 antihistamines and mast cell stabilizers used in selected cases. Topical therapies or phototherapy may be considered in persistent disease.[1,6]

The dermoscopic differential diagnosis includes melanocytic nevi, lichen planus pigmentosus, dermatofibroma, and post-inflammatory hyperpigmentation, which may show a pigment network, whereas papular urticaria and prurigo typically lack a structured network.[7]

Early recognition of urticaria pigmentosa is important for appropriate management and for identifying patients who may require further systemic evaluation.

Conclusion

Urticaria pigmentosa should be suspected in patients with persistent pigmented maculopapular lesions and a positive Darier's sign. Dermoscopy serves as a valuable non-invasive tool, aiding in early recognition by demonstrating a characteristic pigment network. Diagnosis is confirmed by histopathology, and evaluation for systemic involvement, including serum tryptase levels, is essential. Management is mainly symptomatic with antihistamines and avoidance of triggers. Early diagnosis and regular follow-up are important for optimal outcomes.

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Legend Figures

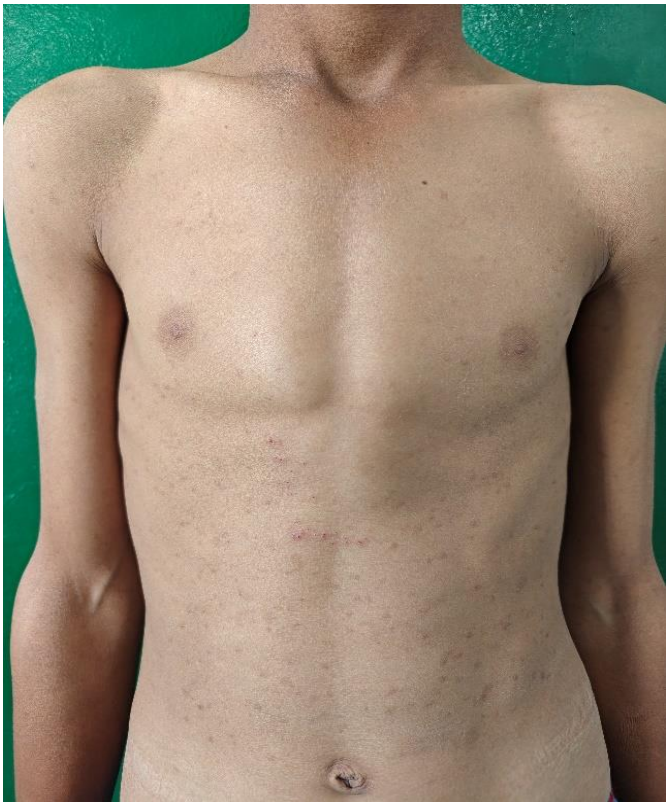


Figure 1 a: Multiple well defined brown coloured macuopapular lesion present over trunk

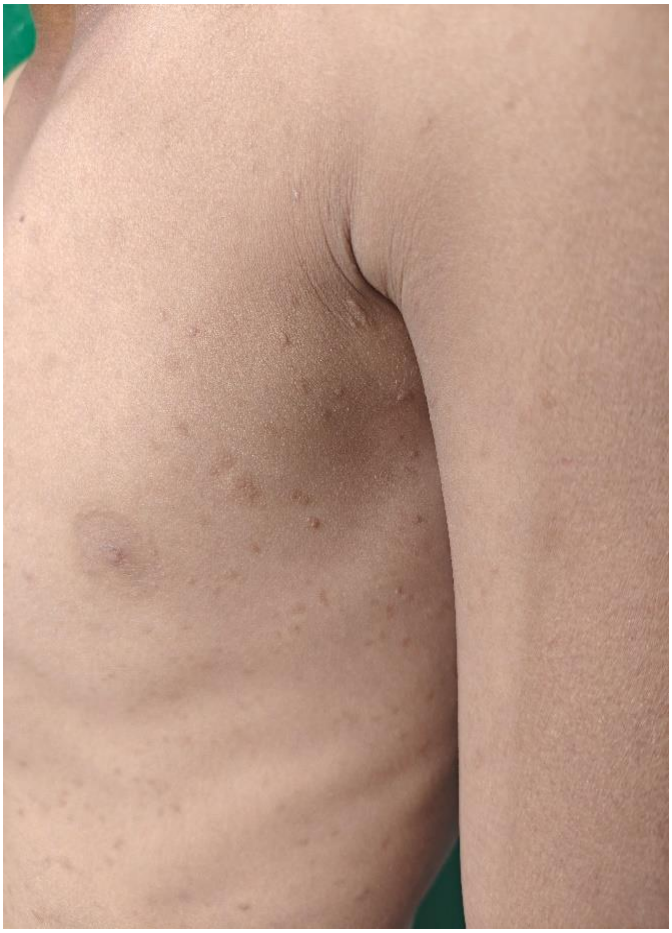
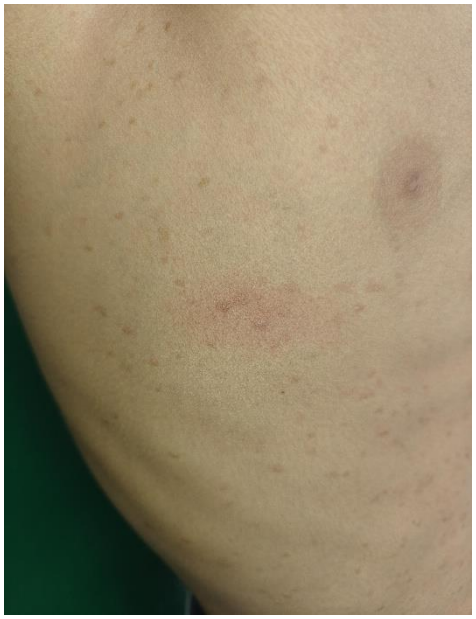


Figure 1b: Macuolopapular lesion present over axilla



Igure 1c: Darier's sign positive

Figure 2: Dermoscopy showed pigment network with a light-brown background

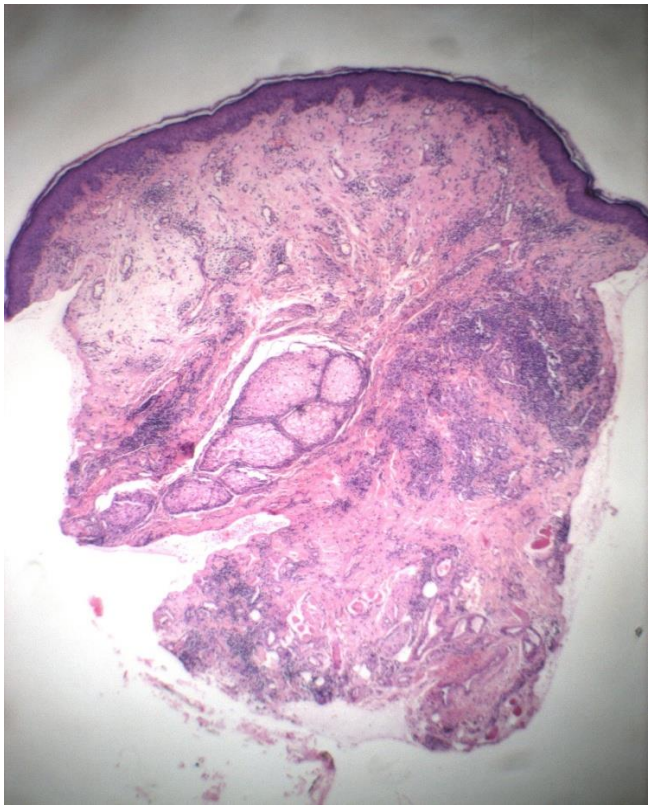


Figure 3a: Epidermis showing basal layer hyperpigmentation with diffuse inflammatory cell infiltrate in the dermis (H&E, 10×).

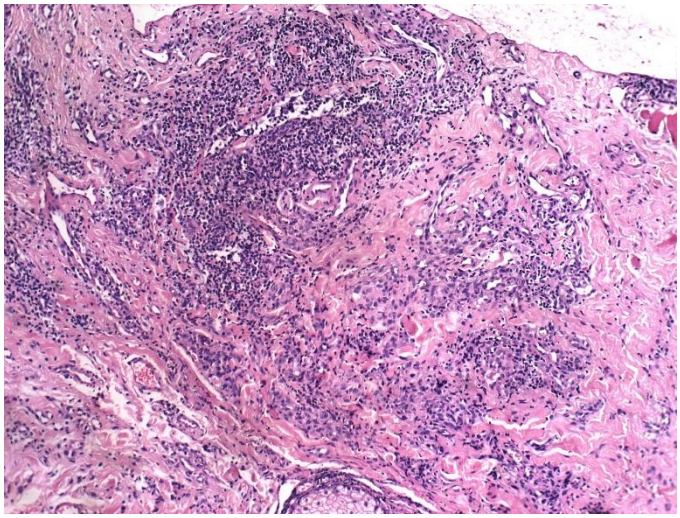


Figure 3b: Low-power photomicrograph showing diffuse dermal cellular infiltrate with sparing of the epidermis (H&E, 10×).

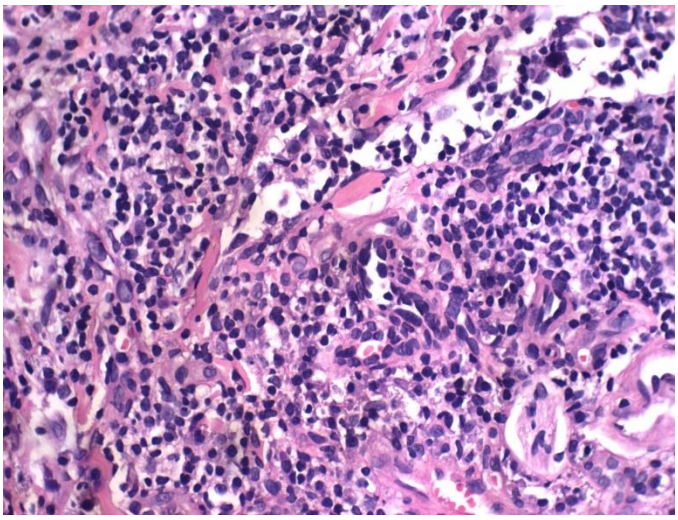


Figure 3c : High-power view demonstrating dense dermal infiltrate composed predominantly of mast cells (H&E, 40×)

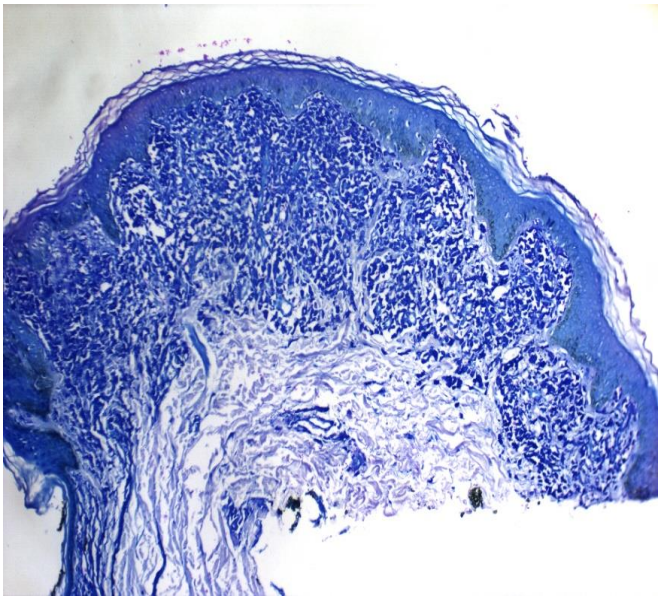


Figure 3d- Special stain showing diffusely positive for mast cells (Giemsa stain; 10X)