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Case Report

Neonatal-onset maculopapular cutaneous mastocytosis presenting with pigmented plaques in an infant

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ABSTRACT

Cutaneous mastocytosis is a clonal disorder characterized by abnormal accumulation of mast cells in the skin. Maculopapular cutaneous mastocytosis (urticaria pigmentosa) is the most common form in children and usually presents within the first two years of life. Neonatal onset and blistering may complicate recognition and mimic other pigmentary dermatoses. We report a 10-month-old male with six well-defined brown plaques measuring 2–3 cm, two with central scars. A blister was noted at birth on the knee, followed by progressive lesion development. Physical examination revealed no lymphadenopathy or visceromegaly. Histopathology demonstrated dense dermal mast cell infiltration with metachromatic granules on Giemsa staining, confirming maculopapular cutaneous mastocytosis. The patient was treated with oral ketotifen with stable disease at six-month follow-up. Early recognition is essential to ensure appropriate counseling, monitoring, and avoidance of unnecessary investigations.

Keywords: Maculopapular cutaneous mastocytosis, Urticaria pigmentosa, Pigmented plaques

INTRODUCTION

Mastocytosis is a neoplastic disorder characterized by clonal proliferation and accumulation of mast cells in one or more organs.[1] According to the International Consensus Classification, it is divided into cutaneous and systemic forms.[1,2] Cutaneous mastocytosis is the most common presentation in childhood and includes cutaneous mastocytoma, diffuse cutaneous mastocytosis, and maculopapular cutaneous mastocytosis (urticaria pigmentosa).[3,4]

In paediatric patients, lesions usually appear within the first two years of life and often follow a benign course with partial or complete regression over time.[5,6] However, neonatal onset and blistering may obscure the diagnosis and raise concern for other pigmentary or inflammatory dermatoses. Histopathological confirmation is helpful in atypical presentations.

CASE REPORT

A 10-month-old male presented with disseminated pigmented plaques involving the left side of the back, lower abdomen, right inguinal region, and ipsilateral knee. Six brown plaques measuring 2–3 cm in diameter were observed, with well-defined borders and smooth surfaces; two exhibited central eutrophic scars (Figures 1a-c). No lymphadenopathy or visceromegaly was detected. Darier's sign was not elicited.

According to the mother, the child had developed a blister on the right knee at birth, which evolved into a hyperpigmented lesion. Additional plaques appeared progressively over the following months.

An incisional biopsy from a shoulder plaque revealed dense dermal infiltration extending from the papillary to the mid-reticular dermis (Figure 1d). The infiltrate consisted predominantly of mast cells with basophilic cytoplasm and granules. Giemsa staining demonstrated metachromatic granules within the cytoplasm (Figure 1e), consistent with maculopapular cutaneous mastocytosis.

Treatment with oral ketotifen (0.05 mg/kg every 12 hours) was initiated. After six months of follow-up, no new lesions had developed and the clinical course remained stable.

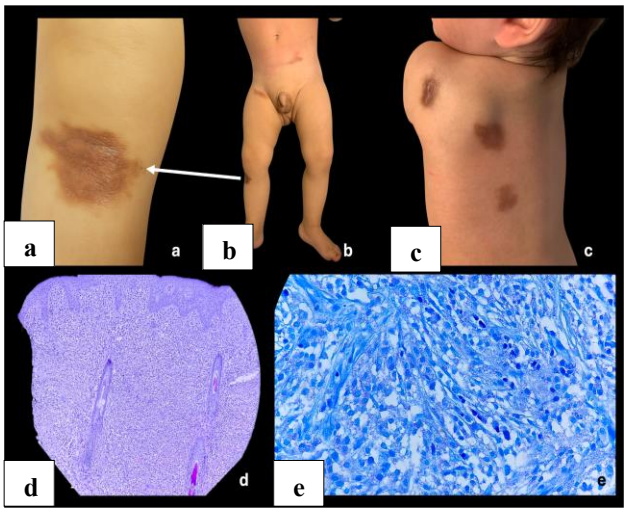

Figure 1: (a) Brown plaque measuring 3 cm in diameter on the right knee, with well-defined borders and a smooth surface, showing a central eutrophic scar; (b) three brown plaques distributed over the lower abdomen, right inguinal region, and knee; (c) three brown plaques on the left side of the back, one exhibiting a central eutrophic scar; (d) skin biopsy specimen from a shoulder plaque stained with hematoxylin and eosin (original magnification ×10), showing dense dermal cellular infiltration; and (e) Giemsa-stained section (original magnification ×40) demonstrating mast cells with cytoplasmic metachromatic granules.

DISCUSSION

Maculopapular cutaneous mastocytosis typically presents as yellow-brown to reddish-brown macules or papules, predominantly affecting the trunk. Mechanical stimulation may induce urtication (Darier's sign) and, in some cases, blistering due to mast cell mediator release.[3,7] Although suggestive, Darier's sign should be elicited cautiously, as mediator release can provoke systemic symptoms in susceptible individuals.[3]

Blistering at onset, particularly in neonates, may complicate recognition and broaden the differential diagnosis.[8] Conditions such as post-inflammatory hyperpigmentation, café-au-lait macules associated with neurofibromatosis, Langerhans cell histiocytosis, pigmentary incontinence, lentigines, and melanocytic nevi should be considered.[7]

Initial evaluation should focus on excluding systemic involvement. Physical examination must assess hepatosplenomegaly and lymphadenopathy. Complementary studies may include complete blood count, liver function tests, and abdominal ultrasound when clinically indicated.[7] Baseline serum tryptase levels are usually normal or only mildly elevated in isolated cutaneous forms; persistent elevation warrants further evaluation.[7]

Histologically, cutaneous mastocytosis demonstrates dermal infiltrates of mast cells arranged in perivascular, interstitial, or nodular patterns. Special stains such as Giemsa highlight metachromasia and facilitate identification.[3] Avoiding vigorous manipulation before biopsy is recommended, as degranulation may interfere with histopathological interpretation.[3]

Management is primarily symptomatic and preventive. Education regarding avoidance of triggers—including friction, sudden temperature changes, infections, and certain medications such as NSAIDs or opioids—is essential.[3,10] Oral H1 antihistamines are first-line therapy for pruritus and erythema.[3] Ketotifen is a dual-action H1 antihistamine and mast cell stabilizer that has been used in the symptomatic management of mastocytosis and other mast cell-related disorders.[9] The prognosis in paediatric cutaneous mastocytosis is generally favorable, with many cases stabilizing or regressing over time.[5]

CONCLUSION

This case highlights an atypical presentation of paediatric maculopapular cutaneous mastocytosis with neonatal blistering onset, which expands knowledge about the clinical diversity of this entity and its implications for an accurate and timely diagnostic approach. Careful evaluation to exclude systemic involvement, combined with symptomatic treatment and avoidance of known triggers, remains the cornerstone of management. The favorable clinical course observed during follow-up is consistent with the generally benign prognosis of childhood cutaneous mastocytosis, underscoring the importance of early diagnosis, appropriate counseling, and regular monitoring.

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