

Visual Treats in Dermatology

## Leonine facies in diffuse cutaneous mastocytosis – A unique feature

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Received: 04 August 2025  
Accepted: 12 August 2025  
Published: 25 September 2025

DOI  
10.25259/CSDM\_142\_2025

Quick Response Code:



A 23-year-old female came with asymptomatic multiple skin colored to red raised skin lesions over the face and body since the 10<sup>th</sup> year of age. There was progressive diffuse thickening of the skin. There was no history of urticaria, angioedema, flushing, anaphylaxis, nausea, vomiting, abdominal pain, diarrhea, bone pain, loss of weight, or appetite. On cutaneous examination, there was diffuse infiltration with skin-colored infiltrated plaques noted over the face with loss of eyebrows (leonine facies) and papules along the inner eyelid margins and pinna [Figure 1]. Diffuse infiltration over the upper limbs, trunk, and lower limbs was present. A few discrete infiltrated papules and plaques with peau d'orange appearance were also seen. Darier's sign was positive. Nails and mucosae were normal. Her hemogram showed thrombocytopenia. Serum tryptase level was 128 µg/L, ultrasound abdomen demonstrated splenomegaly, and dual energy X-ray absorptiometry (DEXA) scan showed osteopenia. Estimation of parathyroid hormone and Vitamin D levels was normal. Skin biopsy report was diagnostic of diffuse cutaneous mastocytosis (DCM). Bone marrow aspirate demonstrated 45% mast cells and flow cytometry, CD117, CD2, and CD25-positive cells. The KIT D816V mutation was present. Based on the above findings, the patient was diagnosed as having DCM with systemic involvement (bone marrow). The patient was referred to the regional cancer center, medical oncology, for possible use of imatinib.

Mastocytosis is a heterogeneous disease characterized by the expansion and accumulation of neoplastic mast cells in various tissues. DCM is a rare and severe form of cutaneous mastocytosis,



**Figure 1:** Diffuse infiltration with skin-colored infiltrated plaques noted over the face with loss of eyebrows (Leonine facies) and infiltration of the pinna (Frontal view-1a, lateral view-1b).

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characterized by a diffuse infiltration of mast cells in the skin, often leading to erythema, blistering, and skin thickening.<sup>[1]</sup> It is typically diagnosed in early infancy and can be associated with systemic complications due to mast cell degranulation. In our case, erythema and blistering were absent, but diffuse infiltration was apparent on the face and pinna, resembling the leonine facies seen in lepromatous leprosy. Treatment of DCM focuses on symptom management, mainly including antihistamines and mast cell stabilizers.<sup>[1]</sup> In extremely severe cases, systemic steroids, tyrosine kinase inhibitors, phototherapy, or omalizumab may be considered. Patients should be equipped with an adrenaline autoinjector to tackle anaphylaxis.

**Ethical approval:** Institutional review board approval is not required.

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent.

**Financial support and sponsorship:** Nil.

**Conflicts of interest:** Dr. Devinder Mohan Thappa has been an editorial board member.

**Use of artificial intelligence (AI)-assisted technology for manuscript preparation:** The author confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

## REFERENCE

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**How to cite this article:** Thappa DM. Leonine facies in diffuse cutaneous mastocytosis – A unique feature. *CosmoDerma*. 2025;5:104. doi: 10.25259/CSDM\_142\_2025