Solitary mastocytoma with positive Darier's sign

Brijesh Nair, Sidharth Sonthalia[1], Ishad Aggarwal[2]

Department of Dermatology, INHS Sanjivani Hospital, Kochi, Kerala, [1]Department of Dermatology, Skinnocence: The Skin Clinic, Gurgaon, Haryana, [2]Department of Dermatology, IPGMER, Kolkata, West Bengal, India

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Address for correspondence:
Dr. Sidharth Sonthalia, Skinnocence: The Skin Clinic, C-2246, Sushant Lok-1, Block-C, Gurgaon - 122 009, Haryana, India.
E-mail: sidharth.sonthalia@gmail.com

A 15-month-old girl child presented with a 6-month-old solitary nodular lesion over her lower abdomen. No symptoms were reported by the parents, except for temporary redness and swelling of the lesion developing while manipulating. Systemic symptoms such as spontaneous hives, flushing, wheezing, unexplained diarrhea, or bone pains were absent. Cutaneous examination revealed a 2.5 × 1.5 cm sized firm, nontender, yellow-tan to light-brown noduloplaque with a characteristic "peau d'orange" surface and rubbery consistency [Figure 1]. Gentle stroking of the lesion evoked erythema and swelling within 30 seconds [Figure 2], which became pronounced in 2 minutes [Figure 3], signifying a positive Darier's sign. Systemic examination was unremarkable. Histopathology of the lesion revealed mast cell aggregates in the papillary dermis, which was confirmed on toluidine blue staining. A diagnosis of solitary mastocytoma was made. Parents were reassured about its benign nature and advised to avoid triggering factors such as rubbing the lesion.

Mastocytosis is characterized by abnormal proliferation of mast cells, involving only the skin (cutaneous mastocytosis or CM) or extracutaneous organs (systemic mastocytosis). Skin is the most commonly involved organ, especially in children. Solitary mastocytoma (SM), the second most common type of CM, accounts for 10%–15% of the total cases of CM.[1] The other variants of CM include urticaria pigmentosa (the most common variant), diffuse CM, and telangiectasia macularis eruptive perstans (TMEP). A mastocytoma is usually solitary that presents as a yellow to tan-brown nodule or plaque, ranging 0.5–3 cm in diameter. A shiny peau d'orange or "orange peel" appearance with a rubbery consistency is characteristic of a classical lesion. Common locations include the trunk, head and neck, and extremities. Although lesions may rarely appear in adulthood, majority present during infancy; with only 10% appearing after 2 years of age.[2] The lesions may be asymptomatic or become itchy and swollen on rubbing. The clinical differential diagnosis includes melanocytic naevi, xanthomas, and xanthogranulomas.[3]

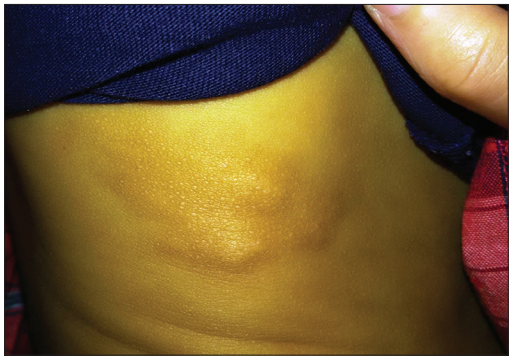

Figure 1: Yellow-tan to light-brown nodule over the right lower abdomen of a 15-month-old girl child, with an "orange peel" appearance

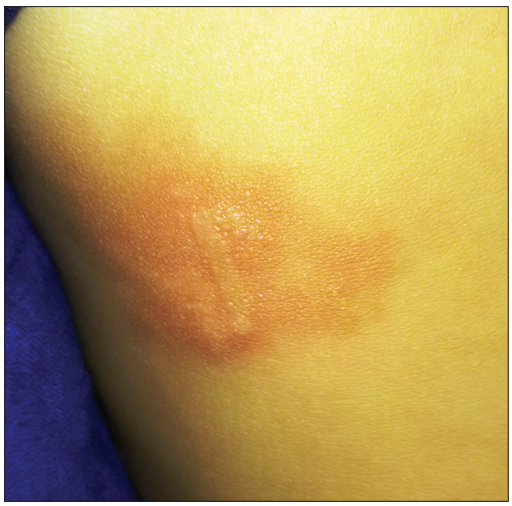

Figure 2: Appearance of lesional erythema and mild edema after 30 s of gentle stroking with a key in the center of the lesion (the vertical mark in the center being the site of stroking

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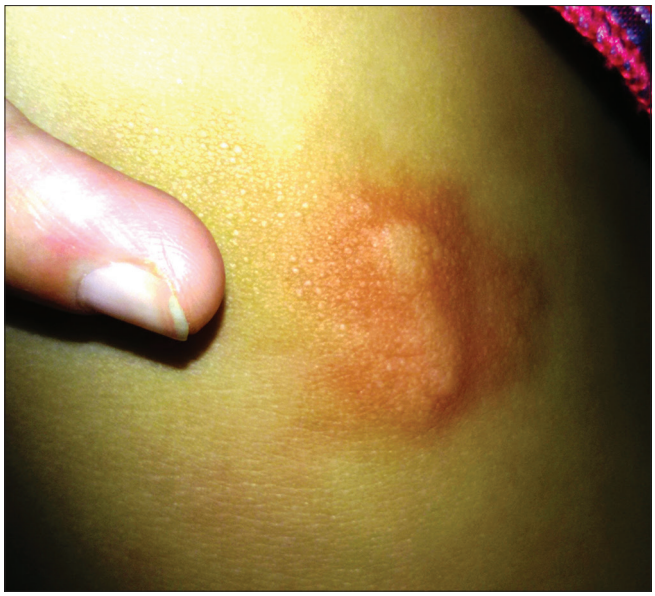

Figure 3: Pronounced lesional and perilesional erythema and edema within 2 min signifying a strongly positive Darier's sign

Darier's sign, named after the French dermatologist Ferdinand-Jean Darier, is considered to be a foretoken of CM. Gentle rubbing or stroking of lesions of cutaneous mastocytosis is followed by local itching, erythema, and weal formation within 2–5 min, which may last from 30 min to several hours. The pathophysiology of Darier's sign involves stroking-induced degranulation of the increased number of functionally normal dermal mast cells, followed by exocytosis of effector molecules such as histamine and heparin.[4] Of all the variants of CM, Darier's sign is most commonly encountered in urticaria pigmentosa, with up to 94% positivity reported in children with urticaria pigmentosa in a clinical analysis of 71 childhood cases with CM.[5] However, its positivity has been reported in only 50% cases of SM.[3] Intriguingly, Darier's sign has also been reported in conditions other than CM, such as leukemia cutis, juvenile xanthogranuloma, histiocytosis X, and rarely in cutaneous large T-cell lymphoma. Darier's sign needs to be severalized from *Pseudo-Darier's sign*, an augury of congenital smooth muscle hamartomas, characterized by transient piloerection and elevation or increased induration of the lesion induced by rubbing.[4]

The diagnosis of SM is confirmed on lesional biopsy with histopathology revealing mast cell aggregates in the papillary dermis, confirmed by metachromatic stains such as toluidine blue. The condition is benign and self-limited, with systemic involvement being uncommon and spontaneous involution of the lesion expected by puberty, rendering laboratory evaluation de trop. Reassurance along with avoidance of triggering factors such as pressure, friction, and intake of mast cell degranulating agents (eg, aspirin) suffice in most cases. Medical treatment with antihistamines, topical or intralesional steroids, or topical tacrolimus is indicated in select cases.[2,3]

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Conflicts of interest

There are no conflicts of interest.

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