



Darier's sign

urtication and erythematous halo that are produced in response to rubbing or scratching of lesions of cutaneous mastocytosis.

In classical Darier's sign, gentle rubbing or stroking of the lesions, is followed by local itching, erythema and weal formation within 2-5 min. This may persist from 30min to several hours. In young children, there may be vesiculation in the stroked lesion. Although classically positive in lesional skin, this sign may even be demonstrated on clinically normal skin in patients of mastocytosis.

In pseudoxanthomatous mastocytosis, a variant of diffuse cutaneous mastocytosis, there will be only erythema without urtication on rubbing as against a classic Darier's sign.

In cutaneous mastocytosis, there is an increased number of functionally normal mast cells in the dermis. When the skin is stroked, there is degranulation of the mast cells with the release of inflammatory granules that contain histamine, slow-releasing substance of anaphylaxis (SRSA), eosinophil chemotactic activating factor (ECAAF) and heparin . It is the histamine that is responsible for the response.

It is also suggested that besides exocytosis of single or multiple granules, there is another pattern of degranulation when mast cells rupture. This happens following physical stimuli such as stroking. A gentle action gives rise to exocytosis, while a strong action leads to rupture of the cells.

Exact pathophysiology of Darier's sign in lymphoma and leukemias, is not known. One hypothesis suggested that mast cell proliferation may be related to lymphokine release, thereby explaining their close relation with lymphocytes. [5] Another hypothesis suggested that laminin increases mast cell attachment to the lymphocytes. [6] In juvenile xanthogranuloma, the histiocytes of the lesions undergo macrophagic differentiation to dermal dendrocytes, and the distribution of mast cells in the skin coincides with that of dermal dendritic cells.

Conditions Associated With Darier's Sign

Cutaneous mastocytosis: In urticaria pigmentosa, the most frequent clinical form of cutaneous mastocytosis, Darier's sign was present in 94% of cases.

Leukemia cutis: Leukemia cutis occurs in 25-30% of infants with congenital leukemia and is frequently associated with acute myeloid leukemia than with acute lymphoblastic leukemia.

Urticaria-pigmentosa-like lesions have been reported in acute lymphoblastic leukaemia.

Juvenile xanthogranuloma: It is the most common form of non-Langerhans cell histiocytosis.

Nagayo et al. reported Darier's sign in this disorder.

Histiocytosis X : Foucar et al. described positive Darier's sign in a patient with 'mast cell rich variant' of histiocytosis X.

Lymphoma: On rare instances, Darier's sign has been reported in cutaneous large T-cell lymphoma and in non-Hodgkin's lymphoma.

Pseudo-Darier's sign : It is a transient piloerection and elevation or increased induration of a lesion induced by rubbing and is observed in congenital smooth muscle hamartomas. A positive pseudo-Darier's sign can be helpful in clinically distinguishing congenital smooth muscle hamartoma from congenital hairy nevus.

Dermatographism ("skin writing"): It is a form of physical urticaria that consists of local erythema due to capillary vasodilatation, followed by edema and a surrounding flare due to axon reflex induced dilation of arterioles, which is observed after the firm stroking of skin. The cause of this phenomenon is thought to be hypersensitivity of the mast cells rather than an increase in the mast cells, as observed in mastocytosis