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Photodermdiagnosis
What caused this plaque?

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A 10-year-old girl was bitten by a wasp on the back of her right chest about 4 years ago. Ever since she complained of a stinging sensation over the affected site off and on, but it was not until a few days earlier that her mother noticed a hyperpigmented slightly scaly plaque on the back of the right chest of her daughter. The itching worsened after intake of spicy foods. Malaise, fever and upper abdominal discomfort were also associated with episodes of increased pruritus over the plaque. However, rest of the systemic review was unremarkable.

On physical examination, a well defined dark brown slightly scaly plaque was present (**Figure 1** and **2**). There was no hepatosplenomegaly and lymphadenopathy. Her differential eosinophil count was 7% and absolute count was 602/cmm. A biopsy was also performed (**Figure 3**).

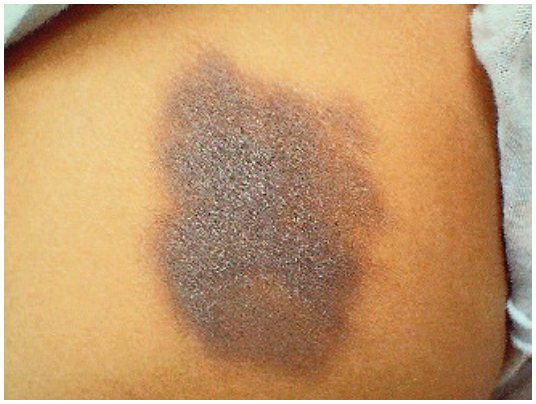

Figure 1

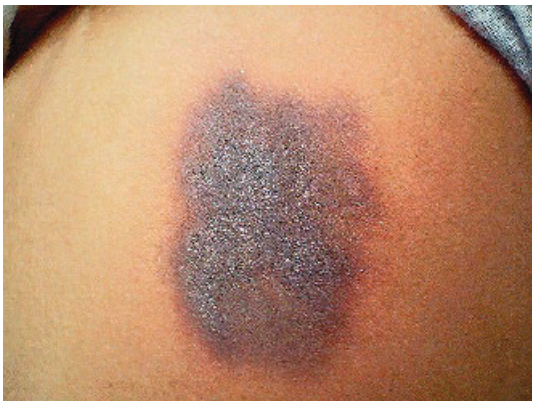

Figure 2

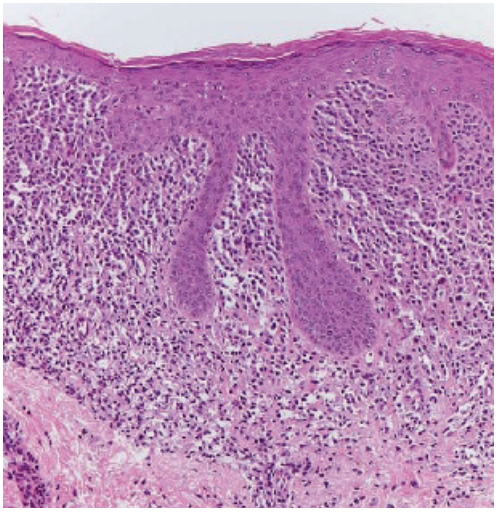

Figure 3

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Diagnosis

Solitary mastocytoma

The histopathology revealed mast cells infiltrating the whole papillary dermis.

Discussion

Mastocytosis is a heterogeneous group of disorders characterized by proliferation of mast cells infiltrating only skin or other tissues like gastrointestinal tract, liver, spleen, bone marrow, lymph nodes and skeletal system. When only skin is involved, in the absence of systemic features, the condition is referred to as cutaneous mastocytosis. Different morphological variants of cutaneous mastocytosis include urticaria pigmentosa, telangiectasia macularis eruptiva perstans, mastocytoma and diffuse cutaneous mastocytosis.[1]

Mutations in the proto-oncogene c-*kit* occur in certain forms of mastocytosis. c-*kit* is expressed on mast cells, hematopoietic stem cells, germ cell lineages, and melanocytes. The regulation of mast cell number and mast cell differentiation is controlled by factors produced both in the bone marrow and by cells in the tissues in which mast cells finally reside. c-*kit* ligand, also known as *stem cell factor (SCF)* is an important mediator for human mast cell differentiation. Final maturation and granule composition depend on factors produced in the tissue microenvironment, including SCF produced by fibroblasts and stromal cells. Inhibition of local production of SCF leads to mast cell apoptosis. The receptor for SCF is c-*kit.* Mutations involving c-*kit* are associated with enhanced receptor function and may contribute to the increase in mast cell number that is characteristic of mastocytosis. Insect stings like many drugs e.g. aspirin, narcotics, iodinated contrast media, alcohol, exercise, and infections, can provoke flushing and vascular collapse in preexisting mastocytosis. However, their role in initiating mastocytosis de novo has to be explored.

Solitary mastocytoma represents uncommon variant of cutaneous mastocytosis.[3] The disease usually starts in infancy; however, onset in adults has been reported.[4] Clinically, it is characterized by yellow, skin-coloured, erythematous or brownish plaque which shows typical Darier's sign. Occasionally, blistering may occur on vigorous rubbing. Clinically, the disease may resemble pigmented epidermal nevus, Becker's nevus, pigmented spindle cell/or epithelioid cell nevus, pigmented dermatofibroma, pigmented actinic keratosis, seborrheic keratosis, large cell acanthoma, granuloma faciale, Kaposi's sarcoma or pigmented basal cell carcinoma. However, all these conditions can be differentiated on the basis of their duration and peculiar histopathology using special stains. The study of biopsy tissue in patients with suspected mastocytosis requires the use of appropriate stains. Tryptase is the stain of choice, as toluidine blue and Giemsa stains are more likely to be affected by tissue processing and may not always produce reliable results. On histopathology, using Giemsa stain, there is accumulation of monotonous mast cells extending from upper dermis to subcutaneous tissue. Electron microscopy may reveal Charcot-Leyden crystals.[5]

The diagnosis of cutaneous mastocytosis is based on; 1) typical skin lesions - urticaria pigmentosa, diffuse cutaneous mastocytosis, or mastocytoma; and 2) positive biopsy of affected skin, with typical infiltrates of mast cells in a diagnostic pattern

Therapy is generally conservative and aimed at alleviation of symptoms with H1 and H2 antagonists, avoidance of mast cell degranulating stimuli and potent topical steroids.

The prognosis for mastocytoma is usually excellent; lesions usually involute in a few years and no systemic involvement has been reported.

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