Case 15

A 20 year-old Thai woman

Chief complaint: Multiple yellow-tan macules on face and upper chest since 3 years of age.
Present illness: The lesions gradually increased in number with occasionally pruritus. Scratching caused swelling and redness of the lesions.
Past history She had normal growth and development and had no known of underlying disease
Family history No other family members were affected with similar lesions
Physical examination A young alert woman, Not pale no jaundice, no enlargement of cervical lymph nodes. Liver and spleen were not palpable
Skin examination Multiple discrete oval shape yellow-tan macules distributed over the face upper chest and trunk.(Fig 15-1 Diascopic view of forehead lesion) Darier sign was positive after stroking the lesion.

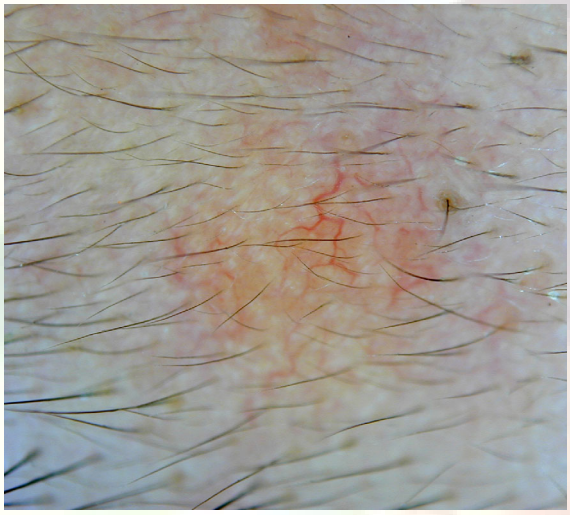

Fig 15.1

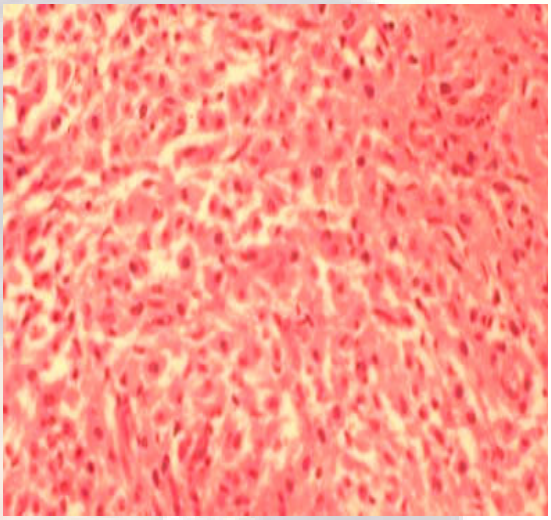

Fig 15.2

Histopathology (S02-8416)

The section shows monomorphous superficial perivascular and interstitial infiltration of predominantly mast cells and some eosinophils

Diagnosis: Urticaria pigmentosa

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Discussion

Mastocytosis is a disorder characterized by mast cell proliferation and accumulation within various organs, most commonly the skin. Urticarial pigmentosa is the most common form. Most patients with mastocytosis are children. Incidence peaks again in patients aged 30-49 years.

Mastocytosis probably is a hyperplastic response rather than a neoplastic condition. Increased local concentrations of soluble mast cell growth factor in lesions of cutaneous mastocytosis are believed to stimulate mast cell proliferation, melanocyte proliferation, and melanin pigment production. The induction of melanocytes explains the hyperpigmentation that commonly is associated with cutaneous mast cell lesions. Associated systemic manifestations are believed to reflect the release of mast cell–derived mediators, such as histamine, prostaglandins, heparin, neutral proteases, and acid hydrolases. Symptoms and signs include headache, flushing, dizziness, tachycardia, hypotension, syncope, abdominal pain, and diarrhea. The skeletal, hematopoietic, gastrointestinal (GI), cardiopulmonary, and central nervous systems may be involved either directly, via mast cell infiltration, or indirectly, via mast cell mediator release.

The skin manifestation in mastocytosis is characterized by red-brown oval macules, papules or plaques ranging in number of a few to thousands. When the lesion is stroked, it typically urticates, becoming pruritic, edematous, and erythematous. This change is referred to as the Darier sign.

Histologic examination reveals dermal mast cell infiltrates, especially in the papillary dermis around blood vessels . Diagnosis of UP may require demonstration of mast cell granules using Giemsa stain or

toluidine blue stain. Most cases of UP in children resolve spontaneously. Patients with adult-onset and adolescent-onset UP are more likely to have persistent disease and are at greater risk for systemic involvement. Juvenile-onset systemic mastocytosis has a malignant transformation rate as high as 7%, while adult-onset systemic mastocytosis has a malignant transformation rate as high as 30%.

Therapy is conservative and aimed at symptom relief with H1 and H2 antihistamine because the prognosis for most patients with mastocytosis is excellent. None of the currently available therapeutic measures induces permanent involution of cutaneous or visceral lesions. Advise patients to avoid agents that precipitate mediator release, such as aspirin, NSAIDs, codeine morphine, alcohol, thiamine, quinine, opiates, gallamine, decamethonium, procaine, radiographic dyes, dextran, polymyxin B, scopolamine, and D-tubocurarine.

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