CASE 14

A 65-year-old Thai man

Chief Complaint:

Asymptomatic brownish macules on upper back and both posterior arms.

Present Illness:

He developed multiple discrete asymptomatic brownish macules on the posterior aspect of both arms for a few weeks. All the skin lesions appeared progressively about two weeks and had remained stable since. He denied having pruritus, diarrhea, abdominal pain, headache, anaphylaxis or bone pain.

Past History:

Two weeks before this visit, he presented with arthritis on the right wrist and had received oral diclofenac for one week.

He has diabetes mellitus, hypertension, benign prostate hypertrophy and dyslididemia.He is taking Aspent M, Ketotifen , Amlodipine and Harnol.

He has never had history of alcohol drinking.

Family History:

Unremarkable

Physical Examination:

Symmetrical, multiple, discrete, 2- to 4-mm tannish brown macules with minimal telangiectasia on the upper back and posterior aspect of both arms. Darier 's sign : negative

Histopathology (S06-09263)

Superficial perivascular and interstitial inflammatory cell infiltration, mild epidermal hyperplasia and epidermal hyperpigmentation.

Most of the cell infiltrate are mast cells.

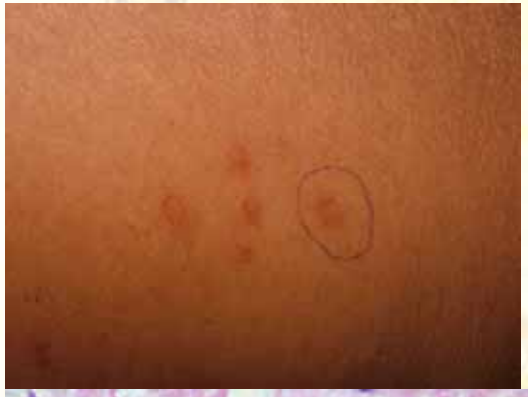

Fig. 14.1

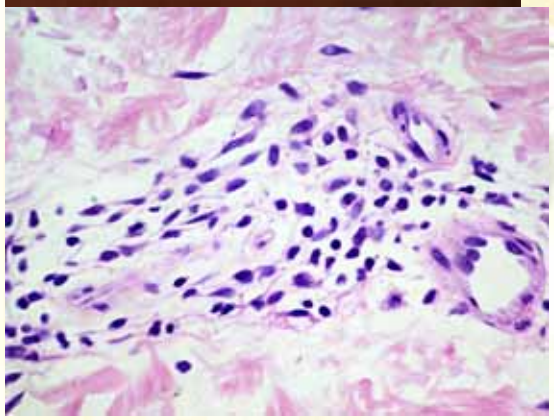

Fig. 14.2

Diagnosis : Urticarial Pigmentosa
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Discussion

Mastocytosis is a condition characterized by abnormal proliferation and accumulation of mast cells in the skin and at various systemic sites[1] cause by c-kit mutation. The prevalence of the disease is unknown and familial occurrence appear unusual, familial inheritance has been reported only 50 families since the mid 1880s. There were four cases of TMEP occurring in three generation of a family[5].

Clinical symptoms are due to the release of mast cell mediators, including flushing, urticaria, angioedema, anaphylaxis, peptic ulcer, abdominal pain, diarrhea and bone pain. In this case, the clinical presentation is local cutaneous without systemic symptoms.

From classification of cutaneous mastocytosis, skin manifestation of this case show symmetrical discrete tan to brown macules and minimal telangiectasia , so the differential diagnosis confines within

1. Urticarial pigmentosa (UP) : is the most common skin manifestation of cutaneous disease [1,3] in both adults and childrens. It usually present with small, yellow-tan to reddish brown macules or slightly raised papules scattered over the body, palm, sole, face. Histopathology show perivascular mast cell infiltration.

2.Telangiectasia macularis eruptive perstans (TMEP): is a rare form mastocytosis and usually seen in adult. It usually present with round oval shaped[2] and telangiectases, hundreds of lesions (trunk>extremities). Histopathology show an increase number of mast cells around the dilated thin-wall blood vessels in the papillary dermis. In both of the condition, mild trauma including scratching or rubbing of the lesion causes urtication and erythema around the lesion. This is known as Darrier's sign. Although Darier's sign is pathognomonic of cutaneous mastocytosis, in IJDVL 2006 reported 2 cases from 6 cases of cutaneous mastocytosis that Darier's test was negative.[4]

Plan for investigation include

1. Direct study: special stains such as toluidine blue, Giemsa, labeled avidin or monoclonal antibodies to tryptase for identifying tissue mast cells.[6]

2. Indirect study: 24-urinary histamine and total serum tryptase for detection of circulating mast cell mediators.

Therapeutic for mastocytosis consist of avoidance of potential mast cell stimuli (alcohol, anticholinergic medications, aspirin, NSAID, polymyxin B sulfate and heat friction). Systemic therapy are H1 and H2 histamine antagonists, oral cromolyn sodium and systemic corticosteroids.[6] Skin-targeted therapies that lead to a resolution of the lesion are PUVA and toplcal corticosteroid therapy either by occlusion or intralesional injection for a limited number of lesion.[7]

References:

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