

Dermato-venereological Quiz

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A one-year old female baby presented to us with multiple itchy brownish papules and macules over the body since 3 months of age. The lesions started at the nape of neck and spread downwards involving trunk and limbs. She had been seen by various doctors but failed to respond to topical treatments. The baby was not on any long term medications and the parents could not recall specific precipitating events. Physical examination revealed multiple pigmented lesions involving the diaper area, trunk and four limbs. The skin lesions mainly consisted of papules each measuring 0.5-0.8 cm in diameter. There were some macular lesions which were new eruptions. Some of the old lesions may persist as papules with more dark brown colour or healed as post-inflammatory hyperpigmentation (Figure 1). Excoriations were noted. The abdomen was soft and no organomegaly was noted. There was no lymphadenopathy. Apart from the skin lesions, her general condition was good with up-to-date development. The family history was unremarkable.

A punch biopsy was performed. There were superficial perivascular and interstitial infiltrate of mononuclear small cells, having regular oval nuclei and moderately abundant pale cytoplasm (Figures 2 & 3). They were CD117 (Figure 4) and CD68 positive and S100 negative. Chloroacetate esterase stain highlights frequent red granules. Eosinophils were rare. There is mild epidermal basal hyperpigmentation. There is no cellular atypia.

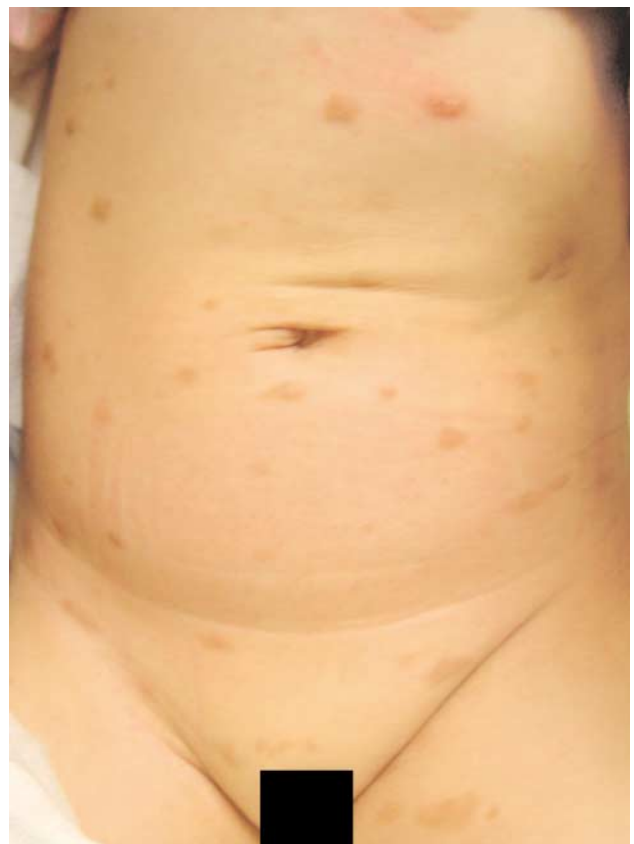


Figure 1.

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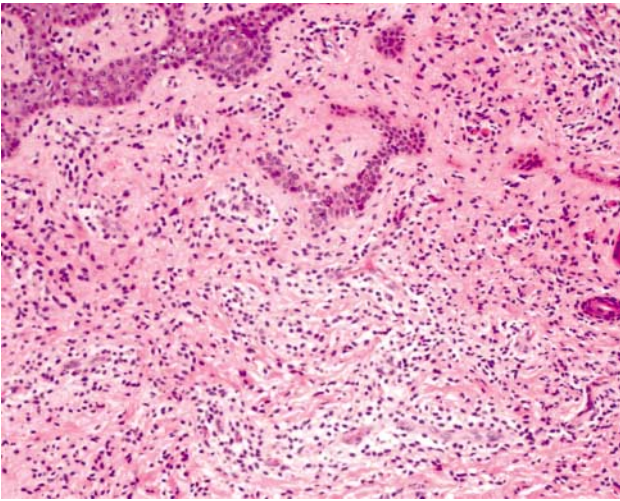


Figure 2.

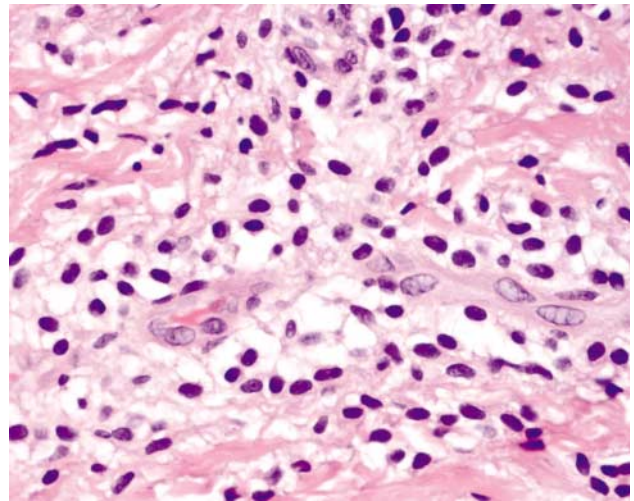


Figure 3.

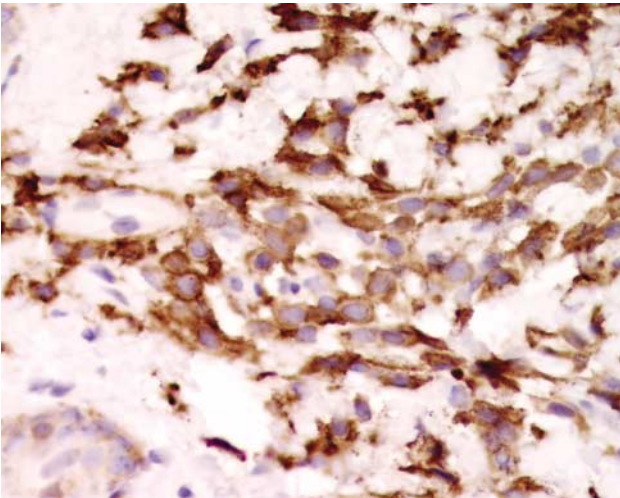


Figure 4.

Questions

1. What are the differential diagnoses and most likely diagnosis?
2. What other investigations that may be helpful?
3. A clinical sign was often mentioned for this condition. How reliable was it in making the diagnosis?
4. What is the long term outlook and what are the treatment options for this baby?

Answers to Dermato-venereological Quiz on pages xx

1. The most likely diagnosis is urticaria pigmentosa (UP). The other differential diagnoses include Langerhan histiocytosis X, juvenile xanthogranuloma, insect bite, scabies, or rarely, leukemia cutis.
2. A skin biopsy is indicated and in this case it demonstrated superficial and perivascular infiltrates of mast cells. The amount of mast cells demonstrated is related to the age of the patient (more in younger patients), the thickness of lesion (more in the nodular forms) and type and age of lesions. Other useful investigations include complete blood picture, renal and liver function tests. Bone marrow examination should be done if abnormalities found in blood picture. Blood tryptase level, 24 hour urine for histamine, histamine metabolites and prostaglandin metabolites may be helpful as well and indicate systemic involvement.
3. Darier sign (Darier's sign, Darier test) was first described by Jean Darier in 1905. It was found to be useful in early days to rule out other skin conditions like lichen planus, psoriasis, syphilitic lesions and tuberculids. However, the Darier sign may not be present in patients with UP, and there are great variations in elicibility. In fact, other urticating conditions can also present with Darier sign like urticaria, preleukemia, nodular scabies, congenital smooth muscle hamatoma and urticating Langerhans cell histiocytosis (Hashimoto-Prikzker).
4. Infants with urticaria pigmentosa will gradually have disease resolution in about 50% of cases. Systemic involvement in children is relatively rare. The mainstay of treatment will be H1 and H2 antagonists, sodium cromoglycate and topical steroid. Other novel treatments for aggressive disease in adult will include interferon-alpha, cyclosporine and nifedipine.