

Dermatology Quiz

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A 1-year-old female was presented to us with multiple itchy brownish papules and



macules measuring 1 to 2 cm in diameter over her body, which had been present since 3 months of age. The lesions started at the nape of her neck and spread downwards involving the trunk and limbs. She had been seen by various doctors but had failed to respond to topical treatments. Physical examination revealed multiple pigmented

lesions affecting the nappy area, trunk and all limbs. Excoriations were noted. The abdomen was soft with no organomegaly, and there was no lymphadenopathy. Apart from the skin lesions, her general condition was good and she had appropriate developmental milestones. The family history was unremarkable.

1. What are the differential diagnoses and most likely diagnosis?
2. What other investigations may be helpful?
3. Skin biopsy demonstrated superficial and perivascular infiltrates of mast cells. Other tests were normal. What is your final diagnosis?
4. A clinical sign is often mentioned for this condition. How reliable is it in making the diagnosis?
5. What is the long-term outlook and what are the treatment options for this baby?

(Answers on p. 123)

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Answers:

1. The most likely diagnosis is urticaria pigmentosa (UP). The other differential diagnoses include Langerhans' cell histiocytosis, juvenile xanthogranuloma, leukaemia cutis, insect bites and scabies.

2. A skin biopsy is indicated as the underlying histopathology is vital in making the final diagnosis. Other useful investigations include a complete blood picture, and renal and liver function tests. Bone marrow examination should be undertaken if abnormalities are found in the blood picture. Blood

tryptase level and 24-hour urine for histamine, histamine metabolites and prostaglandin metabolites may also be helpful and indicate systemic involvement.

3. The final diagnosis is urticaria pigmentosa (UP). The number 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 the type and age of the lesions.

4. Darier's sign (Darier sign, Darier test) was first described by Jean Darier in 1905. It was found to be useful in early days to rule out other lichenoid skin conditions, psoriasis and chronic infections such as syphilis and tuberculosis. However, Darier's 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's sign. These include urticaria, pre-leukaemia, nodular scabies, congenital smooth muscle hamartoma and urticating Langerhans' cell histiocytosis (Hashimoto-Prikzker disease).

5. Infants with urticaria pigmentosa will have gradual 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 adults include interferon-alpha, cyclosporine and nifedipine.

About the Author

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