

QUIZ SECTION

A Segmental Rash in a Young Male: A Quiz

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A 16-year-old student presented with a 4-year history of an occasionally itchy rash on his right shoulder. He subsequently developed a similar eruption on the right submental region. He was otherwise healthy with no systemic symptoms. On examination, a large segmental area of non-scaly hyperpigmented macules on the anterior shoulder extending to the chest was seen (Fig. 1A). An erythematous inflammatory plaque was seen on the submental region (Fig. 1B).

A punch biopsy was taken from both areas. The appearances were similar (Fig. 1C). Histopathological analysis showed a mild dermal perivascular inflammatory infiltrate with an increase in mast cells (45/hpf= 190 cells/mm² [normal range up to 20 hpf= 80 cells/mm²]). There was no evidence of epidermal thickening or elongated rete ridges.

Serum tryptase levels were normal.

What is your diagnosis? See next page for answer.



Fig. 1. Segmental hyperpigmented macules on (A) anterior shoulder (B) submental region. (C) The biopsy revealed a mild dermal perivascular inflammatory infiltrate with an increase in mast cells.

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ANSWERS TO QUIZ

A Segmental Rash in a Young male: A Comment

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Diagnosis: Naevoid urticaria pigmentosa

Urticaria pigmentosa (UP), a form of cutaneous mastocytosis, is a rare disease presenting with brown patches on the skin due to abnormal collections of mast cells (1). Localised itching, erythema and swelling may occur when mast cell degranulation occurs secondary to rubbing the skin or heat exposure. Systemic effects such as flushing and diarrhoea may rarely occur from systemic histamine release. Aggravating factors include systemic drugs that result in histamine release, such as aspirin, NSAIDs, codeine and morphine, in addition to alcohol and emotional stress.

UP most commonly affects infants, the first patches appearing within a few months of age on the scalp, face, trunk and limbs. In children, the condition runs an indolent course and the patches eventually fade away but in adults it tends to persist (2). Clinical examination usually reveals brown-red macules on the trunk and limbs that become itchy and swollen on rubbing (Darier's sign). Telangiectasia macularis eruptiva perstans (TMEP) is a rare variant of cutaneous mastocytosis with diffuse red patches and overlying telangiectasia. Serum tryptase levels are used as a marker of systemic mastocytosis.

Localised UP is a rare entity and has been observed in the context of radiotherapy and microinjections. There has only been one report of spontaneous localised (naevoid) UP following Blaschko's lines in a 6-year-old child (3). The pathogenesis of UP is not fully elucidated. Activating mutations of the C-kit proto-oncogene, a transmembrane protein that binds to mast cell growth factor and promotes

cells division, have been shown to result in abnormal proliferation of mast cells (4). Interleukin 6 and BCL-2 have also been implicated in the pathogenesis (5, 6).

Although there is no curative treatment, antihistamines and H2-receptor blockers may provide symptomatic relief for histamine-induced symptoms. PUVA and to a lesser extent UVB may improve the cosmetic appearance and also slowly decrease the number of mast cells in the skin. Laser treatment may also be used to improve the cosmetic appearance.

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