

Case Report

Adult onset urticaria pigmentosa: a case report

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Abstract Urticaria pigmentosa is a fairly indolent form of cutaneous mastocytosis, which is more prevalent in infants than in adults. Adult onset disease is usually supposed to be associated with systemic disease and has a propensity for polycythemia vera and leukaemia in a certain percentage, though regression has been reported in as many as 19% cases. A useful clue to diagnose indolent forms from malignant forms is that invariably there is thrombocytopenia with leukocytosis in malignant forms. We report an adult female with typical lesions of urticaria pigmentosa, proven by skin biopsy, who showed a good response to H1 and H2 receptor blockage treatment.

Key words

Mastocytosis, urticaria pigmentosa, histamine, adult-onset

Introduction

Urticaria pigmentosa belongs to the category of mastocytosis disorders, where the skin is infiltrated with increased amount of mast cells. (Table 1). It is a disease of infancy and childhood, with more than 50% cases being reported in children.

Case report

A 46-year old lady presented to us with a 6 year history of brownish patches occurring all over the body, which used to itch and become raised occasionally, especially after pressure or exposure to heat. The lesions had arisen insidiously to reach the present density. She had no systemic complaints and there was no history of any childhood skin problems. On examination she had multiple discrete tan to brown-colored macules on her body ranging in size from 0.5-1 cm scattered over neck, (Figure 1) chest, abdomen (Figure 2) trunk, upper limbs, thighs and legs. On friction with a

Table 1 Different clinical forms of urticaria pigmentosa [1]

Maculopapular
Telangiectasia macularis eruptiva perstans
Diffuse bullous
Diffuse erythrodermic
Diffuse doughy infiltrative
Solitary mastocytoma

rubber eraser, individual macules became erythematous and edematous developing into weals within a time of 5 minutes (Figures 1 and 2) and there was severe itching. We provisionally made a diagnosis of urticaria pigmentosa and investigated her further. A complete blood count, liver and renal function tests, and ultrasonography of the pelvis and abdomen were normal. She refused urinary and plasma 5 HIAA level estimation due to the high cost thereof. A 4 mm skin punch biopsy specimen was obtained with due precautions and sent for histopathological examination and for special stains including toluidine blue. It revealed a hyperkeratotic epidermis with sparse perivascular mononuclear inflammatory infiltrate in the upper dermis composed of lymphocytes and mast cells with oval to spindly nucleus and eosinophilic cytoplasm. On toluidine blue stain, positive granules could be demonstrated, which confirmed our diagnosis (Figures 3, 4). As she had no severe

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Figure 1 Neck lesions - brownish pigmented macules and slightly raised papules along with wheals produced by rubbing with an eraser



Figure 2 Brownish pigmented macules on the anterior abdomen

symptoms and associated systemic complaints she was treated with H1 and H2 receptor blockers (oral cetirizine 10 mg daily along with oral ranitidine 150 mg twice daily). She has achieved good control of itch as well as wheals with the above measures.

Discussion

Mastocytosis is a group of conditions of mast cell proliferation which result from specific missense mutations in the proto-oncogene *c-kit*, which encodes for mast cell growth factor and stem cell factor.² The clinical features were first described by Tay and Nettleship in 1869, eight years before Ehrlich discovered the mast cell.³ Urticaria pigmentosa

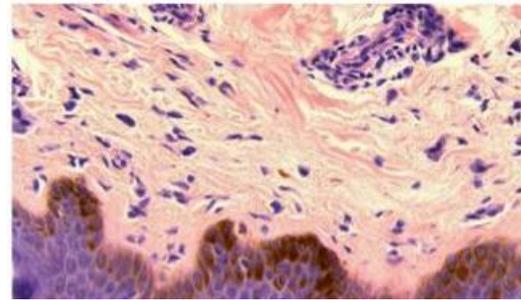


Figure 3 Histopathology from the lesion 40X view showing slightly hyperkeratotic epidermis and sparse mononuclear inflammatory infiltrate in the upper dermis composed of lymphocytes and mast cells with oval to spindle nucleus and eosinophilic cytoplasm.

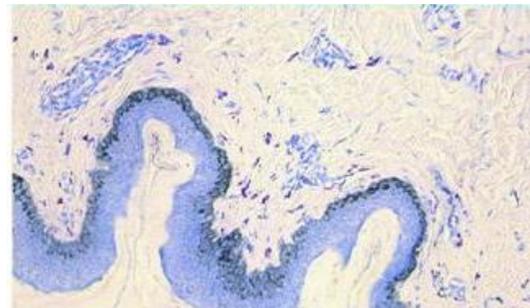


Figure 4 Toluidine blue stained section, 10X view showing the spindle shaped mast cells which have taken up the stain.

is the most common mastocytosis syndrome in both children as well as in adults, and accounts for over 90% cases of indolent mastocytoses.⁴ More than 50% cases occur in children below 2 years of age, and in about 90% of patients no clinical evidence of systemic involvement is detectable.⁵ The condition manifests as intensely itchy monomorphic yellow tan to reddish brown pigmented macules, papules and nodules in a widespread distribution which become itchy, raised and reddish due to local wheal formation on gentle rubbing (Darier's sign).

In patients with cutaneous involvement the only confirmatory diagnosis is skin biopsy, whereas in those with suspected internal involvement bone marrow and liver biopsies may be additionally performed. Elevated levels of mast cell degranulation products like histamine and methyl imidazole acetic acid,

plasma tryptase and PGD2 levels are all markers of mastocytosis.² Histologically there is mild to moderate perivascular infiltration with dendritic mast cells in the papillary dermis; a band like infiltrate or sometimes even nodular infiltrates extending to the subcutis may be seen, especially with special stains like toluidine blue and chloracetate esterase.⁴ In general, urticaria pigmentosa associated with systemic mast cell disease cannot be clinically differentiated from urticaria pigmentosa with purely skin involvement. Travis *et al.* suggest that the presence of dense infiltration by mast cells in urticaria pigmentosa with cytologic atypia may correlate with the presence of systemic mast cell disease.⁶ There are reports of fatal mast cell leukemia presenting as urticaria pigmentosa.⁷ A clue to malignant mastocytosis is a lower erythrocyte sedimentation rate and thrombocytopenia along with leukocytosis. In view of this, at initial presentation, along with a detailed history and physical examination, only liver function tests, complete blood count and skin biopsy is warranted in a patient without any systemic complaints.¹

Treatment is mainly symptomatic with avoidance of all known mast cell degranulation products (morphine, codeine, acetyl salicylic acid, radiograph contrast dyes, scopolamine, d-TC). For uncomplicated urticaria pigmentosa a combination of H1 and H2 receptor blockers, mast cell stabilizers or even PUVA may be tried. Higgins *et al.* suggest intermittent application of topical steroids as a cheap and effective alternate treatment modality in adult onset urticaria pigmentosa.⁸

Adult onset disease has a relatively worse prognosis with a small, but significant risk of occurrence of polycythemia vera or monoclastic or myelocytic leukaemia,³ though regression has been reported in 7-19% of cases.⁹

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