

A case of urticaria pigmentosa

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Abstract

Urticaria pigmentosa is the most common variant of cutaneous mastocytosis. A three and a half-month-old female child presented with generalized eruption of multiple brownish macular and papular lesions on the trunk, limbs, face and neck of two months duration. On stroking the individual lesion, there was formation of wheal and erythema (Darier's sign positive). Histopathological examination of lesional skin and staining with Giemsa stain and hematoxylin eosin showed infiltration of numerous spindle shaped mast cells in perivascular and periappendageal location in the upper dermis. These findings were suggestive of mastocytosis. There was no systemic involvement. We present a typical case of urticaria pigmentosa."

Key words

Mastocytosis, Darier's sign, Giemsa stain, mast cells, urticaria pigmentosa.

Introduction

Urticaria pigmentosa is the most common variant of cutaneous mastocytosis. Mastocytosis includes urticaria pigmentosa, mastocytoma, telangiectasia macularis eruptiva perstans, diffuse cutaneous mastocytosis and systemic mastocytosis. Mastocytosis is uncommon in any form.¹ In about 70-75% of cases, lesions of mastocytosis are evident before the age of 2 years.² We are presenting a typical case of urticaria pigmentosa.

Case Report

A three and a half-month-old female child presented with generalized eruption of multiple brownish macular and papular lesions on the trunk, limbs, face and neck of two months duration (**Figure 1** and **2**). There was no history of diarrhea, fainting episodes and bulla formation. There was no family history

of similar disease and the general health, growth and milestones of the child were within normal limit.

On examination, there were multiple hyperpigmented macular and papular lesions on the face, neck, trunk and limbs. Few brownish plaques were also seen. The mucosae, palms, soles and groins were spared. On stroking the individual lesion, there was formation of wheal and erythema (positive Darier's sign). There was no systemic involvement. Histopathological examination of lesional skin and staining with giemsa stain and hematoxylin eosin showed infiltration of numerous spindle shaped mast cells in perivascular and periappendageal location in the upper dermis (**Figure 3** and **4**). Complete blood count, liver and renal functions, urinalysis and chest roentgenogram were normal. Lesions regressed with topical application of mometasone furoate ointment 0.1% applied twice daily along with administration of hydroxyzine, 7 mg thrice daily for four weeks. Necessary advice to parents was given regarding avoidance of excessive scrubbing and massage of the skin.

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Figure 1 Clinical photograph showing the distribution of hyperpigmented macules on the body.

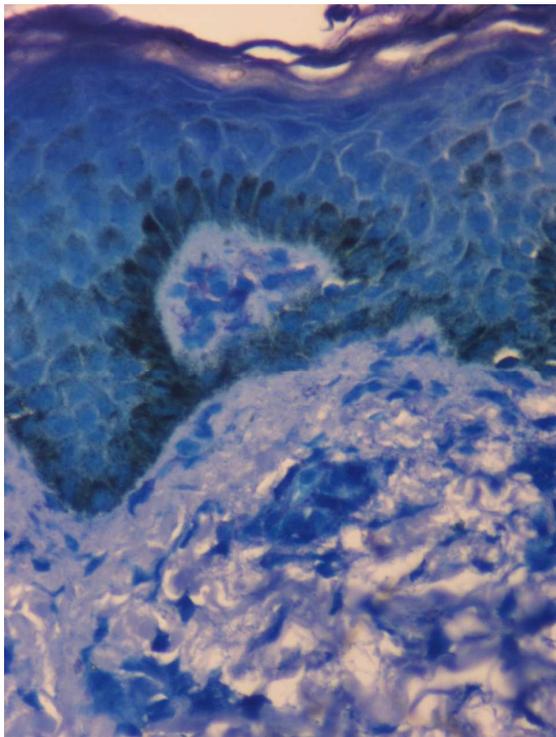


Figure 3 Histopathological image showing the spindle shaped mast cells in the upper dermis (Giemsa, 40 X)



Figure 2 Clinical photograph showing the brownish macules on the trunk.

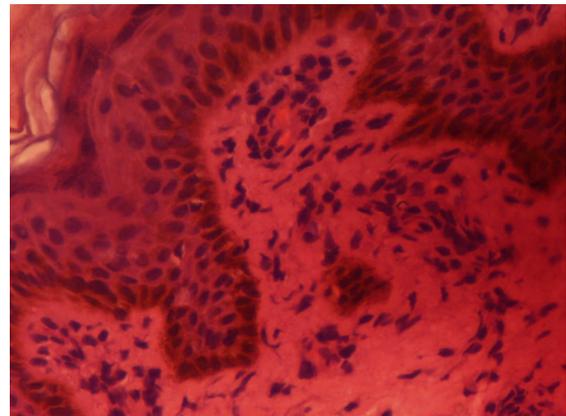


Figure 4 Histopathological image showing the spindle shaped mast cells in the upper dermis (H & E stain, 40 X)

Discussion

Urticaria pigmentosa (UP) is a type of cutaneous mastocytosis characterized by aggregates of mast cells in the dermis, leading to the development of dark yellow to brown macules.³ Mastocytosis encompasses a spectrum of disorders that range from a solitary cutaneous nodule to diffuse infiltration of skin with involvement of other organs. UP is a common type of mastocytosis in children and is considered a hyperplastic, rather than a neoplastic, disorder.^{3,4}

Numerous reddish-brown or yellow-tan monomorphic maculopapules, plaques or nodules appear on the skin of the body. The palms, soles, face and scalp may be free of lesions.⁵ Lesions urticate within a few minutes of gentle rubbing (Darier's sign).⁵ On histopathology of skin biopsy, there are

increased numbers of mast cells in the dermis. The mast cells are oval or spindle-shaped and are well demonstrated by Giemsa stain. Mast cell infiltrates are predominantly found around blood vessels and skin appendages in the papillary dermis. In UP, mast cells may be increased up to 15-fold above normal.⁶

Systemic involvement is rare in UP. Most lesions of UP resolve spontaneously in adolescence.⁷ UP has a good prognosis.

Our patient had the typical clinical features and the skin biopsy also demonstrated increased mast cells so a diagnosis of UP was confirmed.

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