Ind J Dermatol Venereol Leprol 1996; 62 : 175-7

CONGENITAL MULTIPLE CUTANEOUS MASTOCYTOMA

V K Somani, V K Sharma, V N V L Sita, Fatima Razvi, V Sucharita,

A rare case of multiple cutaneous mastocytoma presenting at birth with multiple skin coloured to hyperpigmented papulonodules and plaques all over the body is being reported.

Key Words : Mastocytoma, Congenital

Introduction

Mastocytosis is a condition characterised by mast cell hyperplasia in the bone marrow, liver, spleen, lymph nodes, GIT and skin. Clinically, the diseases is often accompanied by the evidence of mast cell activation which includes pruritus, flushing, urtication, abdominal pains, vascular instability and neuropsychiatric difficulties.

Cutaneous mastocytosis is an uncommon disease. The clinical types seen are solitary mastocytoma, urticaria pigmentosa,diffuse cutaneous mastocytosis and telangiectasia macularis eruptiva perstans. Other unusual cutaneous mastocytosis reported are erythrodermic mastocytosis[1] mimicking staphylococcal scalded skin syndrome[2] and familial mastocytosis.[3]

Cutaneous mastocytosis generally presents during the first 2 years of life. The most common manifestation is a solitary mastocytoma followed by urticaria pigmentosa. The most common symptoms are pruritus, bullae-formation, GIT bleeding, flushing. There is a tendency for regression with age. Darier's sign ie urtication on stroking the lesion is usually present.

From the Department of Skin and VD,Deccan College of Medical Sciences, Princess Esra Hospital, Shah Ali Bunda, Hyderabad, India.

Address correspondence to : Dr V K Somani "Sanskar" 2-1-438/A, Nallakunta, Hyderabad-500044.

Case Report

One month old infant was brought to the skin department with the history of multiple swellings on the body since birth. Patient was 3rd sibling born after full term normal delivery to non-consanguineous parents. The previous siblings were healthy. Mother did not give history of any drug intake or fever during the pregnancy.

The infant had multiple skin coloured to hyperpigmented papulo-nodules and plaques over the extremities, scalp, face, trunk and buttocks. The consistency was firm. Some of the lesions regressed leaving behind hyperpigmented macules and patches. The Darier's sign was negative. There were no mucous membrane lesions or lymph-adenopathy or organomegaly (Fig. 1).

Routine laboratory investigations like haemogram, complete urine examination, VDRL in both mother and infant and X-Ray chest were normal. Ultrasound abdomen was also within normal limits.

Biopsy of a nodule revealed papillary dermis filled with a cellular infiltrate composed of round to oval cells arranged in sheets extending through the entire dermis and even in the subcutaneous fat at some places (Fig. 2). Alcoholic toluidine blue stain showed metachromatic granules thus identifying the infiltrate to be composed mainly of mast cells and confirming the diagnosis of mastocytoma.

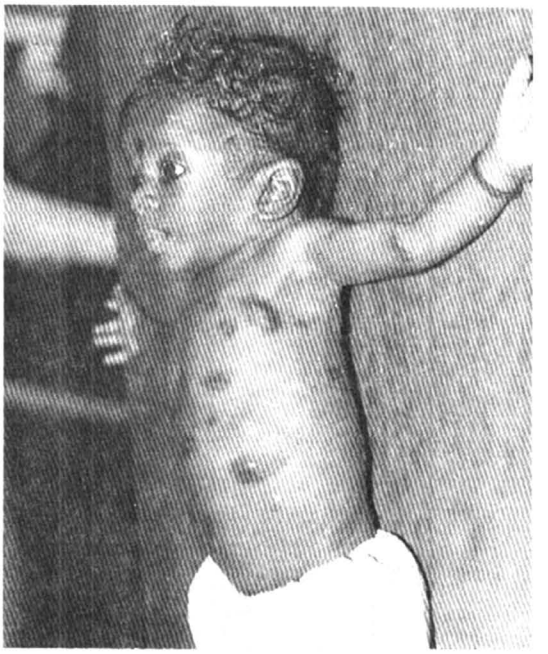

Fig. 1. Showing multiple papulo-nodules and plaques all over the body.

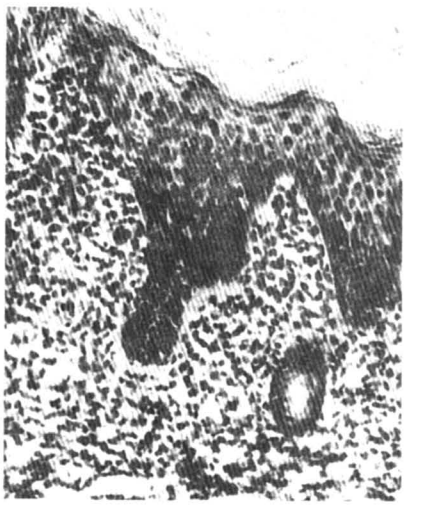

Fig. 2. Showing dense mast cell infiltrate in the dermis. (Toluidine Blue x 100)

Discussion

Presence of papulo-nodules and plaques at birth is quite rare. The differential diagnosis considered at the time of presentation included juvenile xanthogranuloma, sub-cutaneous fat necrosis, cutaneous mastocytosis, congenital leukemia and Blueberry-Muffin syndrome.

Juvenile xanthogranulomas are usually solitary or micronodular, if multiple. Plane of the lesions and softish consistency was against sub-cutaneous fat necrosis. Congenital leukemia and Blueberry-Muffin lesions reveal a purpuric component in the lesions.

About 15% of patients with cutaneous mastocytosis present with localised lesions. These are red, pink or yellow nodules, usually solitary but occasionally multiple, which appear in infancy or early childhood. Lesions usually weal when rubbed and may blister, although occasionally symptomless.[4] This, to the best of our knowledge, is first report of multiple mastocytoma presenting at birth.

Though, Darier's sign is pathognomonic of this condition and is regularly present, some patients have little or no wealing or itching even when skin reveals a dense population of mast cells.[5] The case is being presented for the rarer presentation of non-urticating multiple nodules and plaques present at birth.

References

1. Requena I. Cutis 1992;49 : 189-91.
2. Oranje AP, Soekanto W, Sukari A, et al. Paediatr Dermatol 1991; 8: 147-51.
3. Anstey A, Lown DG, Kirby JD,et al. Dermatol 1991; 125: 583-7.
4. Johnson WC, Helwig EB. Solitary mastocytosis (urticaria pigmentosa). Arch Dermatol 1961; 84: 806-15.

5. Beare M. The pathology of the mast cells in man. In: Rook AJ, Walton GS, eds. The comparative physiology and pathology of the skin. Oxford: Blackwell, 1965: 401.