

A Case Report : Pseudoxanthomatous Diffuse Cutaneous Mastocytosis in A Newborn



Medical Science

KEYWORDS : PSEUDOXANTHOMATOUS DIFFUSE CUTANEOUS MASTOCYTOSIS FEBRILE SEIZURES, Newborn

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INTRODUCTION-

Cutaneous mastocytosis is a rare disorder. Clinical varieties are urticaria pigmentosa, mastocytoma, diffuse cutaneous mastocytosis, and telangiectasia macularis eruptiva perstans in order of decreasing frequency of occurrence.¹ Diffuse cutaneous disease has two clinical variants: bullous and pseudoxanthomatous or xanthelesmoid types.²

Pseudoxanthomatous mastocytosis was first described by Tilbery in 1875.³ It is characterized by multiple, pale, flat, yellowish papules and nodules, varying in size from 1 mm to 2 cm.⁴ This multinodular nonpigmented variety of mastocytosis is extremely rare. In this variety, as against a classic Darier's sign, only erythema without urtication is elicited by rubbing⁵

CASE REPORT: A 9 day old female child was brought by relatives to the paediatric OPD with complaints of elevated lesions all over the body since birth which was progressive in nature. The baby was referred to the dermatology opd for the cutaneous lesions. There were no other associated complaints and no significant antenatal or natal history. She had no family history of any dermatological diseases.

Picture 1

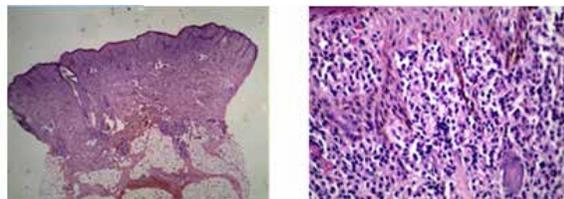


Examination revealed multiple papules and nodules all over the body with diffuse erythematous rash. Baby had increased skin markings with transverse striations over the abdomen. Palms and soles were also involved. Rest of the general examination was normal. Systemic examination revealed no abnormalities.

Hematological and biochemical investigations were within normal limits.

Histopathology of skin punch biopsy revealed dense diffuse infiltrate of numerous mast cells filling up the papillary dermis and most of the upper and mid reticular dermis, moderate epidermal hyperplasia and hyperpigmentation in the overlying epidermis, sparse scattering of eosinophils within the infiltrate and extracellular mast cell granules these findings are suggestive of pseudoxanthomatous diffuse mastocytosis.

Picture 2& 3



Baby was given with anti histaminics and corticosteroids.

DISCUSSION-

Cutaneous mastocytosis is a rare disorder. Clinical varieties are urticaria pigmentosa, mastocytoma, diffuse cutaneous mastocytosis, and telangiectasia macularis eruptiva perstans in order of decreasing frequency of occurrence. Diffuse cutaneous disease has two clinical variants: bullous and pseudoxanthomatous or xanthelesmoid types. Pseudoxanthomatous mastocytosis was first described by Tilbery in 1875. It is characterized by multiple, pale, flat, yellowish papules and nodules, varying in size from 1 mm to 2 cm. This multinodular nonpigmented variety of mastocytosis is extremely rare. In this variety, as against a classic Darier's sign, only erythema without urtication is elicited by rubbing. Skin biopsy reveals dense dermal infiltrate of mast cells. Diagnosis is confirmed on the basis of clinical features and characteristic histopathological findings.

Treatment options include antihistamines, avoidance of trigger factors, topical or systemic corticosteroids, mast cell stabilizers, PUVA therapy, and interferons. The prognosis depends on the age of onset and systemic involvement.⁶ Earlier age of onset and absence of systemic involvement are suggestive of better outcome. Childhood disease may resolve spontaneously. With advancing age, there is risk of extracutaneous involvement and of development of malignancies.⁵

CONCLUSION: The reported case has all the features suggestive of Pseudoxanthomatous diffuse cutaneous mastocytosis. It has not yet been reported to be present since birth.

REFERENCE

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