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KERATOLYSIS EXFOLIATIVA CONGENITUM

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A middle aged male had keratolysis exfoliativa congenitum (continual skin peeling), since his infancy. None of his family members was affected by a similar skin disease. Mild rubbing of the skin with fingers caused rapid peeling of the horny layer. Oral vitamin A, 50,000 units daily for two months and topical liquid paraffin were ineffective in controlling the desquamation.

Key words : Keratolysis exfoliativa congenitum, Familial continual skin peeling, Skin shedding, Deciduous skin.

Keratolysis exfoliativa congenitum is a rare disorder characterized by the continual shedding of the entire stratum corneum in sheets.[1] It has been reported under several diagnostic terms : familial continual skin peeling,[2] deciduous skin,[3] keratolysis or skin shedding,[4] and peeling skin syndrome.[5,6] It is characterized by asymptomatic, chronic shedding of the horny layer of the skin affecting the face, trunk and limbs. The disease usually starts during infancy and persists throughout life. The exact cause of this disorder is not well understood. Familial cases may occur, and an autosomal recessive mode of inheritance has been suggested.[2] We report a case of keratolysis exfoliativa congenitum seen by us.

Case Report

A 40-year-old male developed asymptomatic peeling of his skin since infancy. The peeling had been constant throughout his life and affected his face, trunk and limbs. Sheets of horny layer of various sizes were shed constantly. There was no seasonal variation. There was no parental consanguinity and none in his family suffered from a similar skin disease or ichthyosis. Examination revealed multiple sheets of dry horny layer of various sizes getting detached from the surface of the skin. Mild rubbing of the skin with the fingers caused peeling of the horny layer easily (Fig. 1). It did not leave any erosion or caused oozing of the serum. There was no associated erythema or ichthyosis. The hairs and nails were normal. Oral and genital mucosae appeared normal.

Routine laboratory tests on blood, urine and stools were normal. Examination of the scales in 10% KOH did not show any fungal filament. Histopathology of the skin biopsy was unremarkable apart from the stratum corneum which showed hyperkeratosis with a loose structure. Oral vitamin A, 50,000 in daily given for 2

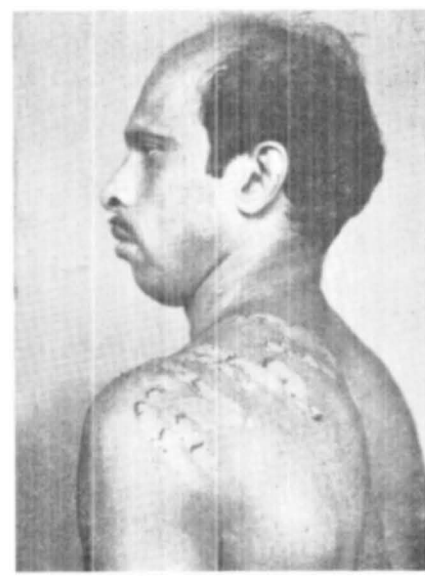

Fig. 1. Multiple sheets of horny layer of various sizes getting detached from the surface of skin following gentle rubbing.

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months and topical application of liquid paraffin were ineffective in controlling the desquamation.

Comments

The clinical features of keratolysis exfoliativa congenitum are so characteristic that elaborate laboratory tests are seldom required for its diagnosis. In the cases reported by Kurban and Azar, 4 of 9 siblings in a family were affected and they used the term familial continual skin peeling for this condition.[2] Levy and Goldsmith considered this as a form of congenital ichthyosis. The cases reported by them had associated pruritus, short stature and easily removable anagen hairs.[5] In our patient these features were absent. Abdel-Hafez et al noted decreased desmosomal attachments in the areas of the split in the epidermis, in addition to disruption and fragmentation of the intercellular lamellar material.[7] The loss of cohesion may be attributed to either or both these factors, but the underlying cause of this disturbance is unknown.[6]

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