CASE REPORT

Peeling Skin Syndrome

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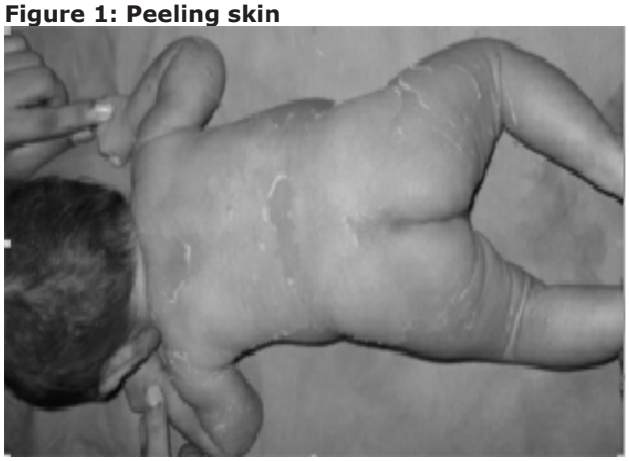
Abstract: A 25 days old male child with peeling of the skin without aminoaciduria is presented.

Keywords: Peeling skin syndrome, Emollient

Introduction: Peeling skin syndrome is a rare autosomal recessive disorder characterized by asymptomatic, generalized, non-inflammatory exfoliation of the stratum corneum (1). It is a rare entity and only few cases have been reported from India (1). It is also named as keratolysis exfoliativa congenita (2), deciduous skin (3), familial continual skin peeling (4) and peeling skin syndrome (5). We report one case of this rare entity.

Case report: A 25 days old male child born of non consanguineous marriage, the second child of the family born as a full term normal vaginal delivery presented with peeling of the skin since 4th day of birth. Mother noticed that the asymptomatic peeling of skin started at the flexures of the upper and the lower extremity and later involved the trunk. There was no history of similar complaints in the family. General and systemic examination was normal. On examination the peeling was present on trunk, extremities with sparing of face, palms and soles, leaving behind non oozing mildly erythematous base. [Fig 1] Nails, hairs, oral cavity and genitals were normal. There was no aminoaciduria. He was treated with emollients after which there was symptomatic relief but on follow up there was recurrence of lesions.

Figure 1: Peeling skin



Discussion: The peeling skin syndrome is an autosomal recessive disorder characterized by lifelong peeling of the stratum corneum. The cause of this disorder is unknown. The defect is reduced adherence of abnormally thick stratum corneum to the stratum granulosum. (6) The skin barrier function and cohesion get disturbed due to abnormal deposition of lipids, which leads to the desquamation of stratum corneum.(7) It may be associated with easy pluckability of hair, shedding of nails; hypogonadism and anosmia (8), low plasma tryptophan levels and aminoaciduria. (9) None of these features were present in our case. Troupe has pointed out that there are two types of peeling skin syndrome. The first was first described by Fox in 1921, and is characterized by shedding of the skin including the face, comparable to the shedding of skin by reptiles. The peeling occurs in the lower part of the stratum corneum. The second type, first described by Wile in 1924, presents with erythroderma at birth, and is associated with features such as growth retardation, and aminoaciduria. Our case is probably of first type but without face and palm and soles involvement.

Emollients such as petrolatum jelly and salicylic acid may be helpful with minimal immersion in water. Drugs such as vitamin A, tretinoin and etretinate, tar, Methotrexate and UVB phototherapy have been tried.

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