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CONTINUAL SKIN PEELING SYNDROME

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Two cases of continual skin peeling syndrome (CSPS) are presented

Key Words: Continual skin peeling syndrome

Introduction

Continual skin peeling syndrome (CSPS) is a rare autosomal recessive disorder characterised by asymptomatic, generalized, non-inflammatory exfoliation of the stratum corneum.[1] Different nomenclatures have been used in the literature, namely keratolysis exfoliativa congenita,[2] deciduous skin,[3] familial continual skin peeling,[4] peeling skin syndrome,[5] etc. We are reporting 2 cases of this rare entity.

Case Reports

Case 1: A 55-year-old Hindu male presented with thickening and peeling of the skin over extensor aspect of both hands, feet, back of the elbow and knee since childhood. Patient reported that his skin over the above sites thickens and peels off in sheets continuously throughout the year but skin over the remaining parts of the body peels off during summers only, which is normal in winters.

No family history of any dermatosis could be elicited. Patient had used emollients, keratolytics, vitamins A and E in past with partial relief. Patient was otherwise healthy.

On cutaneous examination, bilaterally symmetric well defined hyperkeratotic plaques were present on the dorsal aspect of hands, feet, elbow and knees. Rubbing of skin with pressure on these sites induced peeling; large sheets of skin without bleeding and pain leaving behind shiny erythematous area. The skin of remaining body showed fine scaling which could be removed easily. Palms, soles, scalp, hair, teeth, eyes, nails and mucous membranes were normal. Histopathological examination revealed orthokeratotic hyperkeratosis with separation within stratum corneum. Remaining epidermis and dermis was normal.

Case 2: A 35-year-old Hindu male presented with thickened skin over the back of both hands and feet. The skin at above sites got macerated and peeled off during summer or by dipping in water leaving behind glazed erythematous area without any bleeding, oozing or pain. There was no family history of any dermatological disorder. On cutaneous examination, bilaterally symmetric hyperkeratotic plaques were present over dorsum of hands and feet. Remaining skin of body was normal. No other dermatological and systemic abnormality was present.

Discussion

CSPS is a rare dermatological entity and only few cases have been reported from India.[1] The cause of this interesting disorder is not known. Kurban and Azar[4] on the basis of autoradiography suggested CSPS to be a hyperproliferative state. Silverman and Ellis[6] found that level of cleavage in CSPS is

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within the stratum corneum. According to them, the barrier function and cohesion got disturbed due to abnormal deposition of lipids, which leads to the desquamation of stratum corneum. The exact events and nature of lipids have to be explored.

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