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# SKIN PEELING SYNDROME

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We are reporting a case of skin peeling syndrome, a rare disorder in which sudden generalized exfoliation of the stratum corneum occurs. Histologically, there was well formed subcorneal pustule filled with polymorphs and nuclear dust, considering this to be a varient of subcorneal pustular dermatosis, we have put the patient on Dapsone.

**Key Words : Skin peeling syndrome.**

## Introduction

Skin peeling syndrome is a rare disorder characterized by generalized peeling of the stratum corneum. We are reporting a case of skin peeling syndrome with our views about the probable pathogenesis.

## Case Report

A 10-years-old boy was seen for peeling of skin, fissuring of palms and soles. As narrated by the parents, this disorder started when the child was 6 months old. They noticed thickening of skin over palms, soles, elbows, followed by a vesiculopustular rash over the sides of neck, axillae, groins and proximal parts of the extremities. This rash was preceded by a fall in apetite and a bout of fever. These pustules ruptured leading to peeling of skin in sheets with foul smelling serous discharge. Peeling was more pronounced after bath. This was associated with soreness of oral cavity due to presence of erosions. Minimal continual peeling was followed by an episode of acute generalized peeling, approximately once a year. In a few episodes he had lost the nails also.

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This patient was earlier treated with high doses of vitamin A, antibiotics, oral and topical steroids, keratolytics and emollients, which failed to arrest the peeling process. The past medical history was unremarkable. No family history of skin peeling,ichthyosis or other dermatoses could be elicited. The child was borne of non-consanguineous marriage.

The child was otherwise in good health, active and intelligent, though aggressive and angry. Systemic examination did not reveal any abnormality.

Dermatological examination revealed bilaterally symmetrical peeling of the skin (Fig.1) leaving behind hypopigmented areas.

Fig. 1. Bilaterally symmetrical peeling of skin leaving behind hypopigmented areas. Brittle, loosely attached nails with subungual hyperkeratosis.

Flaccid vesiculo-pustules with crusts were seen over the sides of the neck (Fig.2), axillae,

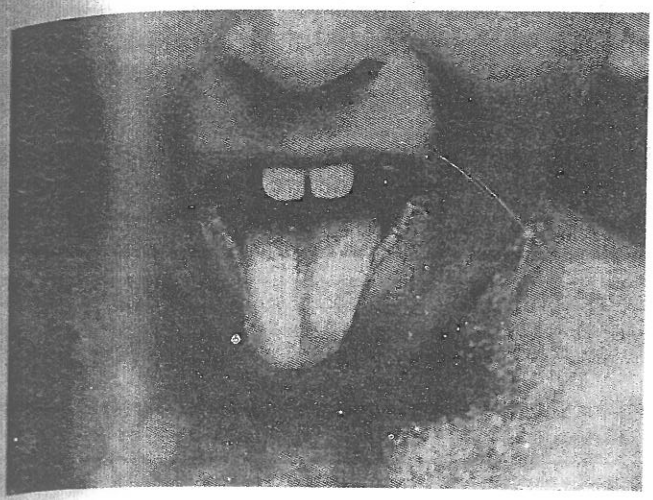

Fig. 2. Pustules with crust over the left shoulder, erosions over the tongue.

groins and other skin folds. Palms and soles were hyperkeratotic with peeling and cracks. Nails were brittle loosely attached to the nail folds, with subungual hyperkeratosis. Scalp and face were relatively spared. Mild rubbing of the skin easily caused peeling. Oral mucosa and angles of mouth showed ill defined erosions. Cervical and inguinal lymph glands were palpable, discrete, firm and non-tender.

The results of laboratory investigations were normal. there was no yield on pus culture. Skin biopsy was taken from a pustule over the left groin. The stratum corneum showed basket-weave hyperkeratosis. There was a well defined subcorneal pustule filled with polymorphs and nuclear dust (Fig.3). In the papillary dermis, infiltrate made up of lymphocytes and polymorphs was seen.

## Comments

Different nomenclatures have been used for this rare disorder in the literature, namely; deciduous skin[1], familial continual skin peeling[2], skin shedding[3], the peeling skin syndrome[4], etc. This is thought to be an autosomal - recessive disorder.[4]

Various authors have put forward various theories of pathogenesis. Azar and Kurban[2] considered this to be a proliferative disorder with abnormal keratinization. Silverman et al[5] are of the view that the pathological changes in this disorder are localized to the stratum corneum. They believe that this disorder is related to intercellular deposition of abnormal lipids which leads to decreased cohesiveness of the stratum corneum.

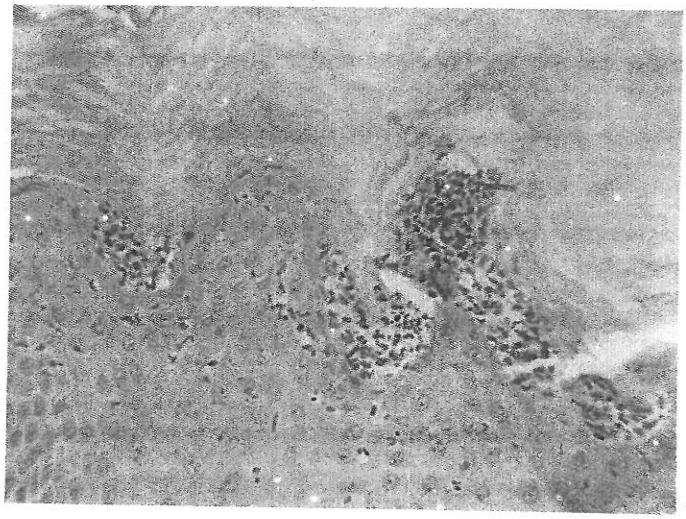

Fig. 3. Photomicrograph of biopsy from a fresh pustule showing basket-weave hyperkeratosis and a well defined subcorneal pustule filled with polymorphs and nuclear dust.(H&Ex400)

All the authors agree with the histological picture of loose hyperkeratosis and subcorneal splitting, but presence of well formed subcorneal pustule has not been documented in any of the case reports. Our case shows presence of subcorneal pustule probably because we have taken the skin biopsy from the earliest presenting lesion, i.e. a pustule. We therefore, think that this could be a varient of subcorneal pustular dermatosis. Our case also showed involvement of the oral mucosa, which was not present in any of the documented case reports.

With subcorneal pustular dermatosis in mind, we have put him on Dapsone 1.5 mg/ kg. Patient responded and the parents

observed that the present episode was of shorter duration compared to several previous episodes. We continued the Dapsone at 1mg/kg dose. There was no major episode during the last 6 months.

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