

Case Report

Acral peeling skin syndrome

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Received: 14 March 2015

Accepted: 21 April 2015

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ABSTRACT

Acral peeling skin syndrome is a rare, congenital subtype of peeling skin syndrome affecting the dorsal and volar aspects of hands and feet. The cutaneous manifestations consist of lifelong, episodic or continuous, painless, non-inflammatory peeling of dorsal and volar aspects of hands and feet. A biopsy specimen shows hyperkeratosis and splitting of the epidermis between the granular layer and the stratum corneum. This chronic disease has no known effective established treatment.

Keywords: Acral peeling, Peeling skin syndrome

INTRODUCTION

Peeling Skin Syndromes (PSS) are rare entities, inherited as autosomal recessive pattern. They are of two types, namely Acral and Generalized peeling syndrome. The Peeling skin syndromes are clinically characterized by an asymptomatic, periodic or continuous exfoliation of the superficial skin owing to a separation of the stratum corneum from the stratum granulosum at the subcorneal level.^{1,2} Acral peeling syndrome is a congenital entity. The peeling of the skin commences on the palms shortly after birth and is lifelong. The peeling can be episodic or continuous, painless and non-inflammatory and is confined to the palmar and dorsal aspects of the hands and feet. It is provoked by immersing the affected parts in water, mechanical trauma or humidity and may be more severe in summers.³⁻⁶ Occasional painful blisters may be present. There is no definite treatment and remissions can be achieved by avoidance of water immersion, use of emollients, topical steroids, keratolytics and calcipotriol. Oral methotrexate and retinoids have been tried without success.

CASE REPORT

A 1.5 year old male child presented to our department with insidious onset of peeling of the superficial layers of skin of volar surface of hands (Figure 1, 2) and feet since 4 months of age.



Figure 1: Superficial peeling of palmar surface of hands.



Figure 2: Superficial peeling with erythema.

The informant was mother and history was reliable. The peeling started on the hands and gradually within a time period of one month involved the feet (Figure 3).



Figure 3: Superficial peeling involving the plantar aspect of feet.

It was present consistently without any seasonal variation. Erythema and peeling increased with immersion of involved areas in water and the patient would peel off the skin consistently with his hands. History of mild pruritus was present. There was no pain or bleeding of the hands and feet. There were no constitutional symptoms associated. Weight of the child was 11 kg. Height was 79 cm. Milestones were normal. Immunization status was complete till date. Dermatological examination revealed that the volar surfaces of hands and feet were erythematous with exfoliation of the superficial layer of skin at multiple areas of palms and soles. Few excoriation marks significant of pruritus were present. No bleeding, tenderness, oozing, ulceration or scarring was seen. Generalized xerosis was present all over body with no other abnormality. The mucosae were unremarkable. No other systemic involvement was seen. Investigations revealed HB 10.7 gm/dl. TLC 8600 per cubic mm. N55, L30, E15, M00. Platelet count 3.8 lakh/cumm. Urine re: shows 1-2 pus cells. LFT and the lipid profile were within normal limits. Skin biopsy (H&E staining at 40x) showed hyperkeratosis and splitting of the epidermis between the granular layer and the stratum corneum

(Figure 4) Child was exhibited topical calcipotriol 0.005%.

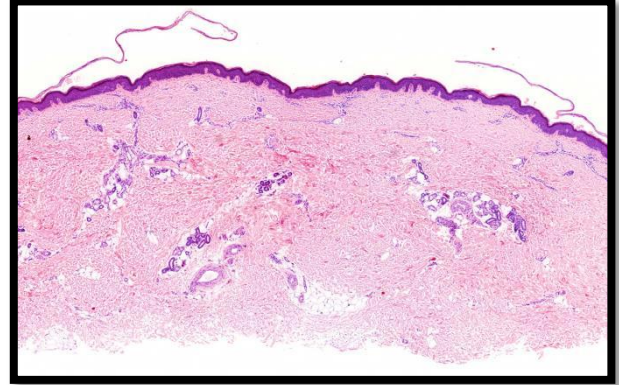


Figure 4: Histopathology – showing hyperkeratosis with intraepidermal split.

DISCUSSION

Congenital acral peeling skin syndrome is a rare entity. It is a subset of peeling skin syndrome; the other subset being the generalized form. The former presents at or shortly after birth and remains lifelong in most of the cases. Superficial peeling of skin of dorsal and volar aspects of hands and feet, associated with mild erythema and pruritus, may be seen especially on immersion in water. Summer exacerbations may be present. Generalized body examination including the mucosae is unremarkable. Associated constitutional symptoms or organ involvement is not present. Intermittent remissions may be seen. Biochemical and serological markers are unremarkable. Skin biopsy shows hyperkeratosis and splitting of the epidermis between the granular layer and the stratum corneum. The diagnosis is mainly based on history and clinical examination with supportive histopathology. As stated by Garg K et al⁷ treatment options are not very satisfactory including emollients, topical keratolytics, urea, calcipotriol, mild steroids. Systemic drugs include corticosteroids at low doses of 1-2 mg prednisone per kg per day for 4-6 weeks with evaluation of response and tapering of dose to prevent HPA axis suppression, growth retardation, cushingoid features, bony defects, abnormal carbohydrate and fat metabolism. Sarma N et al⁸ reported that retinoid being unsuccessful in treating PSS.

Mizuno y et al⁹ reported successful treatment of PSS with calcipotriol. A 9 year old Japanese girl with generalized PSS since birth was treated with 0.005% topical calcipotriol applied once a day on the affected areas for 4 months with improvement.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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DOI: 10.5455/2349-3933.ijam20150520

Cite this article as: Dhillon KS, Sharma D, Singh T, Srivastava R, Agarwal V, Srivastava S, Khan A, Yadav S. Acral peeling skin syndrome. *Int J Adv Med* 2015;2:172-4.