

Biophysical profile of a case of Peeling Skin Syndrome – A model for study of Stratum Corneum function

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Abstract—Peeling skin syndrome (PSS) is a rare recessively inherited ichthyosiform genodermatoses characterized clinically by asymptomatic, localized or generalized, continuous exfoliation of the superficial layer of the skin. A 12-year-old male child was presented with history of asymptomatic peeling of skin from his neck, trunk, and proximal extremities, since the age of 6 years. Histologically, stratum corneum was separated from the underlying stratum granulosum. Clinical and histopathological picture corresponded to the non-inflammatory variant of peeling skin syndrome (type A). Biophysical profile, which includes skin hydration, transepidermal water loss, sebum content, skin pH, of this patient was assessed from 7 different sites using the Corneometer, Tewameter, Sebumeter, Skin-pH-meter. Derangement was observed in all the above parameters. Peeling skin syndrome serves as a model for studying the stratum corneum functions. This article gives a critical review of the structure and various barrier functions of stratum corneum.

Key words: Peeling skin syndrome (PSS), stratum corneum, Biophysical profile

I. INTRODUCTION

Peeling skin syndrome (PSS) is a rare recessively inherited ichthyosiform genodermatoses characterized clinically by asymptomatic, localized or generalized, continuous exfoliation of the superficial layer of the stratum corneum (SC). It has 2 subtypes, acral and generalized form. The later has been subdivided into type A (non-inflammatory)¹ and type B (inflammatory)² Mevorah *et al*³ introduced a new subtype, type C³, which starts in infancy and characterized by atopy, itching, and presence of circular erythematous patches that are encircled by areas of peeling. Histologically it is characterized by separation in the intracorneal or subcorneal layers of the epidermis.⁴ Etiology of this disorder is unknown.⁵ It has been postulated that this is due to reduced adherence of abnormally thick stratum corneum to the stratum granulosum.³ Total number of cases of PSS reported so far is 81 and it consisted of 20 cases of type A PSS, 27 cases of type B PSS, 2 cases of type C PSS, 21 cases of acral PSS, and 11 of undefined phenotypes.^{1,3}

Biophysical profile is a bioengineering method to objectively assess the skin function. Stratum corneum function assessment is done by measuring 4 parameters : Skin hydration, measured by Corneometer CM 825; Skin sebum content, measured by Sebumeter SM 815; Skin barrier function or transepidermal water loss (TEWL), measured by Tewameter Triple TM 330T; Skin pH, measured by Skin-pH-Meter PH 905 (supplied by the manufacturer, Courage & Khazaka electronic GmbH, Cologne, Germany).

Peeling skin syndrome (PSS) serves as a model for studying the stratum corneum functions, as it involves separation of stratum corneum.

II. METHODOLOGY

A rare case of Peeling skin syndrome (PSS) was presented in skin and VD department of SMS hospital, Jaipur (Rajasthan) India, which is attached to a medical college.

This case was entered and examined in details. Along with biophysical profile of this case stratum corneum function assessment is done by measuring 4 parameters : Skin hydration, measured by Corneometer CM 825; Skin sebum content, measured by Sebumeter SM 815; Skin barrier function or transepidermal water loss (TEWL), measured by Tewameter Triple TM 330T; Skin pH, measured by Skin-pH-Meter PH 905 (supplied by the manufacturer, Courage & Khazaka electronic GmbH, Cologne, Germany).

All the relevant data were collected and finally this case was presented as a case report.

III. CASE REPORT

A 12 year old hindu male child, born out of a non-consanguineous marriage, presented at the Department of dermatology, SMS Medical college, Jaipur with the chief complaint of peeling of skin from his neck, trunk, and proximal extremities, especially following friction or rubbing since the age of 6 years. These episodes were asymptomatic and continuous with summer exacerbation. The patient was otherwise healthy and had no history of erythema, blistering, flexural involvement, or other major illness. He had not received any systemic therapy previously. There was no history of similar illness in any of the family members.

On dermatologic examination, there were focal areas of peeling skin patches over the sides of trunk and extremities (Figure 1 & Figure 2). On gentle rubbing of normal-looking areas of skin, peeling of thin, superficial layers was observed. Sheets of superficial epidermis could be easily peeled without bleeding or pain. The underlying skin was not inflamed and no residual hyperpigmentation was noted. Palms and soles were not involved. The teeth, hair, nails, and mucosa were normal.

Figure 1
Superficial peeling of skin over on
flexor aspect of forearm



Figure 2
Superficial peeling of skin over on
back of both the thighs



A skin biopsy specimen revealed slight hyperkeratosis and thinning of the granular cell layer. The stratum corneum was separated from the underlying stratum granulosum. No signs of inflammation

were present. A complete blood count, urinalysis, and routine blood chemistry were within normal limits.

Biophysical profile of the skin was assessed using the Corneometer, Sebumeter, Tewameter, Skin-pH-Meter as supplied by the manufacturer, Courage & Khazaka electronic GmbH, Cologne, Germany. The 4 parameters of stratum corneum function were measured from 7 different body sites ie, face, chest, back, both upper limbs and lower limbs. The test interpretations were as follows : Average TEWL showed a stained skin condition (test value ranges between 25-30 gm/hr/m² ; normal value range is 11.4-14.2 gm/hr/m²). Skin hydration showed dry skin condition (test value ranging from 30-45; normal value >45) as measured by parameters supplied by the manufacturer. Sebum value showed decreased sebum content (test value ranging from 0-6) from various sites and Skin pH showed slight shift towards alkaline side. (Table 1)

Table 1
Biophysical profile of skin of Case of various part affected

S. No.	Biophysical profile parameters	Normal Range	Case Test Value	Remark
1	Average TEWL	11.4-14.2 gms/hr/m ²	25-30 gms/hr/m ²	Increased
2	Skin hydration	>45	30-45	Decrease (dry skin)
3	Sebum	>6	0-6	decrease
4	Skin pH	For males=4.5-5.5 For females =4.3-5.5	6.7	Increased (Alkaline)

IV. DISCUSSION

Stratum Corneum (SC) represents the most superficial and final layer of maturation of the epidermis. It was not too long ago that SC was believed to be “a graveyard of insoluble keratin fibrils”, the “dead” outermost layer of skin, devoid of biological activity and function. The insistent work and research done by various “corneobiologists” in the past 5-6 decades, established the significance of SC in contributing to the physiological homeostasis of the skin, and the so called ‘dead layer’ became ‘alive’ functionally.^{6,7,8} Historically thought to be made up of a collection of sturdy cellular-like structures; with time, the concept of the stratum corneum being viewed as “bricks” and “mortar” emerged, with the corneocytes representing the bricks and the intercellular lamellar lipid membrane representing the mortar between the bricks.⁹ The major structural components of the SC includes the corneocytes (bricks) comprised primarily of keratin macrofibrils, protected externally by a cornified cell envelope, which are cohesively held together by corneodesmosomes. Cornified cell envelope is composed predominantly of proteins (e.g., loricin, involucrin) and a covalently bound outer lipid monolayer that is primarily made up of long chain ceramides.⁹⁻¹¹ The intercellular lamellar lipid membrane comprises three major classes of lipid components : ceramides (40–50%; multiple subfractions), cholesterol (25%), and fatty acids (10–15%), derived from the precursor lipids in the lamellar bodies present in granular layer.^{8-10,12} A mixture of multiple small hygroscopic compounds present within corneocytes (free amino acids, pyrrolidone carboxylic acid (PCA), and urocanic acid, simple sugars and electrolytes, obtained

from the degradation of filaggrin) referred to collectively as natural moisturizing factor (NMF), plays a vital role in the physiological maintenance of stratum corneum hydration.^{6-9,13,14}

The SC is multitasking as evidenced by its inherent ability to provide several barrier properties. The epidermal permeability barrier refers to the ability of the SC to regulate TEWL, retain moisture for proper enzymatic desquamation, and provide selective permeability of exogenous and endogenous substances.^{7,9,10,15} Other barrier functions include immunity barrier, antioxidant barrier, antimicrobial barrier, photoprotection barrier, and hormone receptor functions.^{6,8-10,16}

The normal skin has an acidic pH commonly referred to as the “acid mantle” of the skin. The pH of the SC has a significant influence on the barrier functions of the SC.^{17,18} In addition, acidic SC pH favours growth of normal bacterial flora, as opposed to an alkaline pH, which is more supportive of the growth of pathogens such as *Staphylococcus aureus* and *C.albicans*¹⁸ thus contributing to the antimicrobial barrier. Sebum also contributes to the anti microbial barrier as it is a source of various anti microbial peptides and unsaturated fatty acids, particularly oleic acid, which are the active agent against pathogenic micro-organisms.¹⁹ The SC abnormalities is known to be associated with various dermatological diseases like atopic dermatitis^{20,21} psoriasis^{22,23}, rosacea²⁴, acne²⁵, ichthyotic group disorders and other disorders of keratinisation.

This patient had generalized non-inflammatory (type A) peeling skin syndrome with defective skin barrier functions as evidence by high TEWL, low skin hydration and sebum content, and high skin pH. This derangement is attributed to the split in the stratum corneum and the disordered process of keratinisation, but the genetic defect in PSS type A is not known. However it provides a suitable model to study the barrier function of skin particularly stratum corneum. Understanding the physiological, chemical, and biophysical characteristics of the skin helps us to arrange a proper approach to the management of skin diseases.

V. CONCLUSION

Peeling skin syndrome (PSS) serves as a model for studying the stratum corneum functions, as it involves separation of stratum corneum.

CONFLICT OF INTEREST

None declared till now.

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