

Peeling skin syndrome: 11 cases from Saudi Arabia

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BACKGROUND: Peeling skin syndrome (PSS) is a rare genodermatosis of probable autosomal recessive inheritance. In Saudi Arabia, consanguinity of parents is common and consequently the occurrence of familial disease, including that of the skin, is not uncommon.

METHODS: To characterize the clinical and pathological features of PSS in Saudi Arabia, we reviewed the medical records and clinical photographs of patients with recurring blistering diseases and conducted a histopathologic evaluation of skin biopsies to identify the site of cleavage.

RESULTS: Eleven patients with PSS were seen at King Khalid National Guard Hospital in Jeddah between the years 1986 and 2005. Ages ranged between 2 and 15 years and there were 9 males (81.8%) and 2 females (18.2%). The most common presentation in the majority of patients was localized spontaneous peeling of the skin. Eight patients (72.7%) had a history of vesicles that were small, dry and peeled away. Trauma did not play a role in blister formation. All patients were local from Bedouin tribes where a family history of a similar complaint was documented in 8 cases (72.7%) and consanguinity of marriage was evident in 6 patients (54.5%). Histological examination of skin biopsies showed either intracorneal or superficial subcorneal cleavage above the granular layer in all biopsied patients.

CONCLUSION: Although rare, PSS occurs in Saudi Arabia and is most likely related to consanguinity of marriages. This disease is generally mild and is characterized by intracorneal cleavage within the superficial epidermis. The disease should be recognized and not confused with other vesicobullous disease.

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Peeling skin syndrome (PSS) is a rare genodermatosis, probably of autosomal recessive inheritance with variable age of onset from birth to adulthood.¹ It is characterized clinically by spontaneous superficial peeling of the skin in a localized or generalized pattern associated sometimes with erythema and vesicle formation with seasonal changes, being worse in the summer. Hair, nail and biochemical abnormalities are rare.¹ Histologically, PSS is characterized by cleavage in the stratum corneum or above the stratum granulosum with an essentially unremarkable stratum spinosum or dermis.²

Methods

We reviewed the medical records and clinical photographs of patients with recurring blistering diseases seen at the dermatology department of King Abdulaziz Medical City, Jeddah during the period 1986 to 2005. Skin biopsies were evaluated to identify the site of cleavage.

Results

Eleven patients were seen during the study period (Table 1). Ages ranged between 2 and 15 years. There were 9 males (81.8%) and 2 females (18.2%). The most common presentation in the majority of patients was localized spontaneous peeling of the skin. Eight patients (72.7%) had a history of vesicles that were small, dry and peeled away. Trauma did not play a role in blister formation. The age of onset of disease varied from birth to 5 years and the duration ranged from 4 months to 15 years. All patients were local from Bedouin tribes where a family history of similar complaints was documented in 8 cases (72.7%) and consanguinity of marriage was evident in 6 patients. Histological examination of skin biopsies showed either intracorneal or superficial subcorneal cleavage above the granular layer in all biopsied patients. Immunofluorescence testing, done in 2 patients was negative. Cases 1 and 2 were broth-

ers. Cases 3, 4 and 5 were siblings. The remaining 6 cases came from unrelated families. In two patients the condition was worse during the summer months. Skin biopsies in most patients showed a separation or cleavage within the stratum corneum as shown in Figure 1.

Cases 1 and 2

Case 1 was 3-year-old boy seen in our outpatient clinic due to development of vesicles on the skin of both the upper and lower limbs that ruptured, leaving a superficial continuous peeling of the skin. The condition had been noted since the age of 2 months with no seasonal variation. His general health was otherwise good. He gave a family history of a similar condition. His parents were first-degree cousins. Physical examination of the skin showed numerous areas of hypopigmentation and areas of erythema with superficial peeling involving the elbows, dorsum

Table 1. Summary of findings in the 11 cases of peeling skin syndrome.

Feature	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	Case 10	Case 11
Age (y)	3	6	10	3	2	10	5	6	15	5	10
Sex	M	M	M	F	M	M	M	M	F	M	M
Onset (mo)	2	2	Birth	Birth	Birth	3	24	60	3	60	3
Family history	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	No	No
Consanguinity	Yes	Yes	Yes	Yes	Yes	No	No	No	Yes	No	No
Cutaneous feature											
Vesiculation	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	Yes	No	No
Peeling	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Pigmentary changes	Yes	Yes	No	No	No	No	Yes	Yes	No	Yes	No
Erythema	Yes	Yes	No	No	No	No	Yes	No	No	No	Yes
Symptoms											
Pruritis	No	No	No	No	No	No	Yes	No	No	No	Yes
Seasonal variation (worse in summer)	No	No	No	No	No	No	Yes	No	No	No	Yes
Location											
Generalized	No	No	No	No	No	Yes	No	No	No	No	No
Upper limbs	Yes	Yes	No	No	No	Yes	No	No	No	No	No
Lower limbs	Yes	Yes	No	No	No	Yes	No	No	No	Yes	Yes
Hands and feet	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Histology											
Intracorneal	Yes	Yes	--	Yes	--	No	Yes	No	No	Yes	Yes
Subcorneal	No	No	--	No	--	Yes	No	Yes	Yes	No	No

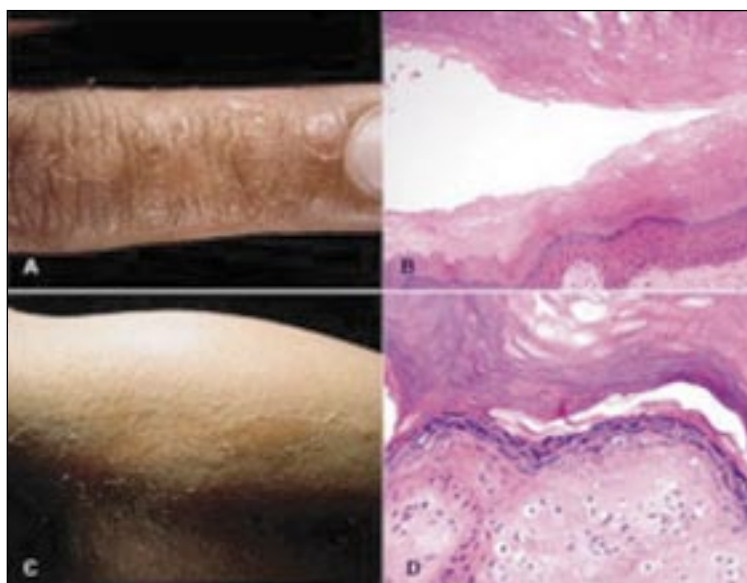


Figure 1. Areas of hypopigmentation and erythema with superficial peeling involving dorsum of hands; close-up of finger (A). Histological intracorneal cleavage (H&E stain x 125) (B) (Case 1). Skin peeling on the leg (C). Histological intracorneal cleavage (H&E x 225) (D)(Case 2).



Figure 2. Skin peeling on palm (A). Intracorneal cleavage of skin biopsy (H&E x 125) (B) (Case 4).

of both hands and the lower limbs (Figure 1). Basic laboratory tests were unremarkable. Histological examination of the skin biopsy specimen showed cleavage within the stratum corneum above the granular layer (Figure 1). Immunofluorescence was negative. His 6-year-old older brother (Case 2) presented with the same complaints, which started at the age of 2 months. His physical and histological findings were similar to his brother (Figure 1).

Cases 3, 4 and 5

Case 3 was a 10-year-old boy with a history of flaccid vesicles that ruptured easily leading to peeling of the skin on both hands and feet since birth. His sister (Case 4), a 3-year-old and his brother (Case 5), a 2-year old had a similar presentation. There was

no history of seasonal variation. Their parents were relatives. Physical examination showed superficial peeling of skin on the hands and feet with histological confirmation of subcorneal cleavage and negative immunofluorescence in Case 4 (Figure 2).

Case 6

This case was a 10-year-old boy who presented with continuous skin peeling over the trunk, knees, and dorsum of the hands and feet, preceded by vesicle formation since he was 6 months of age. No associated cutaneous or systemic manifestations and no seasonal variation was noted. Skin examination showed superficial skin peeling with hypopigmentation on the abdomen, knees, and dorsum of the hands and feet (Figure 3).

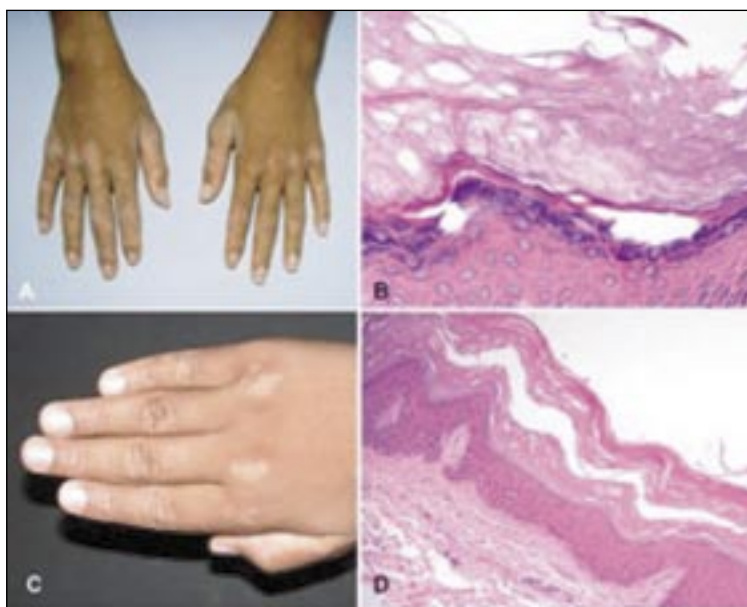


Figure 3. Superficial skin peeling with hypopigmentation on the dorsum of hands (A). Histological section shows subcorneal cleavage above the granular layer (H&E x 125) (B) (Case 6). Desquamation and hypopigmentation over the dorsal surface of hands (C). Histological examination shows intracorneal cleavage within the stratum corneum (H&E x 125) (D) (Case 10).



Figure 4. Superficial vesicle formation over the feet that ruptured, resulting in skin peeling (A). Histological subcorneal cleavage above the granular layer (H&E x 225) (B) (Case 9). Erythema and peeling of the skin over the feet (C). Histological examination shows intracorneal cleavage within the stratum corneum (H&E x 125) (D) (Case 11).

Case 7

This case was a 5-year-old boy who complained of fragile vesicles over the dorsum of both hands and feet that easily ruptured resulting in a continuous superficial peeling of the skin since the age of 2 years. This was associated with itching and erythema that was worse during the summer months. His uncle suffered from the same problem, but consanguinity of parents was not confirmed. Skin examination revealed peeling over the feet with areas of hyperpigmentation. Histopathologic examination of a skin biopsy specimen showed cleavage within the stratum corneum.

Case 8

This case was a 6-year-old boy who presented with depigmentation and peeling of the skin over the hands and feet for one year with no seasonal variation. His younger brother had a similar problem, but consanguinity of parents was not confirmed. Histopathology of a skin biopsy specimen showed subcorneal cleavage above the granular layer.

Case 9

This case was a 15-year-old girl with a complaint of superficial vesicle formation since the age of 3 months over the knees, hands and feet that ruptured resulting in skin peeling (Figure 4). No seasonal variation was noted. Her two brothers had a similar problem since infancy, and their parents were first-degree cousins. No other significant cutaneous or systemic findings were noted. Skin biopsy examination showed cleavage within the stratum corneum (Figure 4).

Case 10

This case was a 5-year-old boy who complained of depigmentation and peeling of the skin over the knees, hands and feet of 4 months duration with no seasonal variation noted. There was no family history of a similar problem. Skin examination showed desquamation over the dorsal surface of hands, knees and feet (Figure 3). Histological examination showed intracorneal cleavage consistent with peeling skin syndrome (Figure 3).

Case 11

Case 11 was a 10-year-old boy who presented with erythema and peeling of the skin over the knees, shin, and hands and feet (Figure 4) since the age of 3 months associated with pruritis, and worsening in the summertime. No family history was re-

corded. Histological examination showed intracorneal cleavage consistent with peeling skin syndrome (Figure 4).

Discussion

Peeling skin syndrome is a genodermatosis commonly seen in communities where consanguinity of marriage is prevalent. The disease is of long duration and is apparently of an autosomal recessive mode of inheritance. Various names have been applied to these conditions and a number of individual cases have been described as the peeling skin syndrome.^{3,4} The differential diagnosis includes all variants of subcorneal disease. Subcorneal pustular dermatosis is clearly excluded since no pustules are seen. Bullous impetigo with subcorneal clefting is an acute condition while the condition described here is a chronic problem with no honey colored crusting or evidence of infection. Pemphigus foliaceus is excluded by the absence of acantholysis, negative immunofluorescence and the fact that the condition under consideration arises in children. Previous reports describe two types of PSS, non-inflammatory (type A) and inflammatory (type B)^{2,3}. In type A, there is asymptomatic peeling, with intact general health; histologically, there is orthokeratosis with a normal epidermis and splitting occurring either within the lower part of the stratum corneum or directly above the granular layer. In PSS type B, there are erythematous migratory patches with a peeling border; pruritis or burning may be conspicuous. Histologically, the erythematous peeling patches have a psoriasiform appearance, and the entire stratum corneum separates from the underlying epidermis.³ PSS commonly shows generalized superficial desquamation, usually beginning at birth, infancy or early childhood and is persistent or periodic in most affected people, involving the trunk, limbs and occasionally the face, leaving mildly erythematous intact skin. Mild pruritis is a problem for some patients and patchy hypo- or hyperpigmentation has been noted. The palms and soles are either spared or are mildly thickened without peeling, and hair, nails and teeth are usually normal, though one patient was reported with severe palmoplantar subcorneal blistering, ichthyosis and keratotic cheilitis.⁵ A few patients show seasonal variation with worsening in the summer months.⁴ In contrast to previous reports the majority of our patients have localized distribution of skin peeling. The onset of symptoms is at birth, or shortly thereafter, and is marked by easy peeling of skin, mild pruritis in a minority of patients and a lack of systemic

manifestations. This relatively benign nature of the condition and its clinical resemblance to other cutaneous disease such as epidermolysis bullosa simplex superficialis suggest that the entity is underdiagnosed. This is particularly true considering the fact that parent consanguinity and intermarriages among families are very common in this part of the world.

Although clinical similarities existed to what Zvulunov et al⁶ described as "a new variant of autosomal recessive exfoliative ichthyosis" none of those patients showed histological intracorneal clefting as noted in our patients, in whom spongiosis was distinctly absent. The differential diagnosis to be excluded is hereditary epidermolysis bullosa simplex superficialis and, particularly the Weber-Cockayne variant, characterized by dominantly inherited, recurrent bullous eruptions of the hands and feet with friction, but with no continual peeling. Light microscopy of skin biopsies should, however, demonstrate

that the bulla unlike PSS is not subcorneal and is formed through the basal cell layer.⁷ For our patients who demonstrated occasional bullous eruptions on the hands and feet, which was worse in warm weather, the level of cleavage was within the stratum corneum on histological examination. Furthermore, no autosomally dominant pattern mode of inheritance was demonstrated in these patients. Management of this condition is difficult as no effective treatment has been reported. Patients are generally instructed to minimize immersion in water and to use absorbing powders or aluminum antiperspirants, especially in the acral variant of the disease. Keratolytic and urea creams might speed up shedding. The disease should be recognized and not confused with other vesicubullous disease, which is achieved by close clinicopathological correlation and adequate information on family history.

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