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Case Report

Peeling Skin Syndrome: A Case Report

Gülsüm Gençoğlan,[1]* MD, Işıl Kılınç Karaarslan,[2] MD, Tuğrul Dereli,[2] MD

Address:
[1]Afyon Kocatepe University Medical Faculty Department of Dermatology, Afyon, Turkey; [2]Ege University Medical Faculty Department of Dermatology, Izmir, Turkey
E-mail: gencoglan75@hotmail.com

* *Corresponding author*: Gülsüm Gençoğlan, MD, Afyon Kocatepe University Medical School Dermatology Department, Afyon, Turkey

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Abstract

Observations: Peeling skin syndrome is a very rare autosomal recessive disease characterized by widespread painless peeling of the skin in superficial sheets. We present a 31-year-old man with a lifelong history of continuous, spontaneous, asymptomatic generalized peeling skin. Histologically, there was epidermal separation at the level of stratum corneum, just above the stratum granulosum. The clinical picture corresponded to the non-inflammatory variant of peeling skin syndrome (type A). Histopathological study confirmed the clinical diagnosis of peeling skin syndrome. A critical review of the literature shows that the case presented here is exceptional.

Introduction

Peeling skin syndrome (PSS) is a very rare autosomal recessive disorder with onset at birth or childhood, characterized by asymptomatic, continuous shedding or peeling of skin in large sheets. Two variants of PSS, non-inflammatory (type A = *Fox*) [**1**] and inflammatory (type B = *Wile*) have been described [**2**]. Skin involvement is usually generalized, rarely localized. Histologically, there is separation at the level of stratum corneum, above stratum granulosum. Several clinical and histological differences exist between these two variants [**3**]. The inflammatory variant presents with erythroderma at birth and clinically shows overlap with *Comel–Netherton* syndrome [**4**]. A third presentation of the disease, with fissured cheilitis, blistering of the palms and soles, and desmosomal anomalies, has been described recently [**5**]. We present a case of Fox type PSS and review the relevant literature.

Case

A 32-year-old man presented with peeling skin that had been present since birth. Sheets of skin were peeling from his neck, trunk, and proximal extremities, especially following friction or rubbing. These episodes were asymptomatic and continuous, without any seasonal variation. 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.

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

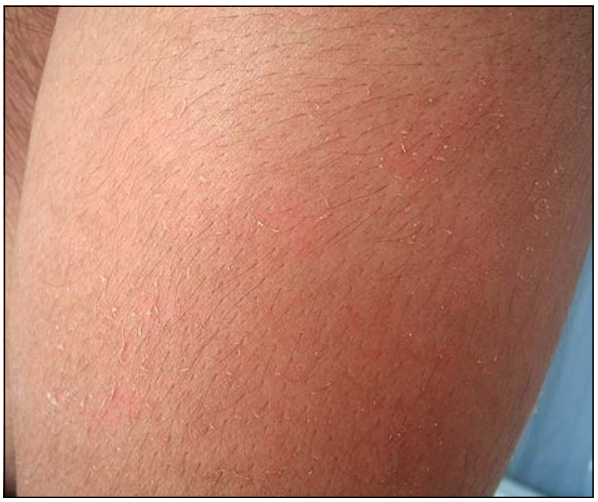

Figure 1. Sheets of peeling skin on the upper extremities. The underlying skin was not inflamed and no residual hyperpigmentation was noted.

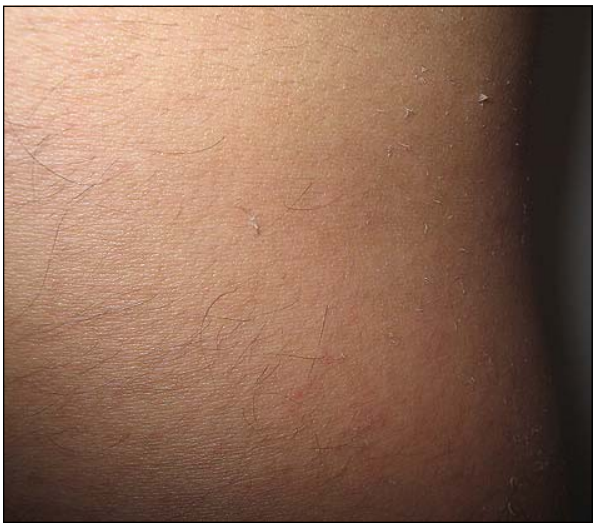

Figure 2. Peeling skin at the sides of the body. Sheets of superficial epidermis could be easily peeled without bleeding or pain.

involved. The teeth, hair, nails, and mucosa were normal. His parents were first-degree relatives, but his parents and siblings were not affected by PSS.

A complete blood count, urinalysis, and routine blood chemistry were within normal limits. Serum iron and copper levels were also normal. Plasma and urinary amino acids analyzed by paper chromatography did not yield any abnormalities. There was no eosinophilia.

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 [**Figure 3**]. Direct immunofluorescence studies did not reveal any immunoglobulin or complement deposition.

Discussion

The first case similar to ours was reported in 1921 and termed by Fox as "keratolysis exfoliativa congenital" [**1**]. In 1924, Wile described three unusual cases of congenital ichthyosiform erythroderma [**2**]. The question as to whether these cases represented similar or different disorders remains unanswered because ultrastructural, biochemical, or genetic studies have not been performed at that time. In 1969, Kurban and Azar reported four siblings in a family, using the term “familial continual skin peeling” [**6**]. Their cases were similar to Fox's case [**1**]. The etiology of this condition is still unknown. However, the increase in epidermal proliferation rate may account for the epidermal abnormality. Considering the cases reported up to date, the disease appears to be generalized, lifelong, and has an autosomal recessive mode of inheritance. The onset of symptoms is at birth, or shortly thereafter, and is marked by easy peeling of skin. Levy [**7**] and Inamadar [**8**] found abnormalities of amino acid metabolism with diminished plasma tryptophan in patients with this syndrome. Minor variations of urine amino acids were detected by Dicken [**9**] and Mevorah *et al* [**10**]. Hacham-Zadeh and Holubar [**11**] described patients with elevated serum copper levels, serum ceruloplasmin, iron and iron-binding capacity. The significance of these findings is uncertain. Our patient showed no amino acid abnormalities. His copper and iron levels were also within normal limits. Eosinophilia was reported in a few cases and it has been proposed that eosinophils, by the local release of cytotoxic cationic proteins, may

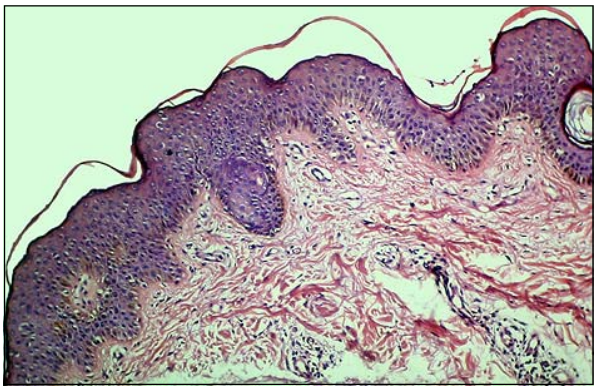

Figure 3. Skin biopsy specimen showed slight hyperkeratosis and thinning of the granular cell layer. The stratum corneum was separated from the underlying stratum granulosum.

play an important role in cutaneous splitting. However one month after birth, IgE and eosinophil levels had normalized despite the persistence of peeling skin [**12**]. The eosinophil level in our patient was within normal limits. Silverman *et al* reported intracellular cleavage and intercellular electron-dense globular deposits that represented abnormal lipids; they suggested that the disorder may present a retention hyperkeratosis rather than a hyperproliferative state [**13**]. Mevorah *et al* reported a keratohyalin abnormality and a four-fold increase in cellular retinoic acid binding protein. They also observed that etretinate had no significant effect on the course of this dermatosis [**10**].

No effective treatment for PSS has been reported. Methotrexate, UVB and oral corticosteroid therapy were found to be ineffective by Levy and Goldsmith [**7**]. Dicken observed that isotretinoin was also ineffective [**9**]. Mizuno *et al* tried 0.005 % calcipotriol ointment applied to the affected area once daily. They reported a decrease in peeling of skin and erythema after 4 months of calcipotriol therapy. However, it should be remembered that continuous application of calcipotriol ointment may cause hypercalcemia [**14**].

PSS is a very rare and not well-understood disorder of keratinization. Diverse clinical presentations of PSS were reported in the English literature. We believe that new cases like ours will help to clarify the pathogenesis and natural course of this syndrome.

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