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A Case of Late-Onset Peeling Skin Syndrome Likely Triggered by Irritation

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Dear Editor:

Peeling skin syndrome (PSS), refers to a rare form of ichthyosis associated with superficial, painless, continual, or seasonal exfoliation. The syndrome generally appears at birth and is clinically characterized by generalized scaling[1]. The condition is usually aggravated by heat, humidity, exposure to water and physical irritation.

A 46-year-old female patient visited the hospital due to slight itching on the trunk with skin peeling symptoms that began one year prior (Fig. 1). She had no noteworthy medial or family history. Before the lesion appeared, she had undergone with massage therapy including exfoliating scrub, massage, packing with moisturizer and steamed towel. After skin shedding began, she went through three additional massage sessions using the same technique.

A skin biopsy from the lesional skin of the left flank was evaluated via electromicroscopic (EM) inspection. Cleavage at the upper stratum corneum (SC) was found on H&E staining, but no abnormalities were observed in the dermis or epidermis (Fig. 2A). EM inspection showed splitting between the corneocyte rows (Fig. 2B).

PSS is a rare dermatosis and can be classified as generalized and localized type. Localized type is characterized by painless peeling of skin predominantly on the hands and feet and also named acral PSS. Three main subtypes of generalized PSS have been suggested based on the

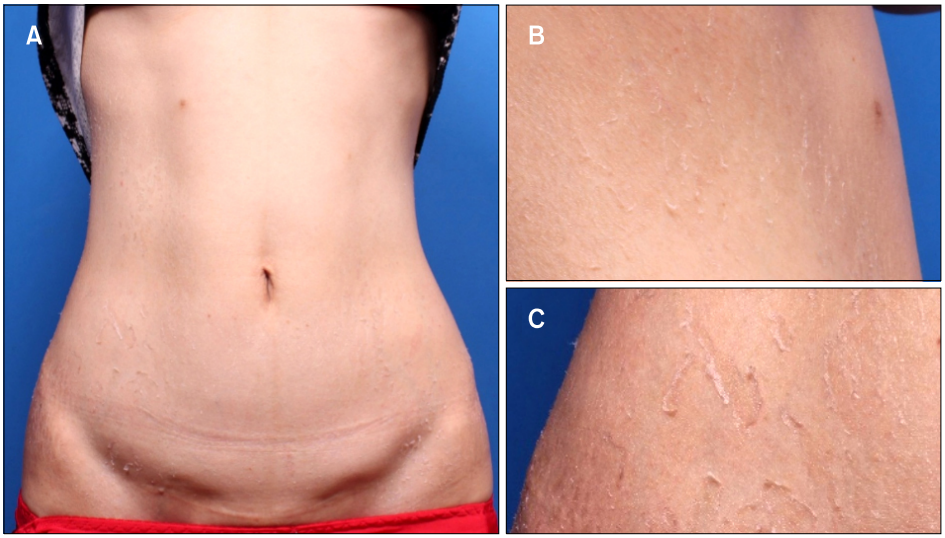

Fig. 1. Clinical features of the patient. (A, B) Diffuse skin shedding on the trunk. (C) Superficial sheets easily removed without bleeding or underlying erythema.

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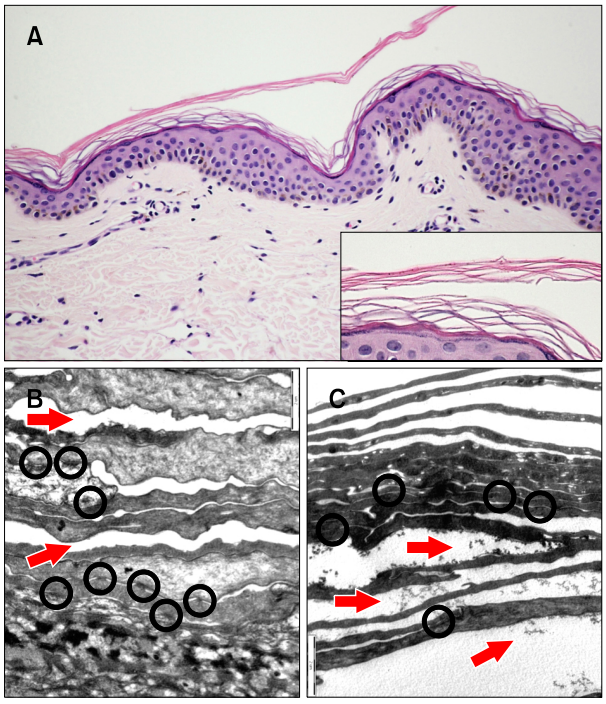

Fig. 2. Histopathological & electromicroscopic (EM) features of the biopsy specimen (collected from the left flank). (A) Epidermal cleavage within the stratum corneum. Orthokeratotic epidermis. Basal, spinous and granular layers were normal (H&E, ×200, inset: H&E, ×400). (B, C) Splitting (arrows) between the corneocyte rows. Corneodesmosomes (circles) were found between the corneocytes (EM, ×10 K).

presence or absence of inflammatory changes[2,3]. Type A is a non-inflammatory variant characterized by asymptomatic, generalized, continuous skin peeling that does not affect general health. Histologically, orthokeratotic epidermis with separation either within the lower part of the SC or just above the granular layer is typically observed. Type B exhibits ichthyosiform erythroderma, migratory erythematous pruritic patches with a peeling border with seasonal variation. Type C usually begins in infancy with erythematous patches surrounded by a superficial peeling collarette. Histologically, type B and C show the absence of the SC or a few layers of parakeratosis, which is separated from the stratum granulosum[2,3].

The exact etiology and pathogenesis of PSS type A has not been clarified[2,4]. Autosomal recessive inheritance is observed in many cases of PSS type A. Croneodesmosin gene mutation has been detected in the inflammatory form of this disease (type B), but genetic data are not available for the noninflammatory form (type A)[3].

Diagnosis of PSS is often delayed owing to late presentation in less severe cases. It is possibly underdiagnosed because of its low clinical expression. Our patient presented with clinical, histological, and EM features of generalized PSS type A, but has no familial history, and the disease occurred later in life and localized to trunk than typical.

Based on the above features, we could not clarify the relationship between cause and effect but supposed that the patient is genetically susceptible to PSS, and that an environmental factor, massage or exfoliating scrub or both in this case, triggered its late-onset. Thus far, few cases of late-onset PSS have been reported[4,5] and no cases of PSS have been reported previously in Korea. Collectively, we report a case of late-onset PSS type A likely triggered by irritation.

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