A Case of Peeling Skin Syndrome

Abstract
Peeling skin syndrome is a very rare autosomal recessive disease characterized by widespread painless peeling of the skin in superficial sheets. Etiology is still unknown with an autosomal recessive inheritance. Less than 100 cases have been reported in the medical literature. A 32-year-old man having asymptomatic peeling of skin since birth. Sheets of skin were peeling from his neck, trunk, and extremities, following friction or rubbing especially if pre-soaked in water but sparing palm and soles. Histologically, there was epidermal separation at the level of stratum corneum, just above the stratum granulosum. This case is being presented due to its rarity.

Keywords: Generalised non inflammatory, keratolytic, peeling skin syndrome

Introduction
Peeling skin syndrome (PSS) is a rare autosomal recessive cornification disorder that starts either at birth or later in childhood. It is characterized by continuous shedding of stratum corneum. Two forms of PSS are recognized, namely, Acral PSS (APSS; MIM 609796), which is caused by mutations in the TGM5 gene, encoding transglutaminase 5, and generalized PSS (MIM 270300). Generalized PSS is further divided into three types, namely, noninflammatory type (type A) and inflammatory types (type B) and C. In both the types (A and B) skin involvement is generalized, however, in type A presentation is erythroderma at birth, showing overlapping with camel netherton syndrome.

Case Report
A 32-year-old male presented to our outpatient department with complaint of repetitive shedding of skin since birth. Peeling of skin was present all over the body [Figures 1 and 2], sparing palms and soles [Figure 3]. Symptoms aggravate on exposure to water. Family history was present; one of his sibling had the same condition. There was no seasonal variation. On dermatological examination, superficial peeling was present. On rubbing of normal looking skin, it peeled off easily without any pain and erythema. Underlying skin was not inflamed. General and systemic examination revealed no abnormality. Routine hemogram and urine analysis were normal. Histopathological examination revealed epidermal separation in stratum corneum, thinning of epidermis, and loss of rete ridges. Mild lymphocytic infiltrate was present in epidermis. Dermis was normal [Figure 4]. He was prescribed emollient along with urea. After 2 weeks, patient was partially relieved symptomatically. However, the patient then discontinued treatment.

Discussion
PSS is a genodermatosis commonly affecting communities where consanguinity of marriage is present. The term peeling skin syndrome was given by Levy and Goldsmith in 1982. Asymptomatic shedding of large sheets of the skin, present since birth or early childhood, is a characteristic of PSS. Peeling can be produced by rubbing skin specially if pre-soaked in water. PSS type A is characterized by noninflammatory and asymptomatic peeling. Onset is congenital or before 6 years of age. Type A PSS has been shown to have its genetic basis in the CHST8 gene encoding a Golgi transmembrane N-acetylgalactosamine-4-O-sulfotransferase (GALNAC4-ST1) in 2012. In histology, hyperkeratosis, parakeratosis, reduction of the granular layer and acanthosis are seen. There is separation of the stratum corneum from the underlying granular layer.

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PSS type B is caused by deleterious mutations in the CDSN gene encoding corneodesmosin. Onset is congenital. The epidermis is psoriasiform with an absent or reduced granular layer, with marked parakeratosis and acanthosis.
The split occurs at the level of the granular layer. Type C starts in infancy and severe palmoplantar subcorneal blistering, ichthyosis, and keratotic cheilitis (PSS typeC). Electron microscopy of skin is helpful. Many patients have elevated IgE.

No effective treatment is reported till date. However, some improvement is seen with keratolytic agents and urea. Topical calcipotriol is also found to be little effective. Other treatment modalities such as topical tar, emollient, topical steroid, methotrexate, and phototherapy had been used but not effective.

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Conflicts of interest
There are no conflicts of interest.

References
1. Sparker MK. Differential diagnosis of neonatal erythroderma. In: Harper J, Oranje A, Prose N, editors. Textbook of pediatric dermatology. Oxford: Blackwell Science; 2000. p. 92-103.
2. Cassidy AJ, van Steensel MA, Steijlen PM, van Geel M, van der Velden J, Morley SM, et al. A homozygous missense mutation in TGM5 abolishes epidermal transglutaminase 5 activity and causes acral peeling skin syndrome. Am J Hum Genet 2005;77:909-17.
3. Sárdy M, Fáy A, Kárpáti S, Horváth A. Comel–Netherton syndrome and peeling skin syndrome type B: Overlapping syndrome or one entity? Int J Dermatol 2002;41:264-8.
4. Levy SB, Goldsmith LA. The peeling skin syndrome. J Am Acad Dermatol 1982;7:606-13.
5. Cabral RM, Kurban M, Wajid M, Shimomura Y, Petukhova L, Christiano AM. Whole-exome sequencing in a single proband reveals a mutation in the CHST8 gene in autosomal recessive peeling skin syndrome. Genomics 2012;99:202-8.