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Acral Peeling Skin Disease: About 2 Cases

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Case Report

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Abstract

Acral peeling skin syndrome is a rare, autosomal, recessive genodermatosis characterized by painless spontaneous exfoliation of the skin of the hands and feet at a subcorneal or intracorneal level. We are reporting 2 cases of spontaneous asymptomatic peeling skin limited to the acral surfaces especially the soles of the feet. Skin biopsy showed slight separation between the granular and corneal layers, with no inflammation. Both of them were treated with emollient and a slight improvement has been noticed.

Introduction

Acral peeling skin syndrome is a rare, autosomal, recessive genodermatosis characterized by painless spontaneous exfoliation of the skin of the hands and feet. It appears most often at birth or later in childhood or early adulthood. Molecular studies have identified mutations in the TGM5 gene that encodes transglutaminase 5, which plays an important role in the crosslinking of cornified cell envelope proteins. We are reporting 2 cases.

Case Report

Two girls aged respectively 5 and 6 years presented, since early childhood, spontaneous asymptomatic peeling skin limited to the acral surfaces especially the soles of the feet. For both of them, heat, humidity or friction exacerbed symptoms, with occasional blisters. On physical examination, the patients presented, fissures associated with peeling on soles, and resolving areas with residual erythema. The rest of the skin and the mucosas were normal. Skin biopsy showed slight separation between the granular and corneal layers, with no inflammation. The typical clinical picture and the biopsy on the feet confirmed the acral peeling skin syndrome. Both of them were treated with emollient and slight improvement has been noticed.

Discussion

Acral peeling skin syndrome is considered a localized variant of Peeling Skin Syndrom. This rare autosomal recessive genodermatosis belongs to the group of hereditary ichtyosis defined by generalized forms of Mendelian disorders of cornification [1]. The age of the beginning is variable, but in the majority of cases reported, the disease develops at birth or in the early years of life [2].

Clinically it is characterized by non-painful, continuous and superficial desquamation affecting mainly the dorsal part of the hands and feet. However, other authors have reported palmoplantar involvement wich was the case with our patients [3]. Exfoliation or blistering can be provoked by heat, humidity and friction

wich was the case in our patients. The resulting lesions can be tender and usually heal without scarring, sometimes leaving residual erythema. Histologic and ultrastructural analyses demonstrate that cleavage occurs at the junction of the stratum granulosum and the stratum corneum, which was the case with our patients Histology can also show hyperkeratosis, orthokeratosis or psoriasiform acanthosis [2].

Molecular studies of families with Acral peeling skin syndrome showed mutations in the gene encoding transglutaminase 5. Indeed Cassidy, et al. located the anomaly on chromosome 15 which contains a group of transglutaminase genes [4]. However, only transglutaminase 5 has a strong cutaneous expression at the extremities and plays a role in the binding of structural proteins and is required for structural integrity of the outermost epidermal layers. The weakness of these links within acral peeling skin syndrome explains the split in the area between granular layer and the stratum corneum [4].

Recently Krunic, et al. identified a mutation within the CSTA gene coding for cystatin A in a case ofacral peeling skin syndrome [5]. Cystatin A is a protease inhibitor present in the envelope cornea [5]. There is no well-codified treatment, topical corticosteroids, emollients and keratolytics have been used with inconsistent results, systemic retinoids being reserved for the handicapping cases.

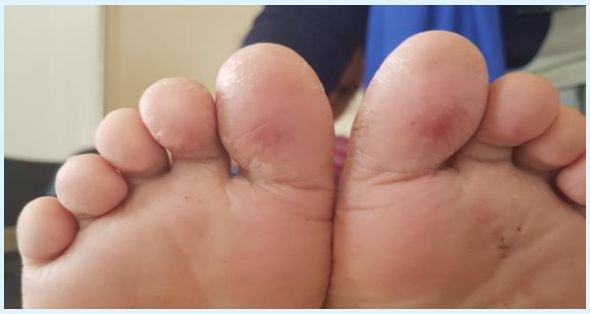

Figure 1: Superficial exfoliation of toes and background erythema.

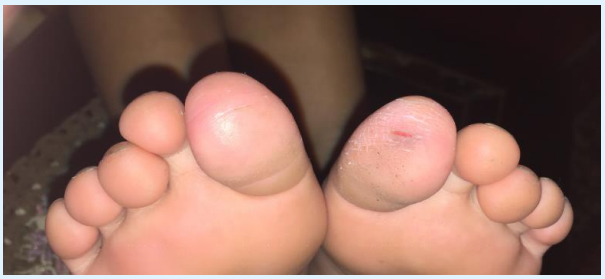

Figure 2: Superficial expoliation with fissuration of toes.

Conclusion

Acral peeling skin syndrome is a rare autosomal recessive disorder. It's a group of diseases that are heterogeneous in clinical and genetic terms with no well codified treatment. The identification of the mutation of the gene encoding transglutaminase 5 allowed a better understanding of the pathology.

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