CASE REPORT

Generalized Form of Peeling Skin Syndrome: A Case Report

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Abstract

Peeling skin syndrome (PSS) is a very rare keratinization disorder, characterized by spontaneous exfoliation of the stratum corneum. Herein, we report a case of non-inflammatory (type A) PSS.

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Key words: peeling skin syndrome, keratinization, stratum corneum

Introduction

Peeling skin syndrome (PSS) is a very rare keratinization disorder, probably inherited as an autosomal recessive trait, characterized by spontaneous exfoliation of the stratum corneum and, histologically, by subcorneal or intracorneal splitting[1,2].

Case Report

A seventeen year-old man was presented with asymptomatic shedding of large sheets of skin since early childhood (Figure 1). The scaling seemed to occur spontaneously, and the patient could peel off sheets of stratum corneum without pain. The patient stated that the dermatosis began at the age of six and worsened in hot weather with bathing. He had used several topical agents without improvement. Physical and motor developments were normal.

His parents were consanguineous and his cousin also had similar lesions. Histopathological examination showed only orthokeratotic hyperkeratosis with an initial separation of the stratum corneum just above the stratum granulosum (Figure 2). No signs of inflammation were detected. All these data were consistent with the diagnosis of peeling skin syndrome (PSS).

Discussion

Asymptomatic shedding of large sheets of skin, present since birth or early childhood, is characteristic of peeling skin syndrome (PSS). Two variants of PSS, non-inflammatory (type A) and inflammatory (type B) have been described [3]. Although classically described as a generalized disorder sparing palms and soles, acral PSS with involving only palms and soles has also been reported [4,5]. Its pathognomonic histopathologic feature is the presence of cleavage within or below the stratum corneum. In type A, the onset is variable (congenital or before the sixth year of life); in type B, it is congenital. Histologically, type A shows orthohyperkeratosis while type B shows a psoriasiform pattern [6].

Erythrokeratolysis hiemalis [7,8], a rare autosomal dominant genodermatosis characterized by palmoplantar scaling, erythema and continuous peeling, begins in early childhood. Its main features, cyclical attacks in the winter and the necrobiosis of the keratinocytes in the Malpighian layer, allowed exclusion of this disease in our patient. Epidermolysis bullosa simplex superficialis (EBSs) is a newly described variant of epidermolysis bullosa with a cleavage plane similar to that of PSS [9]. Nevertheless, EBSs (epidermolysis bullosa simplex superficialis) can be differentiated from PSS

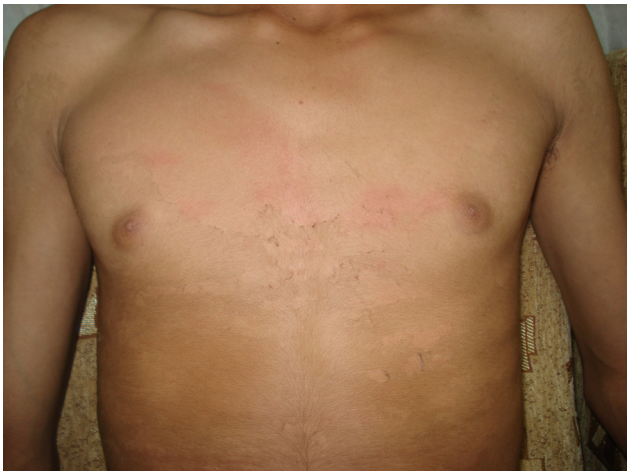

Figure 1. *Shedding of large sheets of skin*

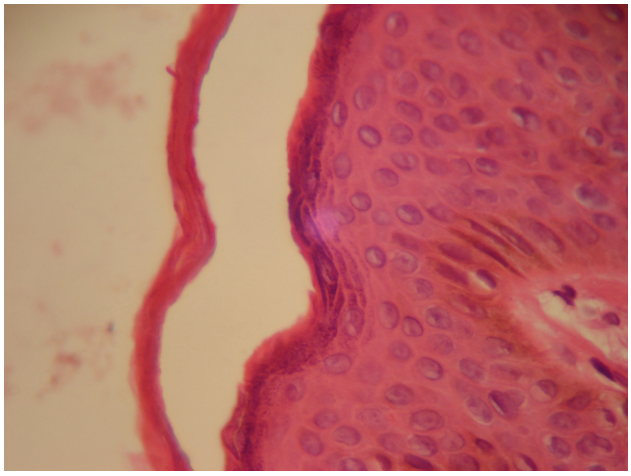

Figure 2. *Histopathology view: orthokeratotic hyperkeratosis with an initial separation of the stratum corneum just above the stratum granulosum (H&E*40)*

through the presence of mechanical blister formation without spontaneous exfoliation, its autosomal dominant mode of inheritance, and the fact that the onset is never after 2 years of age [9].

Thus, the main features of our case, namely the intracorneal splitting without inflammatory changes, the absence of symptoms or seasonal variation, the onset at 6 years of age, and the probable autosomal recessive mode of inheritance because of the parents' consanguinity, and the positive family history supported the diagnosis of PSS type A [10].

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