Case Report

Peeling Skin Syndrome: A Pathologically Invisible Dermatosis

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Abstract

Peeling skin syndrome is a relatively rare clinical case with pathology of apparently normal skin that needs clinical details to reach accurate diagnoses. Hence, this case was used as examples to declare how it is important for both the pathologist and the dermatologist to cooperate to reach an accurate diagnosis.

Keywords: Clinicopathological correlation, invisible dermatosis, peeling skin syndrome

Introduction

Clinicopathological correlation is important in the diagnosis of most of the dermatological diseases. It is often impossible to reach a correct diagnosis without putting the clinical data and the pathological findings in mind together. Nearly normal skin histology can be a finding in some cutaneous lesions, including some that have striking clinical presentation, and, for this reason, these diseases have also been referred to as "Nothing diseases."[1] Because the clinical presentation of such diseases is usually pathognomonic, most of these conditions are not frequently biopsied. For the pathologist, the diagnoses of these diseases are a challenge and may need repeat biopsies, examination of multiple sections, special stains, specific investigations such as immunofluorescence and immunohistochemistry, and proper clinical correlation[2] which may be the only clue.

Case Report

After filling a confirmed consent. A 13-year-old girl presented with peeling skin, and this was manifested since birth. Sheets of the skin were peeling from her neck, trunk, and proximal extremities, especially following friction or rubbing. This process was asymptomatic and continuous, with no seasonal variation. The patient was otherwise healthy. On dermatological examination, there were focal areas of peeling skin patches over the extremities [Figure 1a and b]. On gentle rubbing of the normal-looking areas of the skin, peeling of thin, superficial layers was observed. The sheets of the superficial epidermis that formed could easily be peeled without bleeding or pain. The underlying skin was not inflamed.

The palms and soles were not involved. Her parents were second-degree relatives, but they were not affected; she had a single younger brother who was affected by the same lesions. General and systemic examination revealed no abnormality. Routine hemogram and urine analysis were normal.

A skin biopsy specimen revealed moderate hyperkeratosis. There was a separation between stratum corneum and the underlying stratum granulosum. The dermis was unremarkable [Figure 2a and b].

Discussion

Peeling skin syndrome (PSS) or peeling skin disease is a rare, autosomal, recessively inherited ichthyosiform genodermatoses characterized by asymptomatic peeling of superficial skin in large sheets, which was first described as a case report in 1921 by Fox.[3]

PSS is an example of unobvious skin histopathology; as the separation at the level of stratum corneum, above stratum granulosum; which is the only finding; may pass unnoticed

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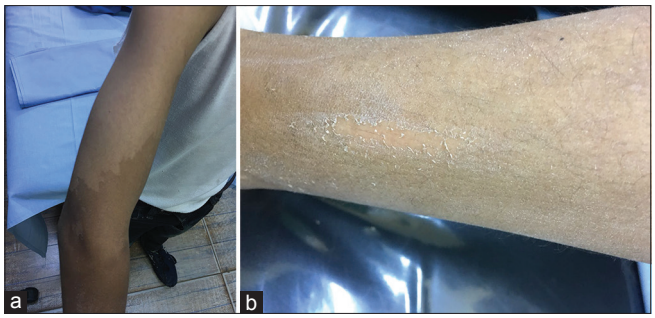

Figure 1: Peeling skin syndrome. Peeling of skin in the upper limb (a) and lower limb (b) with normal-looking underling

unless a good clinical datum is present. Because the histological features are deceptively bland, the general pathologist infers that the changes are nonspecific.

PSS is a relatively rare clinical case with pathology of apparently normal skin that needs clinical details to reach accurate diagnoses. Hence, this case was used as examples to declare how it is important for both the pathologist and the dermatologist to cooperate to reach an accurate diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

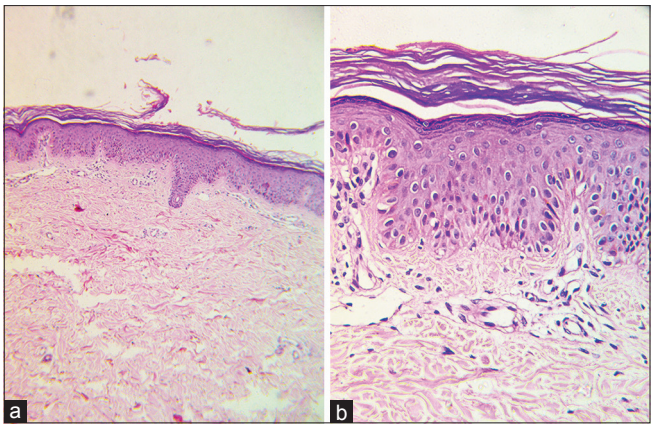

Figure 2: Peeling skin syndrome. Moderate hyperkeratosis. The stratum corneum is separated from the underlying stratum granulosum. The dermis is unremarkable (a, H and E, ×100). Higher power showing the separation (b, H and E, ×400)

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Conflicts of interest

There are no conflicts of interest.

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