

**PROGRESSIVE SYMMETRIC ERYTHROKERATODERMA:
FIRST CASE REPORTED IN THE DOMINICAN REPUBLIC**

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Abstract

Progressive symmetric erythrokeratoderma (PSEK) is an autosomal dominant genodermatosis with incomplete penetrance and variable expressivity. It belongs to the group of erythrokeratodermas where it can be differentiated from erythrokeratoderma variabilis in the absence of migratory erythematous lesions and in a greater incidence of palmoplantar keratoderma. Molecular basis of PSEK has not yet been established although there are reports of mutations in the *loricrin* gene. We report a 13-year-old boy with symmetrically distributed hyperkeratotic plaques over the dorsum of the hands and the extensor aspect of the forearms, elbows and knees. As far as we are aware, we report the first case of PSEK in the Dominican Republic.

Key words: Erythrokeratoderma; genodermatosis; Dominican Republic

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Introduction

Progressive symmetric erythrokeratoderma (PSEK) was first described by Darier in 1911 in his article entitled “Erythroqueratodermie Verruqueuse en Nappes, Symetrique et Progressive” [1]. In 1922 Gottron published an article describing the same entity but this time he calls it as we know it nowadays [2]. Since his initial description less than 50 cases have been published in the literature.

PSEK describes an autosomal dominant mode of inheritance with incomplete penetrance and variable expressivity. It usually develops during early childhood [3] as fixed and slowly progressive erythematous and hyperkeratotic plaques distributed symmetrically over the trunk, knees, elbows, dorsal surfaces of the hands and feet, and sometimes affecting also the face, palms and soles [4,5].

Despite that molecular basis of PSEK has not yet been established, there are reports of mutations in the *loricrin* gene [4,6]. As far as we are aware we report the first case of PSEK in the Dominican Republic.

Case Report

A 13-year-old male that presented to our institution with the onset of symmetrically distributed hyperkeratotic plaques

over the dorsum of the hands and the extensor aspect of the forearms, elbows and knees, from the age of 3, asymptomatic. His past medical history was not relevant, and there were no skin complains in any other family members.

Dermatologic examination revealed, multiple, irregularly-shaped, sharply demarcated and hypopigmented keratotic plaques symmetrically distributed, showing an erythematous border (Fig. 1, 2). Polarized dermoscopy showed white scaly lines over focal areas of hyperpigmentation (Fig. 3).

Skin biopsies were taken from the lesions near the elbows showing epithelial hyperplasia with hyperkeratosis, regular acanthosis, elongation and anastomosis of rete ridges, pigmentation of the basal layer and perivascular lymphohistiocytic infiltrate (Fig. 4). Based on the clinical and histopathological findings, the patient was diagnosed with progressive symmetric erythrokeratoderma.

Discussion

The dermatological picture of PSEK is considerable similar to erythrokeratoderma variabilis (EKV) with symmetrically distributed, fixed or very slowly progressive erythematous, scaly plaques. It can be differentiated in the absence of migratory erythematous lesions and in a greater incidence of palmoplantar keratoderma.



Figure 1. Symmetric hyperkeratotic plaques with well-defined outlines on the dorsum of the hands.



Figure 2. Symmetric hyperkeratotic plaques with erythematous outlines on the elbows and forearms.

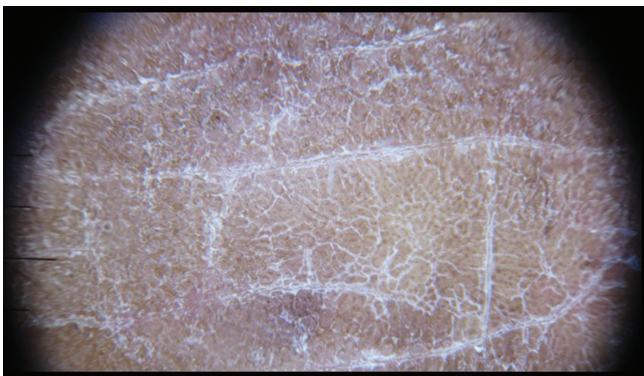


Figure 3. Polarized dermoscopy reveals white scaly lines over focal areas of hyperpigmentation.

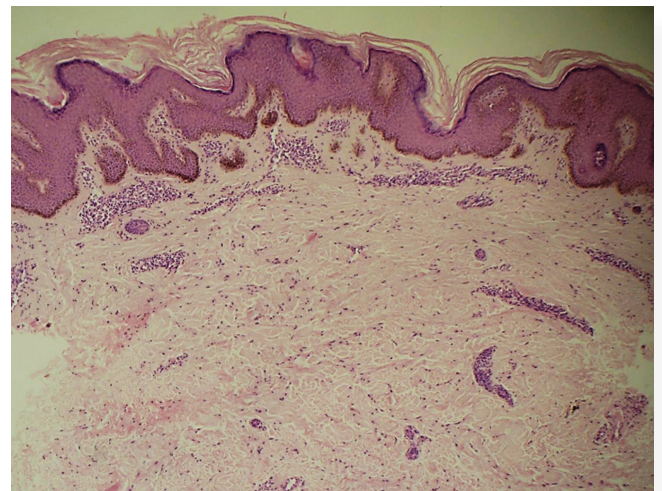


Figure 4. Epithelial hyperplasia observing elongation of the rete ridges, papillomatosis, verrucous hyperplasia and hyperpigmentation of the basal layer. HE 5X.

Also, the symmetry of the lesions in PSEK is more striking than in EKV [5]. These overlapping characteristics have led some authors to propose the alternate term “EKV et progressiva” [7]. The molecular basis of PSE has not been clearly elucidated, Ishida et al reported mutations in the loricrine gene [6] which codifies for loricrin, a major structural component of the cornified cell envelope of the epidermis, which participates in the formation of keratohyalin granules; however similar mutations have not been found by other authors [4]. More studies are needed to clarify the molecular basis of this entity.

It is interesting to observe the presence of white scaly lines

on polarized dermoscopy reminding us the Wickham striae that could correlate to the focal hypergranulosis seen on histopathology. Despite histopathology is non-specific it can help us to exclude other conditions such as pityriasis rubra pilaris that shows alternating orthokeratosis and parakeratosis in both vertical and horizontal directions [8], these features were not seen in this case.

Current treatment options for this condition include keratolytics and topical and systemic retinoids, Bilgin et al reported the use of topical calcipotriol with a remarkable improvements [3].

Conclusion

PSEK is an uncommon genodermatosis. Its pathogenesis is poorly understood however advances in molecular biology have identified mutations in genes that codify structural components of the epidermis yet more research must be done to determine the specific molecular and genetic basis of this entity. Diagnosis of PSEK is done clinically and histologically, however physicians can also be aided by dermoscopy, a practical non-invasive diagnostic tool that can help us to infer histological changes in the epidermis. Patients need to be counseled about the chronicity of their condition and the limitation of temporary improvements with the treatment options available.

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