Progressive symmetrical erythrokeratoderma on the face: A rare condition and successful treatment with calcipotriol

Nagihan Tarikci, MD, Emek Kocatürk Göncü, MD, Tülin Yüksel, MD, Ralfi Singer, MD, İlteriş Oğuz Topal, MD, and İlknur Mansuroğlu Şahin, MD
Istanbul, Turkey

Key words: calcipotriol; erythrokeratoderma; progressive symmetrical erythrokeratoderma.

INTRODUCTION

Erythrokeratodermas are rare genodermatoses that are characterized by well-defined erythematous, hyperkeratotic plaques.[1] They are categorized into 2 main types: progressive symmetrical erythrokeratoderma (PSEK) and erythrokeratoderma variabilis (EKV). Because their histologic findings are very similar, they have to be differentiated clinically.[2]

PSEK, or Gottron syndrome, was first described by Darier in 1911 but takes its name from an article published in 1922 by Gottron.[3] It is characterized by symmetrical, nonmigratory, well-defined, erythematous, hyperkeratotic plaques or reddish orange to brownish color lesions with marked erythema at their periphery. Lesions commonly occur on the knees, elbows, hands, and feet and may rarely involve the face. The trunk is generally spared.[2,3] PSEK usually appears in infancy or childhood and shows an equal sex incidence. Also, as the name suggests, symmetry in PSEK lesions is more remarkable than that in EKV lesions.[4]

PSEK shows a heterogeneous phenotypic variability. Although it shows mainly autosomal dominant inheritance, autosomal recessive and sporadic cases are also reported.[4]

The bioactive form of vitamin D3 (calcipotriol) is found to modulate epidermal proliferation and differentiation. Efficacy, tolerability, and safety of calcipotriol ointment in disorders of keratinization have been shown.[5] In this report, we describe a rare case of PSEK and successful treatment with calcipotriol.

Abbreviations used:

EKV: erythrokeratoderma variabilis
PSEK: progressive symmetrical erythrokeratoderma

CASE REPORT

A 7-year-old boy presented to our clinic with complaints of erythematous patches on his face and elbows that started 1 year after his birth. Similar lesions were not detected in other family members, and no systemic disease was detected in our patient. On physical examination, (Fig 1, *A*), hyperkeratotic erythrodermic plaques of variable thickness with sharply demarcated borders were seen on the face and elbows. No lesions were detected on the palms, soles, nails, hair, teeth, or mucosa. Fungal elements were not detected in potassium hydroxide preparation. The findings on laboratory tests, including complete blood count, liver function tests, glucose, creatinine, serum urea nitrogen, zinc and ferritin levels, and urine microscopy, were normal.

A punch biopsy specimen from the right cheek found nonspecific features of hyperkeratosis, parakeratosis, acanthosis, and superficial perivascular lymphocytic infiltration (Fig 2).

Clinical and histologic findings were consistent with the diagnosis of PSEK. Topical calcipotriol ointment was applied twice a day. No adjuvant steroids or emollients were used while using calcipotriol ointment. One month of therapy with calcipotriol resulted in complete resolution of the

From the Department of Dermatology, Okmeydanı Training and Research Hospital.
Funding sources: None.
Conflicts of interest: None declared.
Correspondence to: Emek Kocatürk Göncü, MD, Okmeydanı Training and Research Hospital, Department of Dermatology, Nadiraga Sok. No: 25/9 Göztepe, 34730, İstanbul, Turkey. E-mail: emekozgur@yahoo.com.

JAAD Case Reports 2016;2:70-1.
2352-5126

http://dx.doi.org/10.1016/j.jdcr.2015.11.010

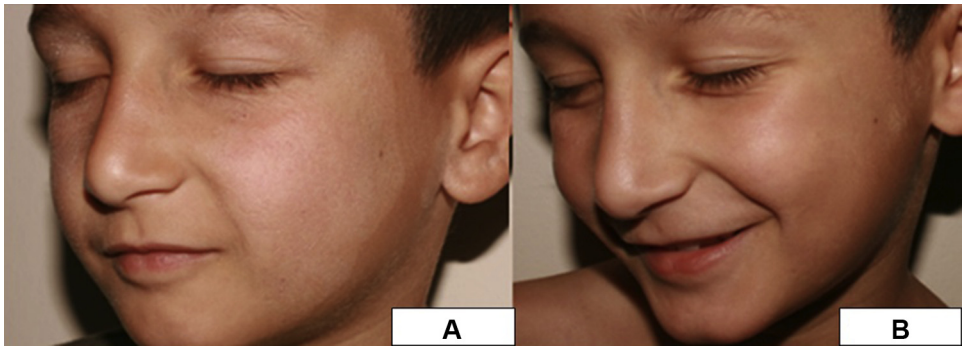

Fig 1. Progressive symmetrical erythrokeratoderma before (**A**) and after (**B**) calcipotriol therapy.

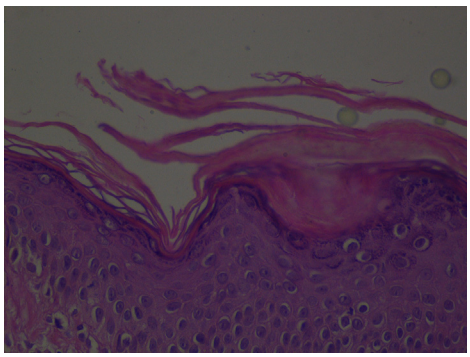

Fig 2. Hyperkeratosis, parakeratosis, acanthosis, and superficial perivascular lymphocytic infiltration.

lesions, and treatment was stopped. There was no recurrence after 4 months of follow-up (Fig 1, *B*).

DISCUSSION

PSEK is one of the erythrokeratodermas, characterized by erythtematous symmetrically distributed plaques. It differs from EKV with the absence of migratory erythematous lesions and its greater incidence of palmoplantar keratoderma. Also, the symmetry in PSEK lesions is more noticeable than that in EKV lesions.[2]

The mechanism of the disease is still mostly unkown. A frameshift mutation in the loricrin gene on chromosome 1q21 is seen in PSEK cases. Loricrin is the major constituent of the cornified cell and envelope of the epidermis, which participates in the formation of keratohyalin granules.[6] Functional studies in transgenic mice find that the mutant loricrin accumulates in the nucleus and destroys the keratinocyte apoptosis process, which leads to a pathologically thickened stratum corneum.[7]

Calcipotriol is officially indicated for the treatment of psoriasis because of its ability to inhibit keratinocyte proliferation and promote their differentiation.[5] Bilgin et al[8] reported an adult PSEK case treated successfully with calcipotriol in 2011.

In our patient, we preferred to use topical calcipotriol because of the presence of hyperkeratosis in PSEK and to avoid side effects of other treatments. A dramatic clinical response was observered through the application of calcipotriol, which mainly acts on the differentiation of keratinocytes.

Here we report a rare case of a erythrokeratoderma with facial involvement that responded well to topical calcipotirol. Topical calcipotriol could be used in the treatment of erythokeratodermas because of its rapid action and good safety profile.

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