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Progressive symmetrical erythrokeratoderma - response to topical calcipotriol

İlgül Bilgin[1], *Kübra Eren Bozdağ*[1], *Seçil Uysal*[1], *Murat Ermete*[2]

1. Atatürk Education and Research Hospital, Department of Dermatology, Izmir, Turkey.
2. Atatürk Education and Research Hospital, Department of Pathology, Izmir, Turkey.

Corresponding author:

Ass. Prof. Kübra Eren Bozdağ

Mithatpaşa Caddesi, No: 388/7

35280-Asansör, Izmir, Turkey

E-mail: *bozdagk@gmail.com*

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Abstract

Background: Progressive symmetrical erythrokeratoderma is characterized by well-demarcated, symmetrically distributed, erythtematous and hyperkeratotic plaques. Treatment options are topical retinoids, emollients, keratolytics and topical corticosteroids with limited or no success. Oral retinoids have been shown to be successful in some cases, but recurrence is to be expected on cessation of therapy. Topical calcipotriol is an established mode of treatment for psoriasis and also reported to be effective in many hyperkeratotic skin diseases.

Main observations: A 20-year-old female patient presented with reddish-brown lesions in her axillae, groins, submammary regions and on the eyelids. Clinical and histological findings were consistent with the diagnosis of progressive symmetrical erythrokeratoderma. She had noted that her lesions did not improve with topically applied steroids and emollients. We recommended our patient to use topical calcipotriol and observed a remarkable improvement within two weeks.

Conclusion: When compared with the other treatment modalities, topical calcipotriol is a safe and effective drug with minimal side effects and it would be a good alternative. (*J Dermatol Case Rep*. 2011; 5(3): 50-52)

Introduction

Erythrokeratoderma is the association of localized hyperkeratotic plaques with overlapping or distinct areas of circumscribed erythema. This clinically and genetically heterogeneus disease group has two major subtypes; erytrokeratoderma variabilis (EKV) and progressive symmetrical erythrokeratoderma (PSEK).[1] PSEK has its onset during early childhood and seems to be inherited as an autosomal dominant trait with incomplete penetrance and variable expressivity.[2] Differentiation of EKV from PSEK can be made by the sharply outlined geographical regions of migratory erythema observed in EKV.[3]

Case report

A 20-year-old female patient presented with reddish-brown lesions in her axillae, groins, submammary regions and on the eyelids. She also had mild palmoplantar hyperkeratosis. Dermatological examination revealed symmetrically distributed erythematous and hyperkeratotic reddish-brown plaques in her axillae (Fig. 1A), groins and submammary regions. She had palmoplantar hyperkeratosis and mild desquamative plaques on her eyelids. Her teeth, nails and hair were normal. Routine laboratory investigations were within normal limits. The histopathological examination of the skin biopsy taken from the inguinal region showed orthohyperkeratosis, irregular acanthosis and focal papillomatosis (Fig. 2).

Clinical and histological findings were consistent with the diagnosis of PSEK and oral isotretinoin (0,5 mg/kg) treatment was initiated. Since there was no improvement at the end of two months, oral isotretinoin treatment was stopped and topical calcipotriol ointment was applied twice a day. No adjuvant steroid cream or emollients was used while using calcipotriol ointment. The patient had noticed a remarkable improvement within two weeks (Fig. 1B). After clearance of the lesions she discontinued the medication and her lesions relapsed within two weeks. Afterwards, we recommended to use calcipotriol twice a day until the clearance of the lesions

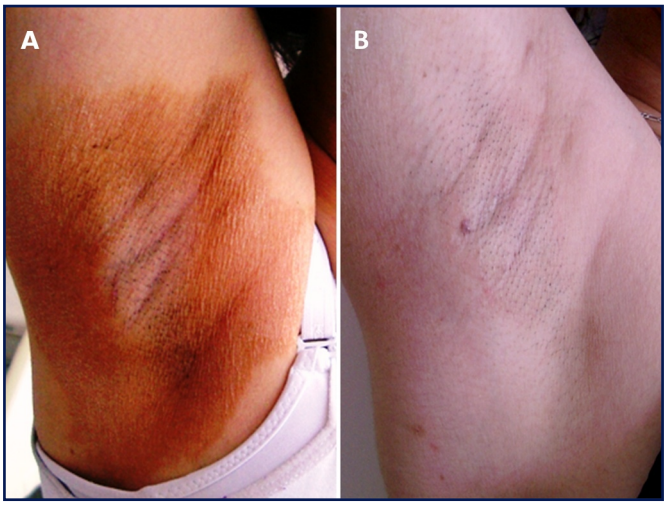

Figure 1

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

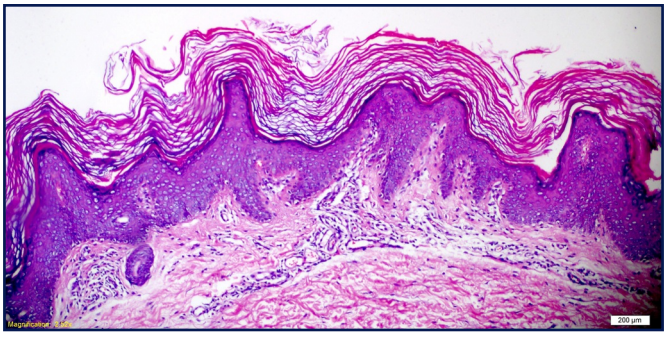

Figure 2

Histopathology (H&E, 120x) showing orthohyperkeratosis, irregular acanthosis and focal papillomatosis.

and than twice a week in order to prevent possible recurrences. But the patient used to stop the treatment when her lesions cleared within fifteen days and then started to use again when the lesions relapsed fifteen days after cessation of the ointment (fifteen days on and fifteen days off treatment).

Discussion

PSEK is characterized by symmetrically distributed, very slowly progressive erythtematous, scaly plaques. The plaques may progress during childhood and then are usually stable.[2,4,5] Palmoplantar keratoderma can be seen in half of the cases and may regress after puberty. Both sexes are equally affected and patients are otherwise entirely normal.[4]

PSEK is first described by Darier in 1886 and presented in 1922 by Gottron.[5] The pathological mechanisms leading to PSEK are still not clearly understood. A frameshift mutation in the loricrin gene, a major structural component of the cornified cell envelope, have been identified.[3] The histopathology of PSEK is non-specific and usually demonstrates acanthosis, papillomatosis, a loose hyperkeratotic stratum corneum, and occasionally parakeratosis.[2]

Treatment options are topical retinoids, emollients, keratolytics and topical corticosteroids with limited or no success.[2,6] Oral retinoids have been shown to be successful in some cases, but recurrence is to be expected on cessation of therapy, and side effect profile limits their long-term use.[2] Topical calcipotriol is an established mode of treatment for psoriasis. It is also reported to be effective in many hyperkeratotic skin diseases, such as nevoid hyperkeratosis of the nipple[7], inflammatory linear verrucous epidermal nevus[8] and confluent and reticulated papillomatosis.[9,10] Calcipotriol inhibits cellular proliferation and induces differentiation of keratinocytes.[8] Presence of hyperkeratosis in PSEK, led us to recommend our patient to use topical calcipotriol and we observed a remarkable improvement. The mechanism of action of topical calcipotriol in PSEK is probably due to its effects on keratinocyte differentiation and proliferation.

Conclusion

When compared with the other treatment modalities, topical calcipotriol is a safe and effective drug with minimal side effects and it would be a good alternative. To our knowledge, this is the first report that calcipotriol ointment is recommended for the treatment of PSEK.

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