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Acitretine Treatment of a Case with Progressive Symmetric Erythrokeratoderma

Güldehan Atış[1], MD, Ersin Aydın[2], MD

Address: [1]University of Health Sciences, Haydarpaşa Numune Trainig and Research Hospital, Department of Dermatology

[2]Tem Hospital, Department of Dermatology

E-mail: guldehan.atis@gmail.com

Corresponding Author: Dr.Güldehan Atış, Tıbbiye cad. Haydarpaşa Numune Eğitim ve Araştırma Hastanesi Dermatoloji Kliniği Üsküdar, İstanbul, Turkey

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Abstract

Observation: Progressive symmetric erythrokeratoderma (PSEK) is a rare herediter genodermatosis. Symmetrical, stable, erythematous and well demarcated hyperkeratotic plaques are common skin finding for PSEK. Treatment agents are topical retinoids, emollients, keratolytics and topical corticosteroids with limited improving . Herein we report a 22-year-old male patient with PSEK who was successfully treated by acitretine without serious side effect.

Introduction

Progressive symmetric erythrokeratoderma (PSEK) or Gottron's disease is a rare genodermatosis. Approximately 100 cases were reported in the literature to date [1]. The inheritance of PSEK may vary, however, the most common pattern is autosomal dominant. Loricrin or connexin mutations have been identified in some cases with PSEK [2]. Sporadic mutations are observed approximately 40% of patients with PSEK [3]. Symme trical, stable, erythematous and well demarcated hyperkeratotic plaques are common skin findings of PSEK. Palmoplantar involvement is seen in about 50% of patients [4]. Treatment agents are topical retinoids, emollients, keratolytics and topical corticosteroids with limited improving [5]. Herein we report a patient with PSEK was successfully treated by acitretine.

Case Report

A 22-year-old man admitted to our dermatology outpatient clinic with pruritic symmetric hyperkeratotic plaques on the antecubital fossa, elbow, back and axilla since his birth (**Figures 1 and 2**). He told that pruritus reduces partially in winter months. The lesions were non-migratory. There was no family history. Palmoplantar involvement was not observed. There was no abnormality of hair, nail or mucosal membranes. Systemic examination was normal. Skin punch biopsy was pe rformed. On histopathological examination; epidermal hyperplasia, mild papillomatosis, irregular acanthosis, hyperkeratosis and parakeratosis were detected. Based on clinical and histopathological findings, the patient was diagnosed with PSEK. Complete blood count, liver enzymes and lipid profile were in normal limits. 25 mg/day acitretin was started. At the end of the first month, all the lesions healed completely. The dose was reduced to

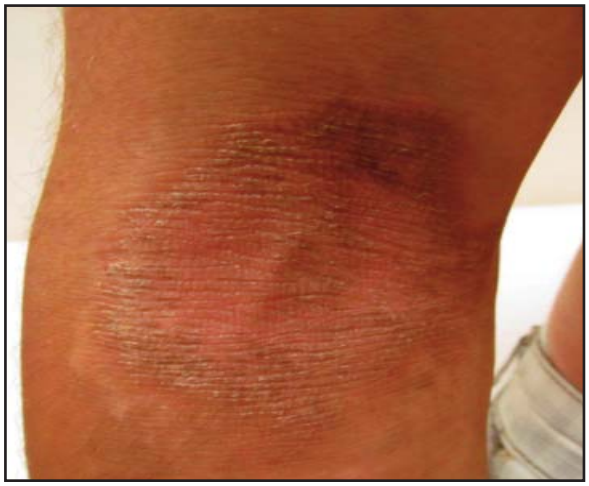

Figure 1. Hyperkeratotic plaque on the antecubital fossa.

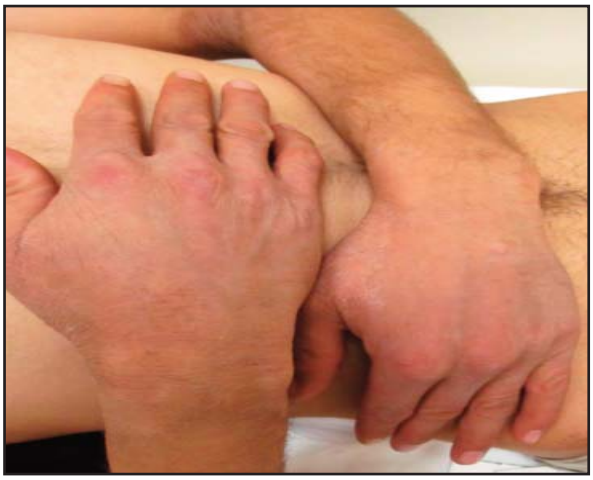

Figure 2. Symmetric hyperkeratotic plaques on the hands.

10 mg/day. The maintenance dose has been continued and he has remained free of the symptoms to date.

Discussion

Erythrokeratodermas are a rare heterogenic genodermatosis characterized with increased epidermal cell proliferation [**6**]. Some of syndromes such as KID (keratitis, ichthyosis, dea fness) and HID (hystrix like, ichtyosis, deafness) can associate with erythrokeratodermas. Two major non-syndromic types were descripted as PSEK and erythrokeratoderma variabilis (EVK) [**4**]. Well demarcated, fixed, erythematous, hyperkeratotic, scally plaques distribute symmetrically on elbows, buttocks, dorsal hand and feet and rarely face in PSEK. Emotional stress, sun exposure, increase temprature, friction may lead exacerbation of lesions [**6**]. Pruritus is reduced partially in winter months in our patient. Pityriasis rubra pilaris, psoriasis vulgaris, especially EVK should be differentiated from PSEK. The lesions are migratuar feature in EVK and it is associated with greater incidence of palmoplantar keratoderma than PSEK [**4**]. Our patient has no n-migratory skin lesions therefore we differentiate EVK. Based on histopathological examination, the patient was diagnosed with PSEK.

Progressive symmetric erythrokeratoderma is characterized by progressive and permanent symmetrically erythtematous plaques. Isotretinoin, emollients, keratolytic agents, topical corticosteroids and topical calcipotriolare used as treatment agents for PSEK in literature [**5**]. Acitretin is a synthetic retinoid which is well-known medication for psoriasis [**7**]. It has antiproliferative and immunomodulatory effects. Providing kerotinocyte differentiation and inhibition of vascular endothelial growth factor from keratinocyte are other important therapeutic effects of acitretin [**8**]. It is suggested for treatment of many hyperkeratotic skin diseases such as palmoplantar lichen planus, hypertrophic lichen planus, and palmoplantar keratoderma. [**9, 10, 11**]. Additionally, erythrokeratoderma variabilis was treated with acitretin successfully [**12**]. Praphou et al. reported an adult onset PSEK patient who was treated with 25 mg/day acitretin [**13**]. In our patient, 25 mg/day acitretin treatment was started because of the hyperkeratosis on his histopathological findings. A remarkable improvement was observed and all the lesions healed completely at the end of the first month without side serious effect. Acitretin probably improves the hyperkeratosis and maturation of keratinocyte. Also retinoids may suppress the expression of mutant loricrin [**3**].

Conclusion

Acitretin is effective treatment agent patients with PSEK. Acitretin provide healing the lesions completely without serious side effect.

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