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# Progressive symmetric erythrokeratoderma with seasonal variation - an unusual case report

Kikkeri Narayanasetty Naveen[1], Varadraj V Pai[1], Ramnathpur Achar Divyashree[1], Sharatchandra.B.Athanikar[1], Ravikala Rao[2].

[1] Department of Dermatology, Sri Dharmasthala Manjunatheshwara College Of Medical Sciences & Hospital (SDMCMS&H), Sattur, Dharwad - 580009

[2] Department of Pathology, Sri Dharmasthala Manjunatheshwara College Of Medical Sciences & Hospital (SDMCMS&H), Sattur, Dharwad - 580009

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**Corresponding Author:** Dr. Kikkeri Narayanasetty Naveen .

**E-mail:** naveenkn80@yahoo.com

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## Abstract

Progressive symmetric erythrokeratoderma (PSEK) is a rare genodermatosis with predominantly autosomal-dominant inheritance, characterized by well demarcated, erythematous, hyperkeratotic plaques that are symmetrically distributed over the extremities and buttocks, and often the face. Like erythrokeratoderma variabilis, it doesn't have seasonal variation. Here we are presenting a classical case of PSEK having autosomal recessive inheritance with seasonal variation.

## Introduction

Erythrokeratoderma (sometimes erythrokeratodermia) is the association of localized hyperkeratotic plaques with overlapping or distinct areas of circumscribed erythema. Many associations and possible syndromes are reported. [1] Erythrokeratoderma has both clinical and genetic heterogeneity. Currently, erythrokeratodermas are divided into two major subtypes: EKV (erythrokeratoderma variabilis) and PSEK (progressive symmetric erythrokeratoderma). [2]

Here we are presenting a classical case of progressive symmetric erythrokeratoderma with seasonal variation.

## Case history

A 22 year old male, product of second degree consanguineous marriage, presented with palmo-plantar keratoderma and skin lesion. According to his parents' statement the palmo-plantar keratoderma started at the age of one year and noticed the skin lesions at the age of seven. It initially started on the dorsa of hands and feet and spread over a period of 2-3 months to involve the legs, knees and elbows and remained static thereafter. The condition waxed and waned yearly without complete resolution and became worse during winter season and nearly resolved by summer leaving hyperpigmented lesions. There was no other triggering factor involved and detailed history revealed no other affected members of the family tree.

On examination multiple, well demarcated, bilaterally symmetrical erythematous plaques with fine scaling of different geographical figures were present over the dorsa of hands **(fig 1)**, foot, legs **(fig 2)** and elbow region with palmo-plantar keratoderma **(fig 3)**. Abdomen, trunk, flexural areas and face were spared. The scalp, nail and mucous membrane were normal.

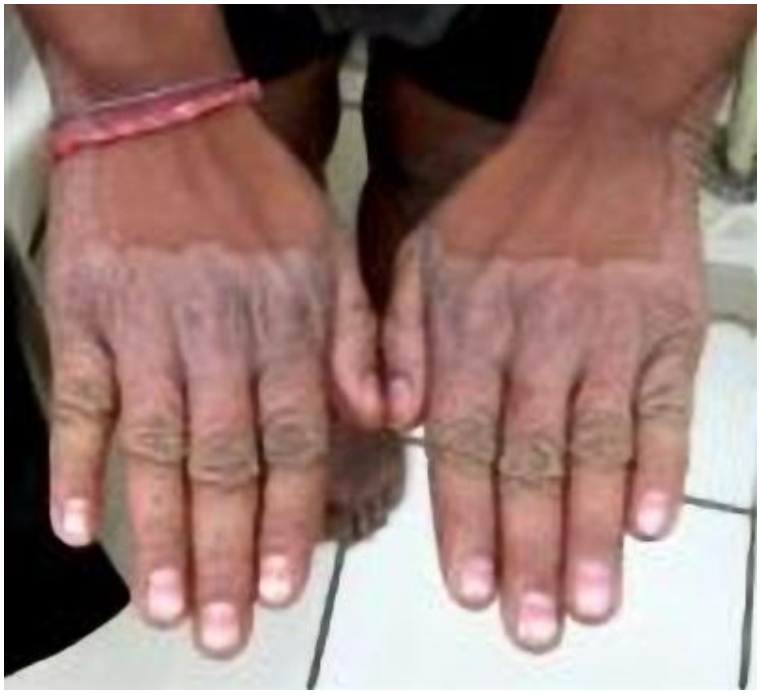

**Fig 1:** symmetric erythematous plaque on dorsum of hand.

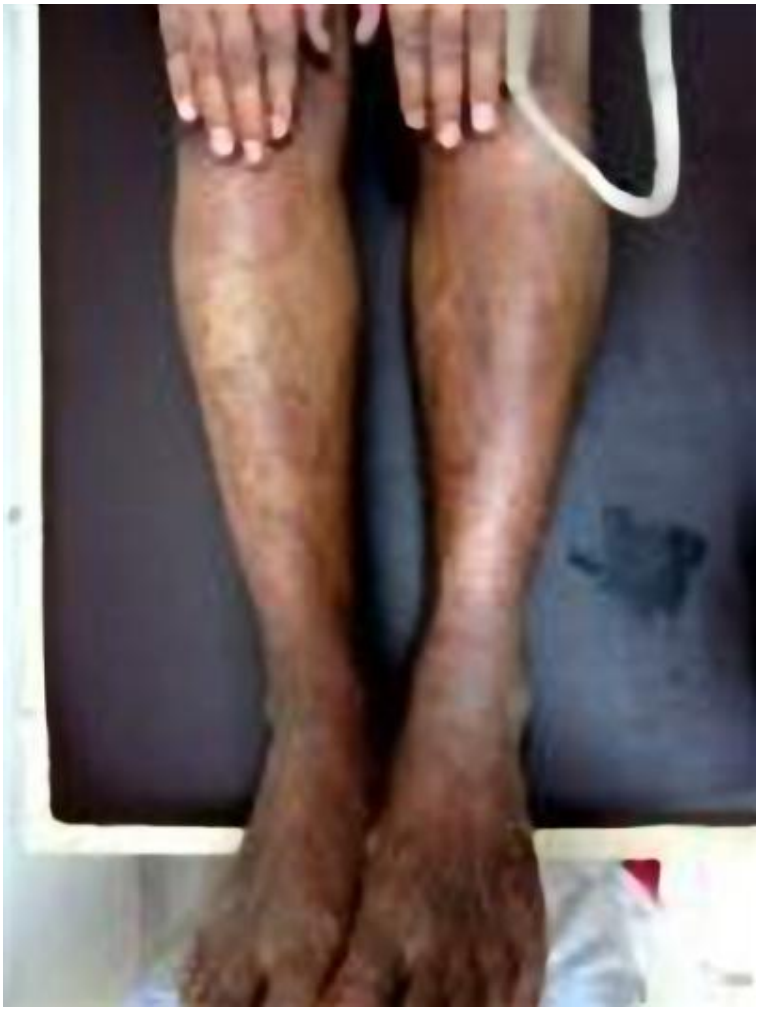

**Fig 2:** symmetric plaques on legs.

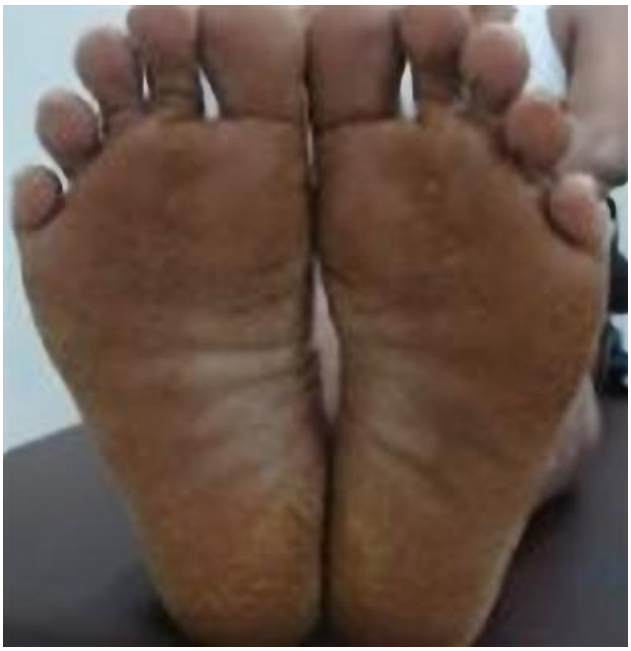

**Fig 3:** plantar keratoderma.

The general and systemic examinations of the patient were within normal limits. Histopathology from the plaque over the knee region revealed hyperkeratosis, normal granular layer and acanthosis with broad rete ridges and in the dermis perivascular lymphocytic infilteration **(fig 4)**.

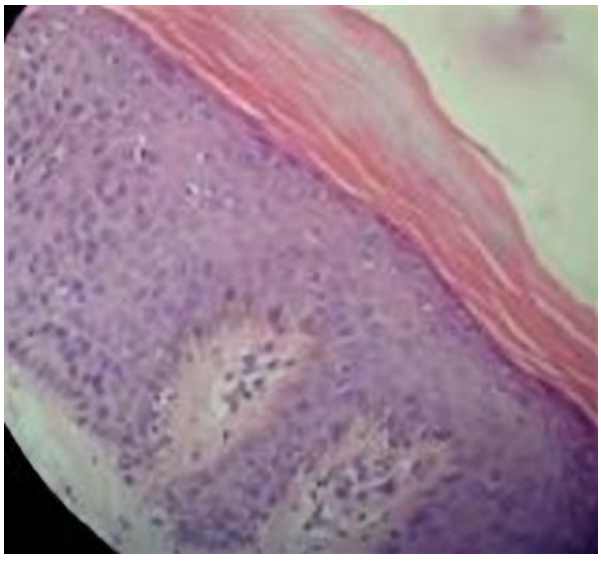

**Fig 4:** Histopathological examination revealing hyperkeratosis, normal granular layer and acanthosis with broad rete ridges and in the dermis perivascular lymphocytic infiltration (H&E. 40x)

A diagnosis of Progressive symmetric erythrokeratoderma was made clinically and was consistent with the histopathological report.

## Discussion

PSEK is a rare genodermatosis with predominantly autosomal-dominant inheritance, although autosomal-recessive transmission has been also observed.[3] Progressive symmetric erythrokeratodermia was first described by Darier in 1911 and is characterized by well demarcated, erythematous, hyperkeratotic plaques that are symmetrically distributed over the extremities and buttocks, and often the face. The trunk tends to be spared, but palms and soles may be involved. [4] Onset usually occurs during infancy with the development of slowly progressive non- migratory erythematous plaques with scale. The plaques tend to progress during childhood, with lesions stabilizing thereafter. [2]

Genetic analyses of large affected families, in conjunction with studies in transgenic mice, have suggested the loricrin gene is associated with the features of this disease. Loricrin is the major structural component of the cornified cell envelope that is formed beneath the plasma membrane of stratified squamous epithelial cells during terminal differentiation. [5]

The diagnosis is invariably clinical, as loricrin gene analysis is difficult in hospital settings. [4]

Erythrokeratoderma variabilis (EKV) which may resemble PSEK differs from the latter, as the lesions of EKV continuously changes and may be induced by external mechanical pressure and temperature changes. [6] Progressive symmetrical erythrokeratoderma (PSEK), resemble those of EKV, but migratory erythema is not seen. Facial involvement and palmoplantar keratoderma are more characteristic of PSEK. [2] In our case there was an autosomal recessive pattern of inheritance, with palmoplantar keratoderma since infancy and symmetric lesions over skin with waxing and waning without complete resolution.

In conclusion, we hereby present a rare and a classical case of progressive symmetric erythrokeratoderma with seasonal variation.

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