# SYMMETRICAL PROGRESSIVE ERYTHRO - KERATODERMA

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*A 13-year-old male child had gradually progressive, bilaterall, symmetrical, erythematous hyperkeratotic plaques over knees, elbows, natal cleft, dorsa of hands and feet with palmoplantar keratoderma. High arched palate, fissured tongue and sternal depression (pectus-excavatum) were unusual associations.*

Key Words : Symmetrical, Keratoderma

## Introduction

Symmetrical progressive erythrokeratoderma or Gottron's syndrome is usually inherited in an autosomal dominant fashion. Sporadic cases are frequent. It starts in childhood and is characterised by large, fixed geographical and symmetrical fine scaly plaques of erythema. Shoulder girdle, cheeks, buttocks are often affected with limited plaques on ankles and wrists.[1] In a few cases Koebner phenomenon is also seen.[2] We are reporting a case of symmetrical progressive erythrokeratoderma who had associated high arched palate, fissured tongue and pectus excavatun.

## Case Report

A 13-year-old male child presented with well defined , erythematous, scaly, bilaterally symmetrical, gradually progressive, plaques over knees and elbows for 10 months. Similar lesions were present over natal cleft, and hyperkeratotic plaques over dorsal aspect of fingers and toes were present for the same duration. Palms and soles showed keratoderma with fissuring. This child also had high arched palate, fissured tongue and sternal depression (pectus excavatum). Hair, teeth and mucosae were normal. None of the family members had similar disease.

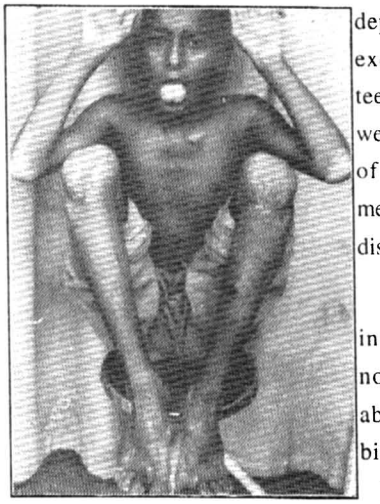

Fig 1. Hyperkeratotic plaques on knees and elbows, fissured tongue and sternal depression.

Routine investigations did not reveal any abnormality. Skin biopsy taken from lesion on the knee showed hyperkeratosis, focal acanthosis and papillomatosis. Dermis showed focal non specific inflammatory infiltrate.

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## Discussion

Onset in childhood, bilaterally symmetrical, asymptomatic erythematous hyperkeratotic, progressive plaques suggest the diagnosis of symmetrical progressive erythrokeratoderma. There was no family history in our case. Positive family history can be obtained in only about 50 percent patients, rest of cases are due to spontaneous mutation.[3]

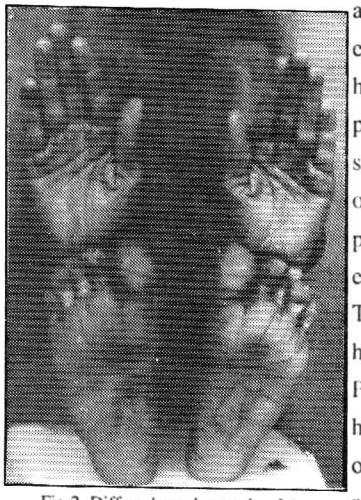

Fig 2. Diffuse hyperkeratosis of the palms and soles

Association of high arched palate, fissured tongue and pectus excavatum seen in our case was unusual and was not seen in earlier case reports.[4-6]

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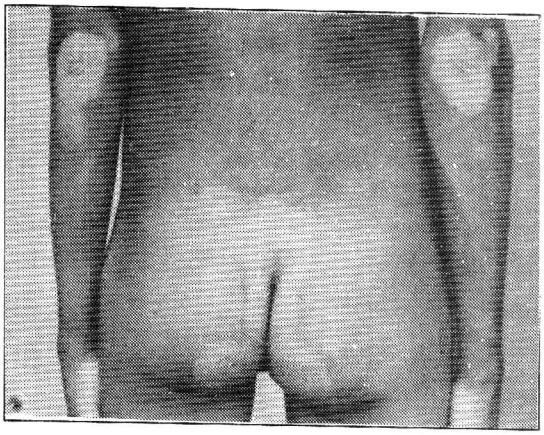

Fig 3. Lesions on natal cleft.