

# SYMMETRICAL PROGRESSIVE ERYTHROKERATODERMA (Gottron's Syndrome) A Case Report

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## Summary

A case of symmetrical progressive erythrokeratoderma is reported in a young male.

**KEY WORDS:** Symmetrical progressive erythrokeratoderma, Gottron's Syndrome.

Gottron's syndrome or symmetrical progressive erythro-keratoderma, is a rare autosomal dominantly inherited disorder of keratinisation characterised by asymptomatic hyperkeratotic plaques, distributed bilaterally and symmetrically on the dorsal aspects of hands, feet, legs, forearms and sometimes on other parts of the body. The palms and soles are spared<sup>1</sup>. The first lesion usually appears in infancy but the onset may be delayed until adult life. Here, we are reporting a case of symmetrical progressive erythrokeratoderma in a young man. To the best of our knowledge, this is the second case report from India on symmetrical progressive erythrokeratoderma, the first one being reported by Kapur et al in 1978<sup>2</sup>.

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## Case report

A 24 years old male, the only son of non-consanguineous parents, was seen in the dermatology department of Medical College Hospital, Kottayam in September 1982, with asymptomatic hyperkeratotic plaques on his limbs which he noticed first at the age 16 years. The lesions had been progressing since then. Patient's mother, who was examined later by us, also had similar skin lesions in the same distribution. There was no history to suggest chronic inflammation or repeated trauma at these sites. General physical examination did not reveal any abnormality. Dermatological examination revealed multiple bilateral symmetrical erythematous hyperkeratotic well defined plaques on the dorsa of the hands and feet, on the elbows and knees and also on the tendo Achillis bilaterally (Fig-1,2,3). There were no silvery micaceous scales on rubbing the plaques with a glass slide and Auspitz sign was negative. Hairs and nails were normal. All other systems were clinically normal.

Routine urinalysis and haemogram were within normal limits. Blood



**Fig. 1**  
Symmetrical progressive erythrokeratoderma. Note hyperkeratotic plaques on the knees and elbows.

VDRL test was non reactive. Repeated scrapings in KOH did not show any fungal filament. Histological study of the lesion from the foot revealed marked hyperkeratosis, irregular acanthosis and sparse mononuclear cell infiltration of the upper dermis. The

patient was prescribed salicylic acid ointment but there was little improvement over a period of 2 months.

### Discussion

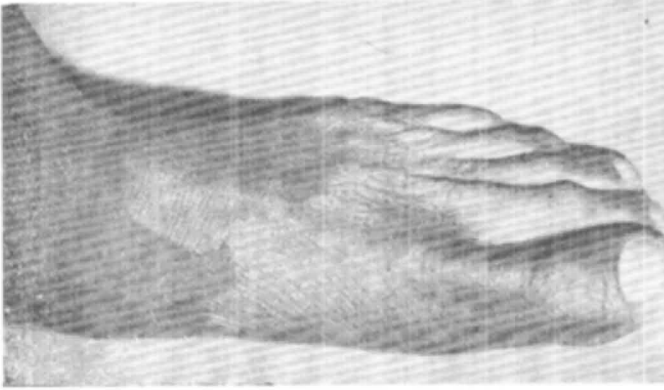
In the present case all the clinical features and family history suggest a diagnosis of symmetrical progressive erythrokeratoderma. It is unlikely to be erythrokeratoderma variabilis, in which the plaques assume bizarre geographic configurations and in short time (hours and days) change their shape and size or involute completely<sup>3</sup>. But the lesions in our case were bilateral, symmetrical and fixed in nature. Psoriasis is excluded, in the present case, by the absence of characteristic micaceous scales and a negative Auspitz sign. The absence of nail changes also mitigates against psoriasis. Histology also did not show any evidence of psoriasis. The sparing of the palms and soles excluded the recessive form of palmoplantar keratoderma (mal de Melada) in which condition also similar keratotic plaques may be noted on the limbs.

### References

1. Ebling FJ & Rook A: Disorders of Keratinisation, Text book of Dermatology, Vol II, 3rd Ed, Edited by Rook A, Wilkinson DS & Ebling FJG, Blackwell Scientific Publishers, London, 1979; 1253.

**Fig. 2**  
Symmetrical keratotic lesions over the Achillis tendons.





**Fig. 3**

Close up view of the well defined plaque on the dorsum of left foot. He had similar plaque on the right foot also.

2. Kapur TR, Ansari MA & Singh OP: Symmetrical Progressive Erythrokeratoderma, *Indian J Dermatol Venereol Leprol*, 1978; 44: 158-159.
3. Domonkos AN, Arnold HL & Odom RB: *Andrews' Diseases of the skin*, 7th edition, WB Saunders Company, London 1982; p 713.

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### **Announcements...**

The Gujarat State Branch solicits good wishes from all association members for a successful State level conference to be held at Rajkot on April 24, 1983.

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The 6th Regional Conference of Dermatology (ASIAN-AUSTRALASIAN) will be held in Singapore between 5.h and 9th May 1984. For details write to :

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