# MUTILATING KERATODERMA WITH DEAF-MUTISM

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A 30-year-old woman presented with typical lesions of mutilating keratoderma. The patient was deaf and dumb. Hyperkeratosis of palms and soles was present since infancy. Constriction of digits started by the age of 5 years. The clinical diagnosis was supported by histopathological examination.

**Key Words : Keratoderma, Mutilating, Deaf-mutism**

## Introduction

Mutilating keratoderma (Vohwinkel's syndrome) is a rare syndrome of hereditary palmo-plantar keratoderma. It is inherited in an autosomal dominant fashion. The keratoderma begins in infancy and is diffuse but "honeycombed" by small depressions. Later on constricting fibrous bands lead to progressive strangulation of digits. Star shaped keratoses on the dorsa of fingers and knees are distinctive.[1] Associated features may include alopecia, deafness, spastic paraplegia, myopathy and ichthyosiform dermatoses.[2] Mental retardation, bony changes and congenital deformities have also been reported in association with mutilating keratoderma.[3,4] We report a case of mutilating keratoderma with deaf-mutism.

## Case Report

A 30-year-old woman developed hyperkeratosis of palms and soles extending to distal part of forearm and legs respectively. The lesion began in infancy with thickening of palms and soles (Fig.1), but since the age of 5 years the disease rapidly progressed. Fibrous bands around fingers mostly on the little fingers were also noticed. These bands led to the constriction of digits. In the last 5 years, thickening had extended to dorsum of hands, feet and flexural aspects of both forearms. Star fish shaped keratosis on the knuckles along with isolated keratotic warty papules on the dorsum of elbows were also seen.

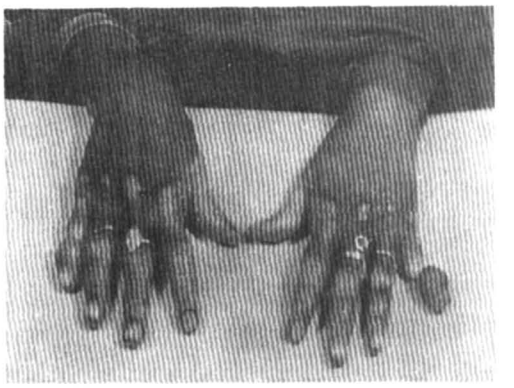

Fig. 1. Palmoplantar keratoderma with constriction bands.

The patient was deaf and dumb since birth. Hair were sparse, thin and lustreless. No other abnormality could be detected. Detailed family history did not reveal any other family member affected with similar disease.

Histology of the palmar lesion showed changes of keratoderma ie, hyperkeratosis, hypergranulosis and moderate acanthosis along with mild perivascular infiltrate of mononuclear cells.

## Discussion

A few cases of mutilating keratoderma

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have been described in the literature but extension of keratoderma into adjacent areas of dorsum of hands and feet and forearm has not been reported earlier. Diffuse thickening of palms and soles along with constriction bands on little fingers of both hands suggested mutilating keratoderma. Absence of erythema and scaling on palms and soles ruled out Mal de Meleda.

## References

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