

## POROKERATOSIS MIBELLI WITH CUTANEOUS HORNS

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The appearance of Cutaneous horns on human beings is an uncommon sight and is generally considered as a freak of nature. It becomes interesting to see such a thing developing on the top of another rare disease like Porokeratosis, which presents characteristic clinical features with a distinctive histopathological lesion.

A case admitted in this hospital showing formation of Cutaneous horns, is described.

### CASE REPORT

#### History and Condition on Examination

A male aged 45 years, a farmer, had generalised itching 25 years back, followed after few days by the appearance of very small light brown macules scattered all over the body. They grew in size in about three months to varying sizes, the maximum being about 2" in diameter. They were of varying shapes. The lesions were bounded by rough margin traversed all along by a longitudinal groove. The central parts of the lesions were of different colours - light brown, dark brown, skin coloured and hypopigmented. There was no scaling. Sensations were normal. He had lesions on scalp, face (including ears, nose, and eyelids) neck, genitalia both extremities and trunk (fig. 1). On the scalp there was some alopecia. He had no lesions on glans penis, palms, below both the

ankles, tongue and nails. The total number of lesions were more than 200.

For the last 4 years he developed intense itching over the lesions of both the lower extremities. Subsequently, the margins of the lesions began to hypertrophy enormously at several places. Twenty five lesions showed such horns, each lesion containing 1 to 5 of them (fig. 2). The base of these was discoid with a maximum diameter of about  $\frac{1}{2}$ ". Colour of the horns varied i.e., ash, yellowish brown, and brown. Surface of all the horns was rough but that on the posterior aspect of left thigh was smooth. Length of the horns varied from  $\frac{1}{2}$ " - 2". Two of them were long and pointed and slightly curved at the tip. All others were short and stumpy and rough with corrugations. They were painful on movement. The skin elsewhere was dry.

#### Family History

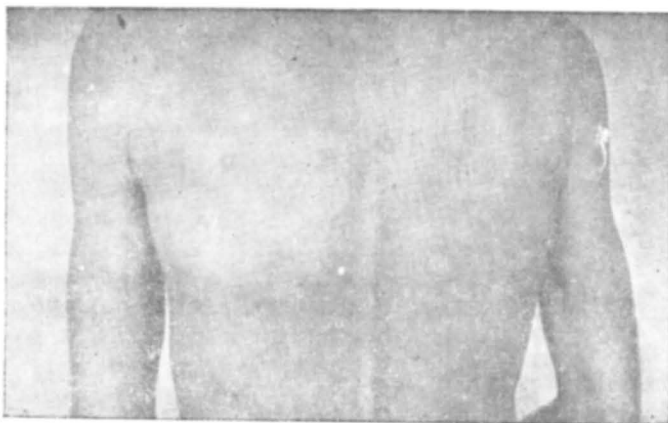
Patient had one brother aged 25 years who was said to be having similar lesions scattered all over the body. The patient in question had nine children, six males, and three females. Seven of them, five males and two females, died due to inter-current disease before they attained five years. This surviving boy was 14 years old and the girl 5 years old, both of them were so far free from such lesions. The patient did not give any history of exposure. His wife, who was not a blood relation, had no miscarriages. She was free from such lesions.

#### Investigations :

Routine blood and Urine examination did not show anything significant.

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**Fig. 1**

Circinate lesions on extremities and trunk



**Fig. 2**

Cutaneous horns



**Fig. 3**

Characteristic Coronoid lamella

Blood V.D.R.L. : Negative.

Blood Group : A. B. group.

Scrapings of the margins of lesions: Negative for fungus. Cross section of a cutaneous horn : No medulla was seen. Serum calcium and biochemical analysis of horn could not be done.

Histopathology of skin piece from right arm (fig. 3): It showed the characteristic "Coronoid lamella". The groove in the epidermis was occupied by a horny plug. In the centre of the horny plug there were prokeratotic cells.

#### Treatment :

He has been put on 'B' Complex parenteral, Vitamin A & D capsules and on salicylic acid 12% ointment.

#### Comments

The clinical appearance of the lesions in this case resembles Keratosis follicularis serpiginea, ring worm and maculoanesthetic leprosy. But the atrophic lesions with keratotic borders, presence of sensations, absence of scaling and of fungus in the scrapings of margins and the characteristic histological picture confirms the diagnosis of prokeratosis.

Cases of Porokeratosis are reported sporadically from all over the world and also from India, Venkatesan (1965). The tissues generally involved in this condition are the skin, mucous membrane and the nails. In this case, only the skin is involved. Formation of cutaneous horns is a very uncommon event in Porokeratosis. Cutaneous horns may arise from normal skin and mucous membrane or over conditions

like rodent ulcer, squamous cell carcinoma, sebaceous cysts, warts or naevi, Belisario (1959). Many cutaneous horns are hypertrophic types of actinic Keratoses and occur in elderly persons with seborrhoea and who were subjected to long continued exposure to sun, Andrews and Domonkos (1964). Common sites are the scalp, forehead, nose, cheeks, ears, lips, fore-arms, hands and penis, Belisario (1959). In this case, the horns have developed on the Keratotic borders of the lesions. The site of the horns is very unusual in that all of them are located only in the lower extremities. True cutaneous horns are medullated but the term is loosely applied to non-medullated excrescences, Andrews and Domonkos (1964). The horns in this case are non-medullated. In this case sun light might have been one of the conditions favouring the formation of horns on both the lower limbs, which are always kept exposed to sun light by virtue of his occupation.

Treatment is not of much avail since it is described as one of the diseases showing remissions and exacerbations.

#### Summary

A case of Porokeratosis is described with formation of Cutaneous horns. The condition began in a male at the age of 20 years. All the Cutaneous horns occurred only on both the lower limbs.

#### Acknowledgment

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3. Venkatesan TV : Porokeratosis of Mibelli, Ind. J. Derm, 10 : 53, 1965.

## WHAT IS YOUR DIAGNOSIS ?

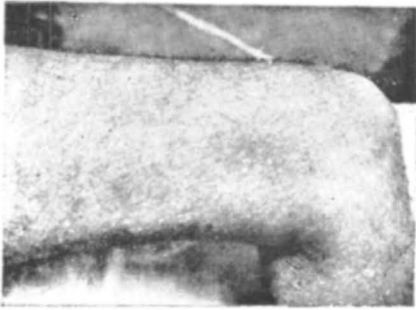


Fig. 1



Fig. 2



Fig. 3



Fig. 4

*(Question & Answer overleaf)*

Fig. 1 : “ Rain drops on a dusty road ” appearance on the skin of thigh.

Fig. 2 : Pigmented warty lesions on the back and hypopigmented spots.

Fig. 3 : ‘ Keratoses ’ on the palms

Fig. 4 : Keratotic lesions and fungating growth left sole foot.

### Differential diagnoses :

1. Xeroderma pigmentosum
2. Arsenical pigmentation and keratosis
3. Malignant Melanoma

He is a case of Chronic bronchial asthma and has taking several medicines including Ayurveda. Biopsy of a keratosis of palm revealed typical features of Arsenical Keratosis. Biopsy of the fungating growth showed Sq. Cell Ca. Final Diagnosis is Arsenical pigmentation and keratosis.

A 62 year old man came with the following manifestations of more than 2 years duration :

1. Generalised mild hyperpigmentation
2. Pea sized hypopigmented macules all over the body
3. Multiple keratosis particularly palms and soles ; and
4. Large fungating growth sole of left foot of 6 months duration.

**DIAGNOSIS**