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# POROKERATOSIS OF MIBELLI IN A FAMILY

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A case of porokeratosis of Mibelli with a zosteriform distribution in an adult male is described. His daughter too had lesions on her face. Histopathology showed the characteristic cornoid lamella.

**Key Words : Porokeratosis, Cornoid lamella**

## Introduction

Classic porokeratosis was first described simultaneously by Mibelli and Lespighi in 1893. This has an autosomal dominant mode of inheritance and is often seen in father and son or daughter. The characteristic histological change is the cornoid lamella which is a thickened column of light stained keratin with parakeratotic cells extending upwards from a notch in the malpighian layer. Other types of porokeratosis have been described viz, disseminated superficial actinic porokeratosis (DSAP) by Chernosky in 1963, porokeratosis palmaris et plantaris dissiminata by Guss in 1971, porokeratosis punctata palmaris et plantaris (PPPP) by Brown et al and porokeratosis plantaris discreta (PPD) which is thought to be essentially a plantar clavus.[1]

We are presenting a case of porokeratosis of Mibelli in a man with a zosteriform distribution and in his daughter.

## Case Report

A male patient aged 40 years came to the Dermatology outpatient complaining of patches on his body for the last 25 years. He first noticed round patches on the front of the left forearm when he was about 15 years old. This increased in number spreading upwards over a period of 1 year and then remained static. There were multiple, well difined annular lesions having a sharply raised border which was pigmented. At places

Fig.1. Porokeratosis lesions in a zosteriform pattern over the left shoulder

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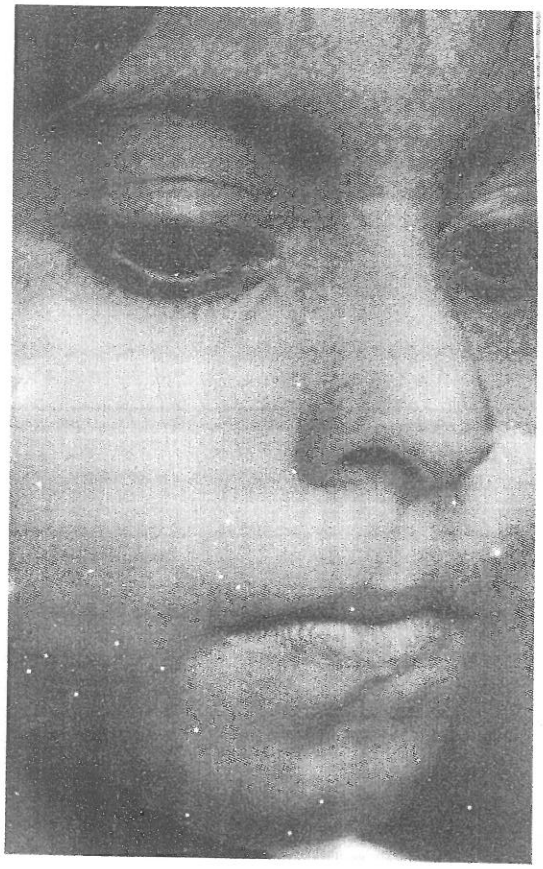

Fig. 2. Patient's daughter having the lesion on the face

the border was warty and was having a groove at the summit. The skin at the centere showed atrophy with hairloss and sweating sensation to temperature, touch and pain was normal. The lesions were arranged in a linear pattern extending upwards from the lower end of the sternum on the left side of the midline to the left shoulder joint. The upper arm was free of lesions. A second group of similar lesions was present extending down from the left elbow crease to the lower third of the forearm anteriorly. There were 3 other lesions, 2 on the right arm and 1 on the right leg.

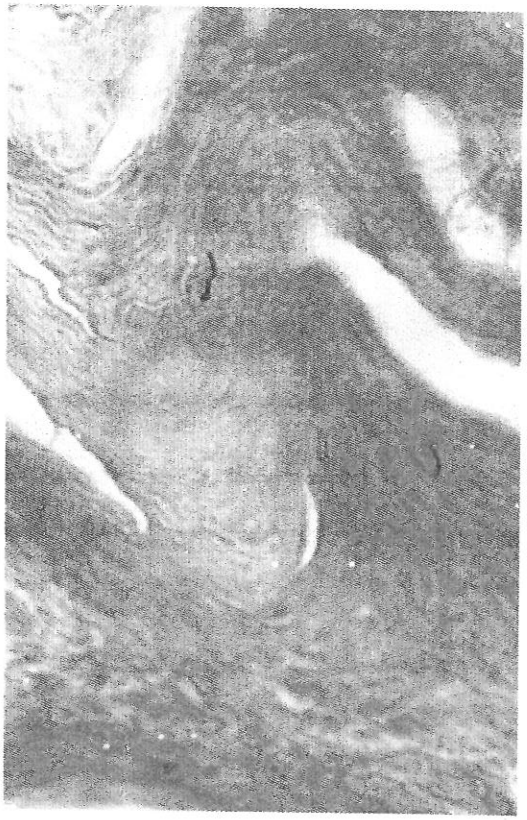

Fig. 3. Histopathology showing cornoid lamella

His daughter 10 years old had well defined tiny lesions lying adjacent to each other about a centimeter below and medial to the inner canthus of the right eye. The lesions had well defined raised border with atrophic centre.

Biopsy was done from the forearm lesion. Histology showed hyperkeratosis, mild acanthosis and non specific dermal infiltrate. The characteristic cornoid lamella was present. Biopsy was not done on his daughter for cosmetic reasons.

## Discussion

The premalignant potential of classic porokeratosis and DSAP is well known. Squamous cell corcinoma arising in a porokeratotic lesion is reported in Indian literature.[2] Association with diabetes

mellitus is also mentioned.[3] Fibroblasts cultured from classic porokeratosis have shown chromosomal instability and this may be relevant to its premalignant potintial. It is interesting to note that porokeratosis has been related to the intake of thiazides, potentially capable of producing photosensitive type of drug reactions. Patients with porokeratosis should be instructed to avoid excessive sunshine, use sunscreens and have periodic examinations by a dermatologist.[1]

Treatment consists of keratolytic ointment application. 5-fluorouracil and etretinate have been tried. Cryosurgery ($CO_2$ laser), electrocautery and excision can also be done.

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