Porokeratosis of Mibelli

- first reported case from Kano, Nigeria

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ABSTRACT

Porokeratosis of Mibelli is a rare genodermatosis with an autosomal dominant mode of inheritance. Males are more often affilicted than females and the onset of disease is usually during childhood. We describe the typical *Mibelli* type porokeratosis in a 12 year old boy, the first from our centre.

INTRODUCTION

THE porokeratoses are a group of hyperkeratotic skin disorders that are characterized clinically by an elevated, thread-like border that expands centrifugally and histopathologically by the presence of cornoid lamellae. Five variants are well-recognized variants. Porokeratosis of Mibelli is the prototype of the group and the commonest.

CASE REPORT

A 15-year-old boy presented with multiple enlarging asymptomatic lesions on the dorsum of his left foot, over the right knee and the right cheek. These first appeared as small flesh-coloured papules when he was 5years old. The lesions subsequently enlarged centrifugally. He denied any symptoms associated with the plaques but noted the lesions got scaly when he was outdoors. He had no other similar lesions elsewhere and no family members were affected.

Results of physical examination revealed multiple well-circumscribed plaques with a hypopigmented atrophic centre over the right knee (figure 1),on the left cheek (figure 2) and on the dorsum of the right foot (figure 3). The lesions had a prominent peripheral hyperkeratotic ridge.

Biopsy specimen of the peripheral ridge and the central atrophic areas revealed a slight acanthosis with invaginations filled with columns of parakeratosis, forming the cornoid lamella, next to the orthokeratotic stratum corneum of the adjacent epidermis with loss of the granular layer with focal lymphocytic infiltrate (figure 4).

The patient was given 3% salicylic acid with bethametasone to apply twice daily. There was no remarkable response. Oral acitretin was prescribed but non-availability in the country precluded its use.

Excision was planned for a later date, but the patient absconded.

The aetiology of porokeratosis is still unknown but it has been suggested that the presence of helper T cells and some Langerhans cells in Mibelli's porokeratosis is evidence for immunological mechanisms induced by antigen presentation.

DISCUSSION

Classic porokeratosis was described by Mibelli in 1893 as one or more localized, gradually progressive, hyperkeratotic, irregular plaques with central atrophy and a prominent peripheral keratotic ridge.[1]There are five well-recognized variants, these include porokeratosis of Mibelli, punctate porokeratosis, linear porokeratosis, porokeratosis palmaris plantaris et disseminata, and disseminated superficial porokeratosis.[2] All the five clinical variants are associated with autosomal dominant inheritance.[1,2] The aetiology of porokeratosis is still unknown but it has been suggested that the presence of helper T cells and some Langerhans cells in Mibelli's porokeratosis, as in this present case, is evidence for immunological

Fig. 1

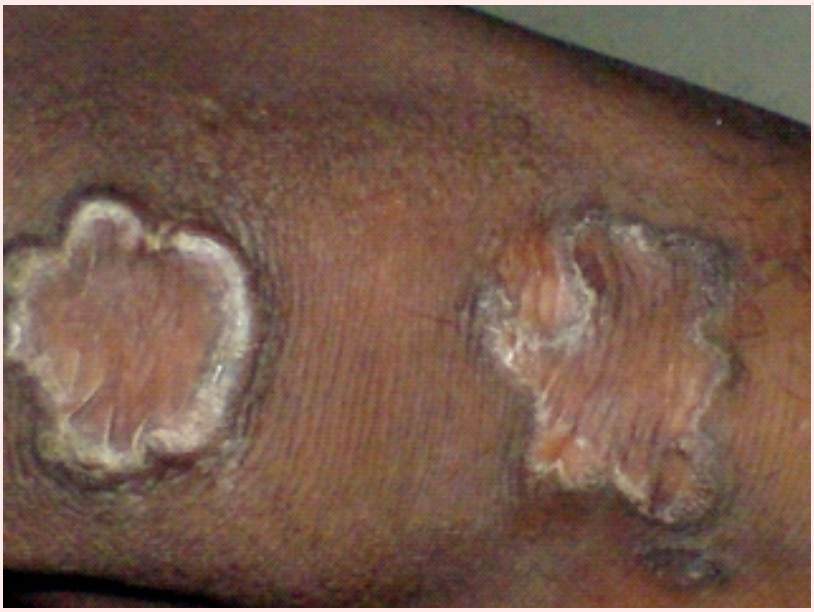
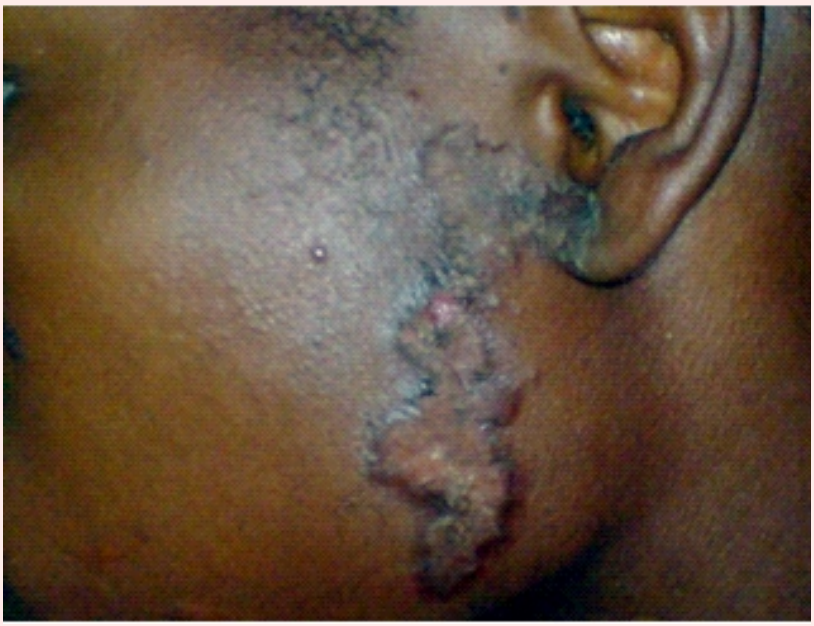


Fig. 1



FIGURES (1 & 2): Classic lesions of Porokeratosis showing the hypopigmented atrophic centre with prominent raised peripheral hyperkeratotic ridges over the right knee and the left side of the face

FIGURE 3: Linear distribution of porokeratotic plaques on the right lower limb.

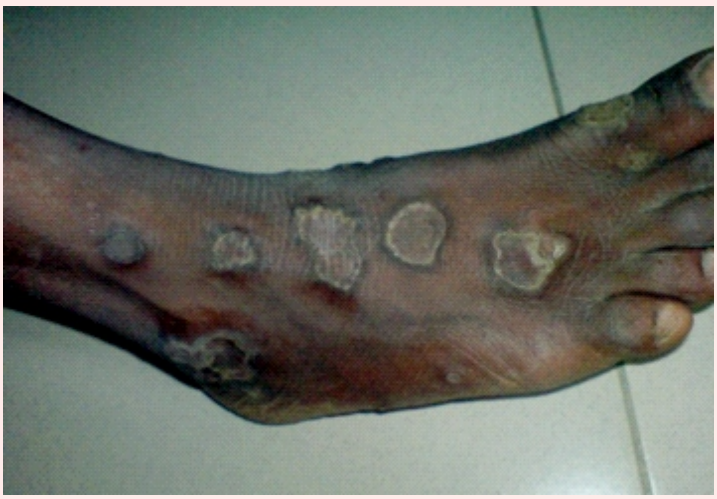
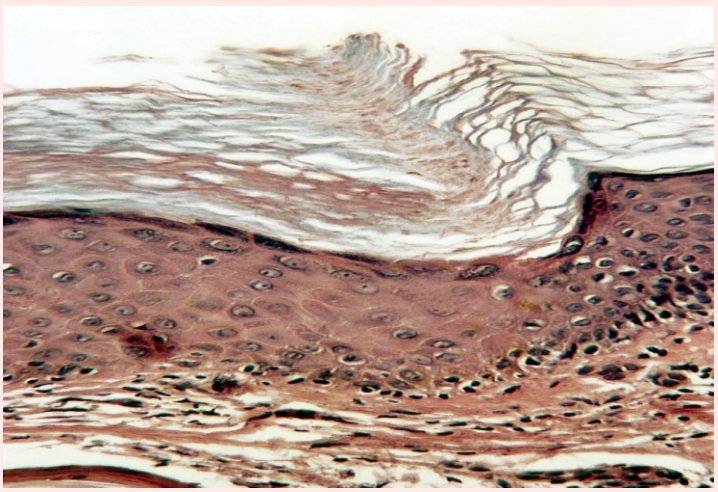


FIGURE 4: Typical cornoid lamellae (H&E, original magnification x 200)



mechanisms induced by antigen presentation.[3,4] Several trigger factors have been suggested these include irradiation, infective agents, trauma and immunosuppression.[2,3]

The usual clinical presentation of this disease is keratotic papules of various sizes, which may coalesce in plaques with irregular boundaries, characterised by a raised border with a well defined longitudinal furrow.[2] The lesions are generally unilateral, involving predominantly the distal part of the limbs, thighs and perigenital region, although they may also appear on other parts of the body including the face.[2,5] Malignant transformation in porokeratosis lesions is more frequent on non-exposed skin.[2]

Porokeratosis has been associated with the development of skin malignancies including Bowen disease, squamous cell carcinoma, and basal cell carcinoma.[6,7]

The differential diagnosis for a plaque of the extremity of similar description may include elastosis performancehb serpiginosa, lichen sclerosus et atrophicus, lichen planus, plaque stage of cutaneous T-cell lymphoma, or punctate keratoderma.[8]

Histologically, the epidermis showed a slight acanthosis with invaginations filled with columns of parakeratosis, forming the cornoid lamella, next to the orthokeratotic stratum corneum of the adjacent epidermis.[9]

Treatment options for PM include excision, cryotherapy, electrodesiccation, dermabrasion, carbon dioxide laser and topical 5-flurouracil.[1,10] Successful treatment also has been observed by using imiquimod cream 5%,[11] and oral retinoids, however, relapse after discontinuation of the medication has been reported.[12] ■

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