Porokeratosis - Head to toe: An unusual presentation

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ABSTRACT

Porokeratosis is a disorder of epidermal proliferation in which many different clinical variants can be distinguished. Herein we report a rare case of porokeratosis involving almost all parts of the body with simultaneous occurrence of various forms of the disease in the same individual (classical porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, hypertrophic porokeratosis, and genital porokeratosis).

Key words: Porokeratosis, rare coexistence, uncommon sites

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INTRODUCTION

Porokeratosis is a rare autosomal dominant type of genodermatosis featuring abnormal epidermal keratinization.[1] Classically, five clinical variants are recognized: Classic porokeratosis of Mibelli (CPM), disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminate, and linear porokeratosis.[2] There are also reports of atypical presentation like hypertrophic variety of porokeratosis.[3] Scalp, mucous membrane, palms, soles, inguinal folds, and genitalia are not commonly involved.[4-6]

CASE REPORT

A 20-year-old male patient presented with multiple, itchy and hyperpigmented annular lesions over scalp, face, neck, oral mucosa, trunk, extremities, genitalia, palms, and soles of two years duration. There was a history of photosensitivity. Initially, the lesions appeared over face and neck, gradually spread to involve the scalp, oral mucosa, extremities, trunk, groin, genitalia, palms, and soles. To begin with, the lesions were small keratotic hyperpigmented papules that subsequently enlarged to form ring like plaques with central atrophy surrounded by a discrete ridge. On family screening, it was found that his mother and brother had similar lesions.

Cutaneous examination revealed multiple, superficial, annular, hyperpigmented, atrophic plaques 1 to 3 cm in diameter with a slightly raised peripheral border over the face and neck [Figure 1]. Lesions over the front of the right ear, palms, and soles showed typical furrows [Figures 2 and 3]. Verrucous annular plaques with central atrophy were noted over the right forearm [Figure 4], scrotum and penis [Figure 5]. The lesion over the scalp was associated with alopecia [Figure 6]. A milky white, raised, cord-like lesion with maceration was seen over mucosal surface of the lower lip [Figure 7]. A vitiliginous depigmented macule was present over glans penis and mucosal surface of prepuce [Figure 8]. Dermoscopic examination showed a "white track" structure with brown pigmentation in the inside of the track [Figure 8]. A provisional diagnosis of porokeratosis was made, which was confirmed by histopathological examination [Figures 9 and 10].

DISCUSSION

Mibelli described classical porokeratosis in 1893.[4] Porokeratosis can occur with a genetic predisposition and it can also be secondary to risk factors such as sun exposure, immune suppression, and ultraviolet exposure. The incidence of Bowen's disease and squamous cell and spinous cell carcinomas is 6.8-11% in this disease, hence follow-up is essential. The risk is greater if the patient has linear-type porokeratosis, or a large or long-standing lesion.[7] Etiopathogenesis of porokeratosis is obscure, but certainly complex and multifactorial. It has been

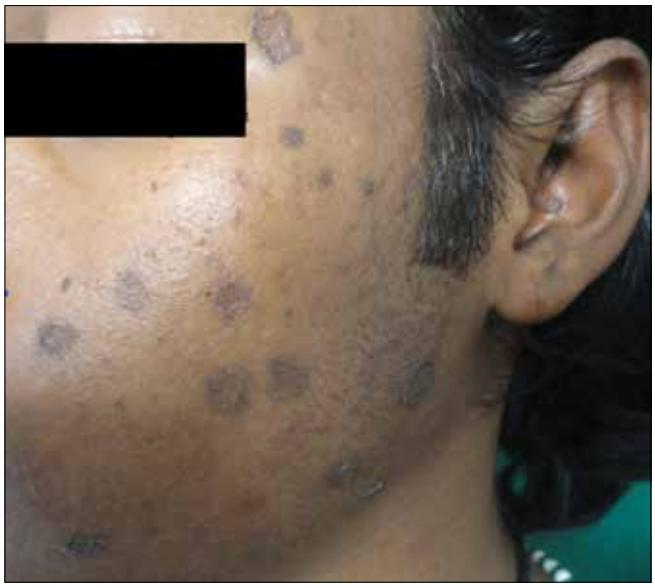

Figure 1: Annular hyperpigmented atrophic macules with slightly raised peripheral border over the face and neck

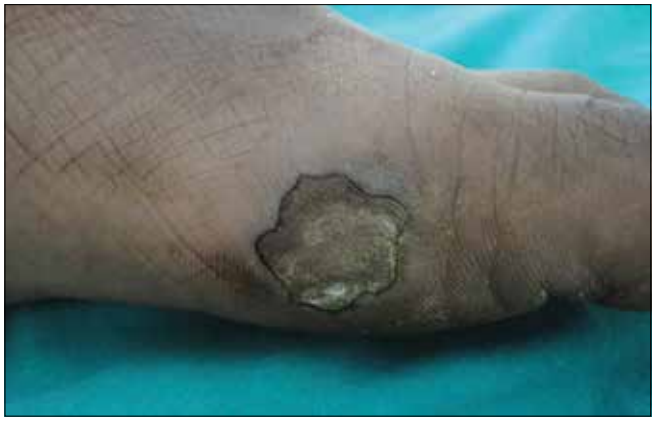

Figure 3: Classical porokeratotic lesion of Mibelli over the medial border of left foot

Figure 5: Multiple verrucous annular plaques seen over the groin, scrotum and penis

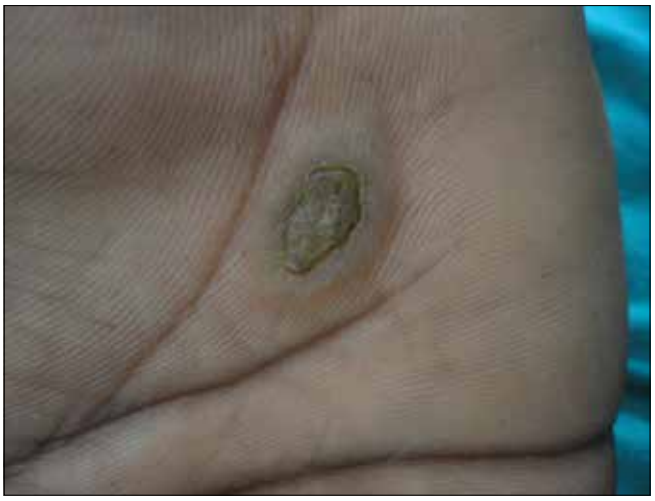

Figure 2: Lesion over the palms showing typical furrow

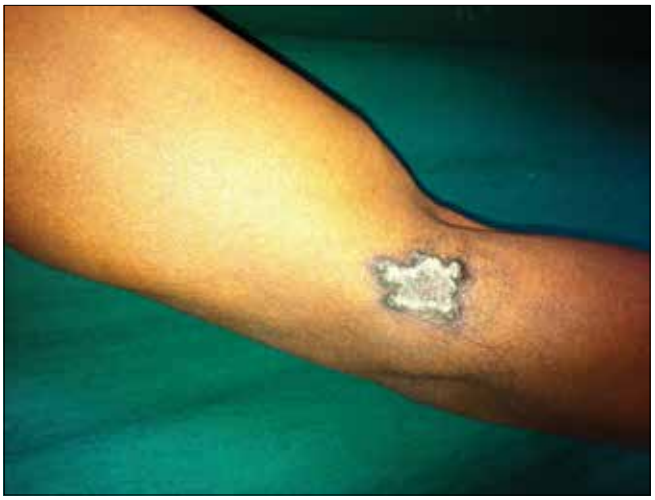

Figure 4: Single, verrucous annular plaque with central atrophy over the right forearm

Figure 6: Scalp lesion with associated alopecia

suggested that the lesions of porokeratosis result from the peripheral expansion of an abnormal, mutant clone of epidermal

Figure 7: Vitiliginous depigmented macule present over glans penis and under the surface of the prepuce with a porokeratotic lesion

Figure 8: Dermoscopic examination showed a "white track" structure with brown pigmentation in the inside of the track

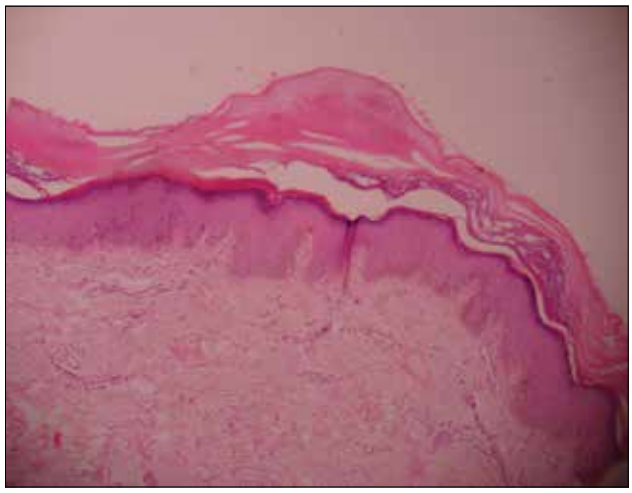

Figure 9: Histopathology in low power view showing parakeratotic column present superficially, overlying an effaced epidermis (H and E, ×10)

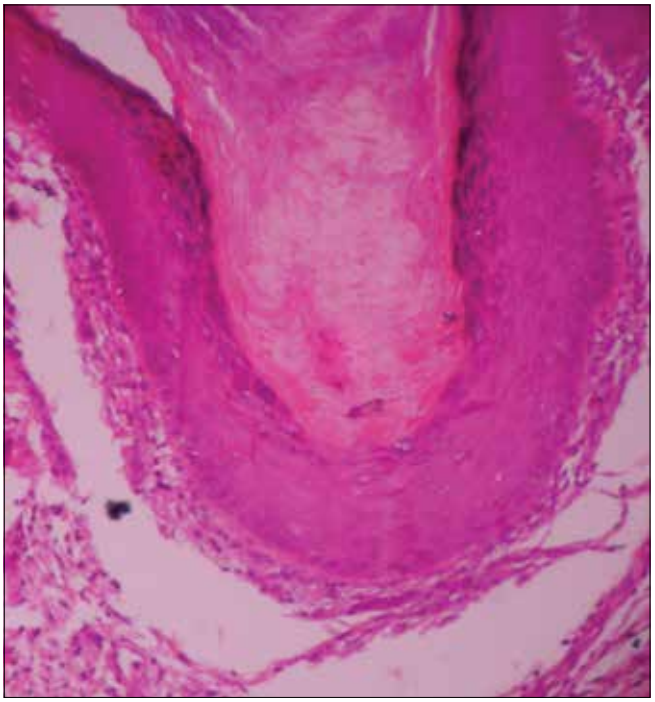

Figure 10: Skin biopsy showing the characteristic coronoid lamellae (H and E, ×100)

keratinocytes (which would be inherited) located at the base of the parakeratotic column.[8]

Clinically, the basic lesion is a crateriform horny papule that gradually extends to form a circinate or gyrate plaque with a distinct ridge like border and atrophic center. Histopathologically, it is characterized by coronoid lamella.[9] Though the lesions of porokeratosis may involve any part of the body, lesions on the scalp, mucous membrane, palms, soles, and genitalia are regarded uncommon.[4-6] There are various reports of coexistence of multiple variants of porokeratosis in the same individual.[10,11]

Keratolytic agents, topical 5-flourouracil, topical and oral retinoid agents, topical imiquimod, cryotherapy, photodynamic therapy, carbon dioxide laser ablation, dermabrasion, and excision are used for treatment,[12] but achieving complete resolution of lesions is difficult. In our case, since the lesions were generalized, we considered oral acitretin 25 mg after the baseline investigations. Cryotherapy was done for hypertrophic lesions, and photoprotection was advised. As the inheritance of the disease is autosomally dominant, the patient was educated of possible future transmission of the disease in his progeny; we lost the patient to further follow-up.

The interesting features of our case report are: (1) Porokeratotic lesions involving almost all parts of the body from head to toe (2) involvement of uncommon sites such as scalp, oral mucosa, palms, soles and genitalia (3) simultaneous occurrence of

various subtypes - CPM, DSAP, hypertrophic porokeratosis (4) association with vitiligo of the glans penis and (5) a positive family history.

To the best of our knowledge, this unusual and varied presentation of porokeratosis in the same individual is being reported for the first time in the literature.

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