

Porokeratosis: a report of three clinical variants

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Summary

Three patients presenting with distinct clinical variants of porokeratosis are reported. This article discusses the etiology, classification and response to different modalities of treatment.

Key words: Porokeratosis, variants, treatment response.

Introduction

Porokeratosis constitute a group of uncommon dermatosis that have a special form of faulty keratinisation which histopathologically manifest as parakeratotic cells placed on top of each other resembling a pile of plates — the cornoid lamella. This clinically appears as an annular, sharply demarcated lesion with a raised hyperkeratotic border.

Case report 1: A 18 year old young Malay woman was admitted for evaluation of an asymptomatic dermatosis that had been present for the last 10 years. Numerous erythematopapular lesions were seen on the outer surface of the right thigh. These lesions were of different shapes and sizes with a reddish-violet central area and hyperpigmented, serpiginous, raised and well-defined margin. They were grouped together with a zosteriform arrangement. On palpation the lesions had an irregular surface and a soft parenchymatous consistency. (Fig. 1).

The patient had no family history of similar lesions. She had been treated elsewhere with various topical corticosteroids without significant benefit. The patient was treated with topical 5-Flurouracil but the response was unsatisfactory and she was advised grafting but refused.

Case report 2: A 42 year old Chinese female presented with numerous small, asymptomatic, superficial, relatively uniform annular keratotic papules over the face (Fig. 2), neck, forearms and feet of one year duration. Individual lesions were dry and anhidrotic and absent from body regions shielded from sunlight. She was initially treated with 5-Fluorouracil topically but without benefit. Subsequently a course of oral etretinate followed by isotretinoin were tried over a period of six months but response was unsatisfactory.



Figure 1. Zosteriform Porokeratosis on the right thigh.

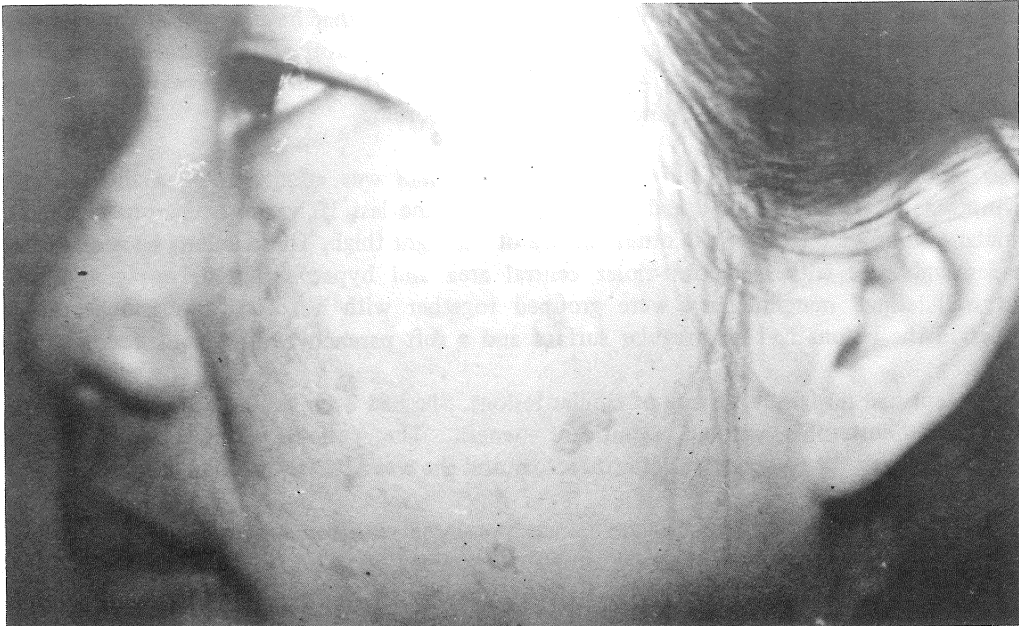


Figure 2. Disseminated superficial actinic porokeratosis.

Case report 3: A 14 year old Malay girl was referred for an asymptomatic annular plaque over the right knee which had been present for six years. The lesion started as a small, brownish keratotic papule that slowly enlarged to form an irregular, annular plaque with a smooth atrophic center and well demarcated, raised, hyperkeratotic border (Fig. 3). She was treated with 5-Fluorouracil but responded poorly and subsequently liquid nitrogen cryotherapy was carried out with minimal improvement.

Results of routine blood tests and investigations were all within normal limits in all three patients. Biopsy specimens were obtained from all three patients. The histopathologic examination gave identical results in each case and showed nucleated horn cells in the stratum corneum (parakeratosis) arranged in a column (cornoid lamella) with hyperkeratosis on either side. Beneath the cornoid lamella the granular layer was deficient and replaced by dyskeratotic and vacuolated cells. In the superficial dermis a scanty perivascular lymphocytic infiltrate was observed (Fig. 4).



Figure 3. Porokeratosis of Mibelli over the right thigh.

Discussion

Porokeratosis is a chronic, progressive disease characterised by the formation of slight atrophic patches surrounded by an elevated, warty border. The etiology of the various types of porokeratosis is unknown. Porokeratosis is a disease of the epidermis in which a mutant clone

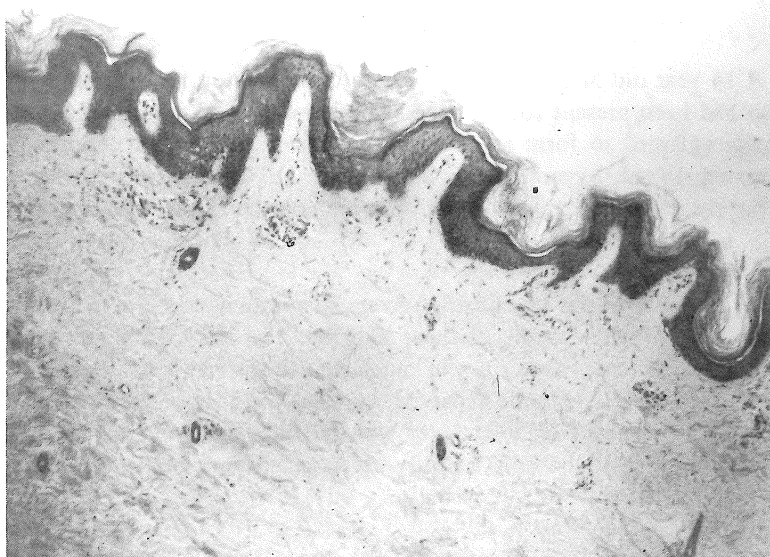


Figure 4. Cornoid lamella arises from a small indentation of the epidermis.

of epidermal cells expand peripherally leading to formation of the cornoid lamella at the boundary between the clonal population and normal epidermal cells.¹ The tendency for the abnormal clones to develop is probably inherited as latent abnormal clones may become clinically overt following solar exposure, indicating that the phenotypic expression of the regulating gene is dependent on uv radiation.² In porokeratosis we are watching the quiet drama of a mutant cell and its progeny pushing aside and replacing the normal epidermis. Each lesion represents a clone of these raw keratinocytes and the keratotic rim, the battle line between the invasive cells and the normal epidermal population. The mutant epidermis is thin and sparsely colonised and this atrophic epidermis after years of sun exposure may account for the degeneration of squamous cell carcinoma in some of these patients.³

A number of clinical variants are being recognised. Generally accepted variants include:

1. Zosteriform porokeratosis (case report 1).
2. Disseminated superficial actinic porokeratosis (case report 2).
3. Porokeratosis of Mibelli (case report 3).
4. Palmarplantar porokeratosis et disseminata.
5. Punctate porokeratosis which is usually associated with the zosteriform or Mibelli variant.

Zosteriform porokeratosis (Fig. 1) represents a rare variety of porokeratosis with only 40 cases being reported worldwide.⁴ The age of onset varies. Although it may present at birth, the condition may appear in childhood or early adulthood. In contrast to other types of porokeratosis, familial cases of zosteriform porokeratosis are rare. Zosteriform keratosis appears as a unilateral, interrupted, linear array of annular hyperkeratotic papules and plaques most commonly on the extremities.

Disseminated superficial actinic porokeratosis (Fig. 2) is the most common form of porokeratosis. It is transmitted by an autosomal dominant gene, expressed most often in women. It is characterised by numerous superficial, annular keratotic, brownish red macules found on sun exposed areas.

Porokeratosis of Mibelli (Fig. 3) is transmitted by an autosomal dominant gene, and the onset of the disease is early in life. The disease is twice as common in males as in females. The disease begins as a small keratotic papule, which spreads peripherally and atrophies in the center. The disease also occurs in the buccal mucosa and on the glans penis.

Treatment of porokeratosis lesion may be initiated for cosmetic improvement or to prevent a potential carcinoma. Carcinoma develops in these lesions not infrequently. James et al⁵ reviewed the literature in 1986 and found 29 patients in whom squamous cell carcinoma (21 patients), Bowen's disease (eight cases), or basical cell carcinoma (three cases) was found within lesions of porokeratosis. Most occurred in the zosteriform type, and most were on the legs.

When lesions of porokeratosis are few they may be excised or destroyed by cryotherapy. Topical 5-Fluorouracil has been found to be effective in porokeratosis of Mibelli, disseminated superficial actinic porokeratosis and zosteriform porokeratosis.^{2,6-8} However our patients did not respond satisfactorily although they were treated for a sufficient duration, and this has been confirmed by others.^{3,9}

The use of oral retinoids has yielded conflicting results. A good response has been observed in some patients with disseminated superficial actinic porokeratosis,¹⁰ whereas failures (as in our patient) have also been confirmed by other reports.¹¹ Treatment of porokeratosis of Mibelli and zosteriform porokeratosis is difficult and excision is the best approach if limited to small areas. In widespread cases patient education, periodic examination and protection from ultraviolet rays is in our opinion the best approach to this interesting disorder.

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