Disseminated linear porokeratosis: a case report and literature review

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Porokeratosis is a heterogeneous group of keratinization disorders with an autosomal dominant inheritance pattern. The etiology and pathogenesis of this disease have not been completely elucidated. The porokeratosis clinically presents with a characteristic prominent peripheral ridge and central dell. The histological feature of the parakeratotic column known as 'cornoid lamellae' is a hallmark feature observed in the peripheral rim. Among the various morphological forms, linear porokeratosis is a rare variant with four subtypes. When compared to other forms, the linear variant carries an increased risk of malignancy, particularly squamous cell carcinoma, warranting a regular follow-up. Though there are multiple treatment options, achieving complete clinical resolution is difficult. We report an interesting case of linear porokeratosis with generalized lesions in a 60-year-old male. The lesions were typical and predominantly distributed in the extremities. The biopsy was diagnostic in our patient. This case is reported for its rare presentation and clinical interest.

Keywords: linear porokeratosis, cornoid lamellae, malignancy.

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INTRODUCTION

Porokeratosis is a rare abnormal epidermal keratinization disorder, characterized clinically by a hyperkeratotic plaque with an elevated border that expands centrifugally and histologically by the presence of cornoid lamella [1]. Porokeratosisis is a heterogeneous group of disorders inherited in an autosomal dominant fashion [2]. Sporadic cases are not uncommon [3]. We present a rare case of linear porokeratosis occurring in multiple sites.

CASE PRESENTATION

A 60-year-old male presented with dark-colored raised lesions over the right thigh region since 5 years of age (Figure 1). At 30 years of age, he developed similar lesions over the lower abdomen and lateral aspect and nape of the neck (Figure 2). Initially, the lesions were present as small hyperpigmented papules, which developed into large itchy hyperkeratotic plaques. There was no family history of a similar disease. There were no hair or nail changes.

On cutaneous examination, there was a well-defined irregular plaque with central hyperpigmentation and mild atrophy with a slightly raised keratotic periphery. The plaque extended from the medial aspect of the knee to the anterior aspect of the right upper thigh in a linear pattern. Similar plaques were present on the right side of the lower abdomen and left lateral aspect of the neck following Blaschko's lines. He also had a diffuse hyperpigmented plaque over the nape of the neck region. The histopathological examination from the periphery of the lesion revealed epidermal invagination with parakeratotic columns of cells (cornoid lamella). The granular layer was absent below the invagination, and the upper dermis showed a lichenoid infiltrate (Figure 3). Hence, a diagnosis of generalized linear porokeratosis was made. The patient refused to undergo any

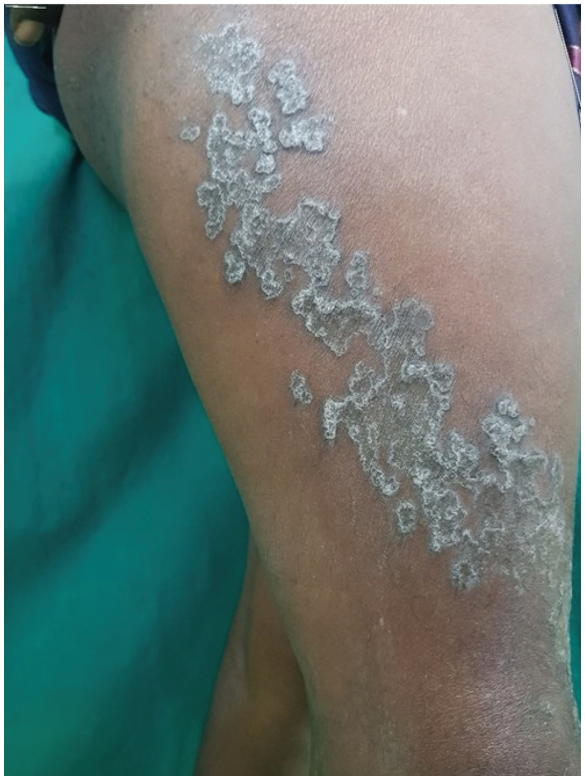

Figure 1. Linear hyperkeratotic plaque over the right thigh region.

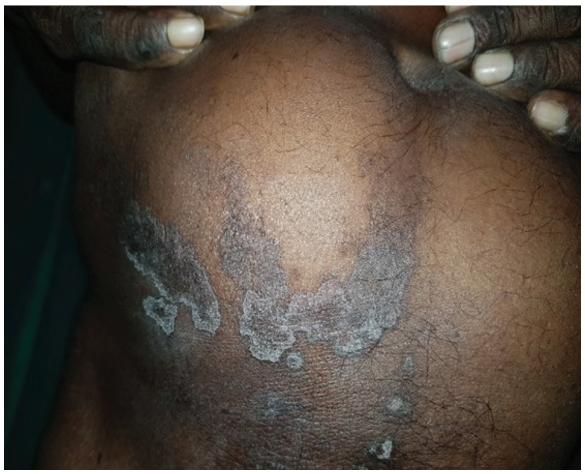

Figure 2. A well–defined plaque with ridging over the lower abdomen.

modality of treatment.

DISCUSSION

Porokeratosis is a disorder with varied morphological presentations. It includes clinical variants such as classical porokeratosis of Mibelli, linear porokeratosis, disseminated superficial porokeratosis, disseminated actinic superficial porokeratosis, punctate porokeratosis of the palms and soles, porokeratosis palmaris, and plantaris et disseminate [4]. Other rare reported variants include facial, giant, follicular, hypertrophic/verrucous, bullous, ptychotropica, hypopigmented, pustular, pruritic papular, hypopigmented, and erosive porokeratosis [5].

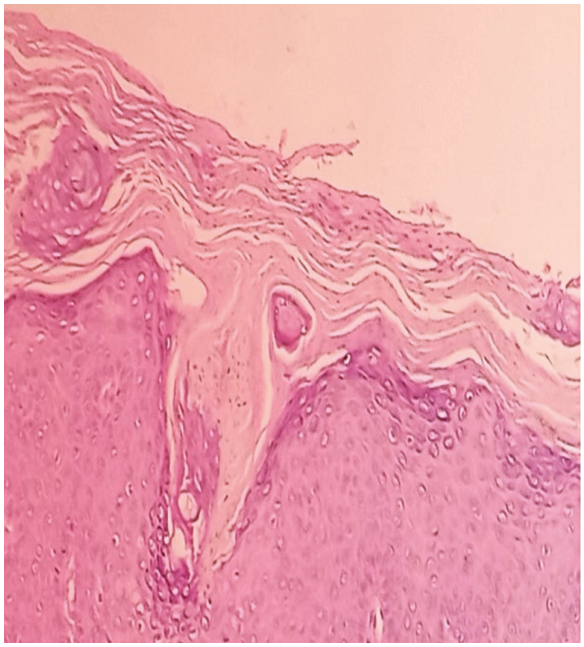

Figure 3. Hyperkeratosis, parakeratotic column, and a keratin-filled invagination (H & E 10×).

Among the clinical forms, linear porokeratosis is a rare and unique variant 1. It is further classified into the localized, zosteriform, generalized, and systematized subtypes [4]. The localized form is most common, where lesions are unilateral and confined to a distal extremity following Blaschko's lines. The generalized form is rare, and lesions are multiple affecting extremities as well as the truncal region [1].

The allelic loss and overexpression of p53 predispose to various cutaneous malignancies in linear porokeratosis [2]. Linear porokeratosis carries a 19% risk of malignancy, whereas, in other types, the risk is between 3–10%. The most common associated malignancy is squamous cell carcinoma, followed by Bowen's disease and basal cell carcinoma [5].

The conditions to be ruled out for diagnosing

linear porokeratosis are linear lichen planus, lichen striatus, linear verrucous epidermal nevus, linear psoriasis, incontinentia pigmenti, and pityriasis rubra pilaris [1,4]. Of these, linear lichen planus usually presents with extensive lesions and have a malignant potential similar to linear porokeratosis [6].

Localized porokeratosis can be treated with topical 5-fluorouracil, keratolytics, retinoids, calcipotriol, and imiquimod cream [2,7]. Ablative therapeutic options include ablative laser, chemical peeling, cryotherapy, and photodynamic therapy [5,7]. However, they may cause scarring as the ablation reaches the mid-dermis. In diffuse cases, topical management does not give desired results and causes relapse. Surgery is not preferred as it may lead to scarring [7]. In our case, the disease was extensive and bilateral, ruling out topical and surgical management. Systemic retinoids are helpful in such scenarios and may be used as prophylaxis against malignancy [7].

Similar cases are summarized in Table 1 [2,4,7-10].

Regular follow-up is required for these patients, even if the lesion does not show features of malignancy.

CONCLUSION

This case is reported for the rarity of generalized linear porokeratosis. Usually, linear porokeratosis is unilateral, but our patient had an unusual bilateral presentation. A regular follow-up on a strict basis is required as there is an increased risk of malignancy. The possibility of malignancy should be kept in mind even if the initial pathology is benign.

Conflict of interest: None declared.

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Table 1. A summary of the previous cases of disseminated porokeratosis [2,8,4,7,9,10]

No	Age	Sex	Age at onset of the lesion	Distribution	Associated features
1	23 years	Female	2.5 years	Right gluteal region, posterior aspect of right leg, right side of the trunk, both arms, left leg, dorsum of the hands and feet, and both soles	Nail dystrophy with longitudinal ridges, pterygium, and subungual keratosis
2	36 years	Male	At birth	All the extremities and anterior part of the trunk	Hyperkeratotic fissured plaque present over the dorsal aspect of both feet
3	24 years	Female	6 months	Face, neck, right arm, lower right leg, palms, and soles	Nail changes; dorsal pterygium
4	54 years	Female	Since childhood	Left side of the body over face, neck, arm, forearm, hand, gluteal region, and thigh.	-
5	57 years	Male	12 years	Right side of the body over the arm, posterior back, lateral leg, and gluteal region.	-
6	25 years	Female	Since birth	Left sole, left leg, left thigh, and left gluteal region.	-