Case Report

A rare case of disseminated superficial porokeratosis-Case report

Ramachandran Ramakrishnan, T. Arun Vignesh, Priya Cinna T. Durai, Murali Narasimhan

Department of Dermatology, Venereology and Leprosy, SRM Medical College Hospital and Research Centre, Potheri, Chennai, Tamil Nadu, India

Abstract

Porokeratosis is a keratinization disorder characterized by hyperkeratotic sharply demarcated plaques with central atrophy and histopathologically, by cornoid lamella. A 30-year-old male presented with multiple pruritic dark raised skin lesions over the trunk, face, and upper limbs for past 3 years. Cutaneous examination revealed hyperkeratotic annular plaques with raised margins over face, trunk, and arms. Histopathology revealed marked hyperkeratosis with irregular acanthosis and papillomatosis. Vertical parakeratotic foci and focal hypergranulosis were seen. Hence, a diagnosis of disseminated superficial porokeratosis was made. We present this rare case which may have association with systemic disease, immunosuppression, and malignant transformation.

Keywords: Cornoid lamella, disseminated superficial porokeratosis, porokeratosis

Introduction

Porokeratosis is a disorder characterized by clonal expansion of keratinocytes, which differentiate abnormally but are not truly neoplastic. Mibelli described the classical form which bears his name, in 1893. This was followed by descriptions of superficial and disseminated forms of porokeratosis, linear porokeratosis, disseminated superficial actinic porokeratosis (DSAP), disseminated palmoplantar porokeratosis, and punctate porokeratosis by others subsequently. All forms of porokeratosis show thin vertical tier of parakeratosis, the cornoid lamella, representing the elevated border. The progression of porokeratosis to squamous cell carcinoma is noted in about 6.9--30% of cases.[1] Primary care physicians need to be familiar with the typical cutaneous manifestation of such rare dermatoses with typical and easily identifiable skin lesions. This can help in early referral to the dermatologist and thereby reduce the risk of complications.

Address for correspondence: Dr. Ramachandran Ramakrishnan, Department of Dermatology, Venereology and Leprosy, SRM Medical College Hospital and Research Centre, Potheri, Chennai - 603 203, Tamil Nadu, India.
E-mail: drram.srkn@gmail.com

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Case Report

A 30-year-old male patient presented to dermatology OPD with multiple dark raised skin lesions over the trunk, face, and upper limbs for the past 3 years. He started developing occasionally itchy, dark raised skin lesions, initially over the face, followed by chest, back, and upper limbs. The lesions gradually increased in size and number. There was no photosensitivity and no similar skin lesions among other family members.

Cutaneous examination revealed both hyperkeratotic plaques and typical annular plaques with raised margins over both exposed and non-exposed parts of the body [Figure 1a and b].

Skin biopsy revealed marked hyperkeratosis, irregular acanthosis, and papillomatosis. Multiple foci of vertical layered parakeratosis

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and few foci of hypergranulosis were seen. Base of the parakeratotic columns contained vacuolated keratinocytes with few apoptotic keratinocytes and dyskeratotic cells. Lymphohistiocytic infiltrates and melanophages were noted in the upper dermis [Figure 2]. With the clinico-pathological findings, patient was given a diagnosis of DSP. After all relevant pre-treatment investigations, patient was started on oral Isotretinoin, topical keratolytics, and oral antihistamine medication.

Discussion

Porokeratosis is a clonal expansion of keratinocytes which are not hyperproliferative, but differentiate abnormally. Porokeratoses may present as single or multiple lesions and can be of the disseminated or localized form. The raised border, represented histopathologically by the cornoid lamella, is composed of a thin vertical tier of parakeratoses.[2,3] Dermoscopic features include central brown pigmentation with blue-gray dots, encircled by a single hypopigmented band and peripheral white track.[4]

The different types of porokeratosis commonly manifest in specific age groups. Classical porokeratosis of Mibelli and linear porokeratosis appear during infancy or childhood. Punctate palmoplantar porokeratosis and disseminated palmoplantar porokeratosis usually appear in adolescence, whereas DSAP generally first manifests in adult life. Porokeratosis of Mibelli, genital porokeratosis, and punctate porokeratosis are more common in males, whereas DSAP is more common in women.

Drug-induced immunosuppression and exposure to UV radiation are triggering factors for DSAP and porokeratosis of Mibelli.

Porokeratosis usually presents as single or multiple papules or plaques which develop into annular lesions with a thin raised border.

They are divided into localized and disseminated forms [Table 1].

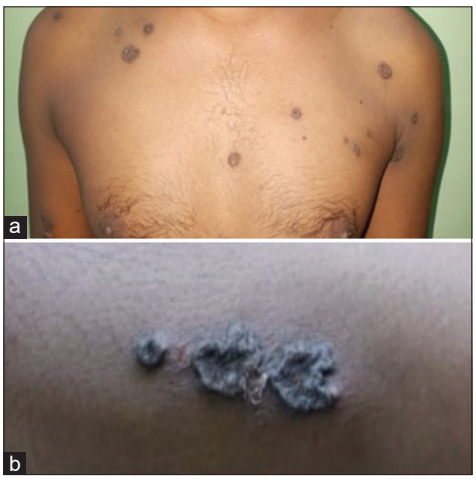

Figure 1: (a) Multiple hyperpigmented annular plaques over the chest, shoulders and upper arms. (b) Closer view of annular plaques with raised borders in the upper arm

Localized forms

Porokeratosis of Mibelli starts as single or multiple keratotic papules. Lesions are most commonly seen over the extremities.

Linear porokeratosis presents during infancy, as unilateral plaques or streaks of reddish-brown papules along limbs, sides of the trunk, head or neck in a Blaschkoid distribution, indicating underlying somatic mosaicism. There is a high risk of malignant transformation.[5] Malignant transformation to squamous cell carcinoma has been noted in about 6.9% to 30% of these cases.[1] With regular use of sunscreens risk of malignant transformation can be reduced.

Punctate palmoplantar porokeratosis is the rarest type presenting with seed like lesions over palms and soles.[3] Genital and perianal porokeratosis are other rare forms.

Disseminated forms

DSAP presents as single or multiple skin colored, pink or red macules with fine scales and a characteristic well-defined raised border. It favors lower legs and arms.

Disseminated superficial porokeratosis (DSP) can present with skin lesions in both sun-protected and sun-exposed sites, occasionally including the oral mucosa and genitalia. It is associated with immunodeficiency and may also develop sporadically during childhood.

Table 1: Clinical classification of porokeratoses[2]

Localized forms	Disseminated forms
Porokeratosis of Mibelli	Disseminated superficial actinic porokeratosis
Perianal porokeratosis	Disseminated palmoplantar porokeratosis
Genital porokeratosis	Systematized linear porokeratosis
Linear porokeratosis	Disseminated superficial porokeratosis
Punctate palmoplantar porokeratosis	

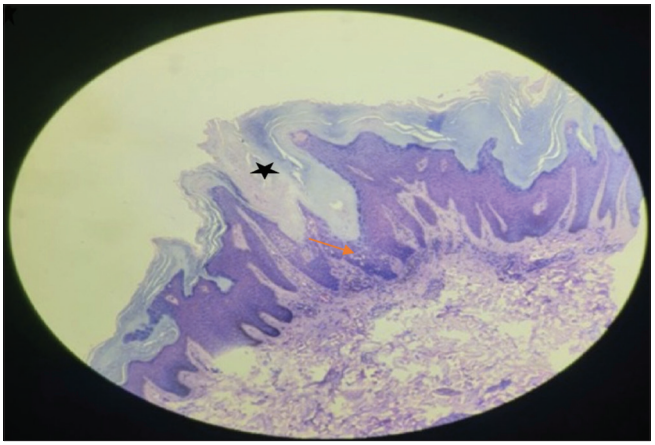

Figure 2: Histopathology of the trunk lesion showing vertical layered parakeratosis, the cornoid lamella (black star) and vacuolated keratinocytes below the cornoid lamella (red arrow) {H and E stain- 10×}

Systematized linear porokeratosis is a disseminated variant of linear porokeratosis, which may be unilateral or generalized, and follows the lines of Blaschko.

Disseminated palmoplantar porokeratosis is a rare generalized form of punctate palmoplantar porokeratosis.

The main differential diagnoses for disseminated Superficial porokeratosis include psoriasis, tinea corporis, granuloma annulare, generalized lichen planus, and annular syphilides.

Causes of immunosuppression, including HIV and hematological malignancies, may have to be ruled out, in cases of sudden onset of porokeratosis. All clinical types of porokeratosis have a potential for malignant transformation especially squamous cell carcinoma.[6,7]

Treatment options for porokeratosis, include topical, systemic, and surgical modalities. Topical 5 -fluorouracil, imiquimod cream, diclofenac gel, cryotherapy and oral retinoids are used to treat porokeratosis of Mibelli.[8] Linear porokeratosis shows excellent response with topical or systemic retinoid therapy.[8] Q-switched ruby, Nd:YAG, CO2, Grenz ray and fractional photothermolysis lasers, have been used to treat DSAP.[7] Oral retinoids such as isotretinoin and acitretin have been advocated for treatment of immunosuppressed and/or those with the linear or disseminated forms of the disease, in order to reduce the risk of malignancy. DSP has a good prognosis and shows excellent response to treatment. Tendency to remit spontaneously is rare and regular follow-up is needed. Topical therapy with diclofenac, steroids, and retinoids may provide symptomatic relief.[9] Topical diclofenac has been used to treat DSAP with a good safety profile.[10] Surgical interventions or cryotherapy may be used in scenarios where prolonged use of topicals is difficult or contraindicated.[8]

There have been reports of association of DSP with systemic diseases like diabetes and liver disease, drug intake and immunosuppression. Our case is an example of occurrence of DSP without evidence of any immunosuppression or underlying systemic disease.

If the Primary care physicians are aware of the typical skin manifestations of this rare dermatoses, it can lead to provisional clinical diagnosis of this condition at the primary care level. Prescribing regular use of sunscreens and subsequent prompt referral for specialist management can prevent complications due to longstanding disease, such as malignant transformation.

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Conflicts of interest

There are no conflicts of interest.

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