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LINEAR POROKERATOSIS ALONG THE BLASCHKO'S LINES- A CASE REPORT

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ABSTRACT
Porokeratosis is a rare chronic progressive disorder which usually presents as an asymptomatic, hyperkeratotic papules and plaques surrounded by an elevated and raised margins that expands centrifugally. We report a case of linear porokeratosis in a 19 years old male who presented with asymptomatic pigmented scaly plaques over his right arm for the past 6 years.

INTRODUCTION

Porokeratosis is a rare and chronic progressive disorder of keratinization usually presenting as an asymptomatic, hyperkeratotic papules and plaques with raised margins that extends centrifugally over years. There are five types of porokeratosis which includes Plaque type of Mibelli, Disseminated superficial actinic porokeratosis, Porokeratosis plantaris, palmaris et disseminate, Linear porokeratosis and Punctate porokeratosis of which Linear porokeratosis is not a common variant and usually it presents in early childhood and rarely since birth and may represent a segmental form of actinic disseminated porokeratosis They are usually asymptomatic and have classical histopathology of cornoid lamella.

Case report:

A 19 year old male presented to our OPD with complaints of dark coloured skin lesions over the right arm of 6 years duration. Initially a single, small lesion was present over the lower part of the right arm and subsequently progressively increased to the present size. There was no history of itching, no history of similar lesions else were on the body. There was no history of similar complaints in the family. Dermatological examination revealed well-defined multiple, pigmented, Scaly plaques with raised borders in a linear distribution over the extensor aspect of the right arm [fig 1]. Palms, soles, nails, oral mucosa and the scalp were normal. General and systemic examinations were normal. Routine investigations done were within normal limits. Histopathology from the elevated margins revealed hyperkeratotic invagination of the stratum corneum within the epidermis, with parakeratosis confined within it. Granular layer was missing and a very few lymphocytes were seen in the dermis. Thus our diagnosis was confirmed based on clinical and the histopathological findings.

DISCUSSION

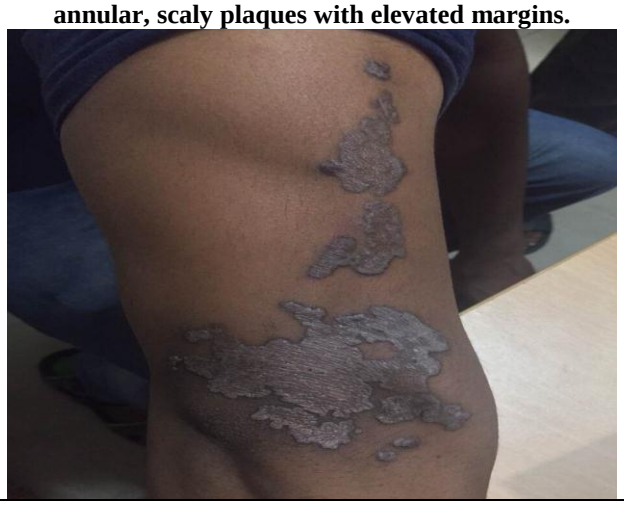
Porokeratosis is a rare and progressive keratinization abnormality. Linear porokeratosis is not a common variant that usually presents in early childhood and rarely since birth and may represent a segmental form of actinic disseminated porokeratosis [1]. Incidence wise females outnumber the males. Clinically lesions may be hyper or hypo pigmented and/or erythematous plaques that

are disposed in an interrupted linear fashion confined to an extremity following one Blaschko's line. Rarely generalized forms with lesions occurring on the trunk face and soles are seen. Unilateral and linear form resembles linear verrucous epidermal nevus [2]. Lesions affecting the distal limb can be associated with nail dystrophy such as pterygium and longitudinal ridging. Linear porokeratosis carries the highest risk for developing malignant degeneration among the other types of porokeratosis and especially develops squamous cell carcinomas [3].

The cause has been postulated as to occur sporadically and in some as autosomal dominant. The centrifugal expansion of individual lesions is reflected due to the migration of a mutant clone of keratinocytes. They have the highest potential for malignant degeneration of all the types of porokeratosis. Histologically, the peripheral raised hyperkeratotic ridge corresponds to the cornoid lamella [4] which shows invagination of the epidermis, filled with keratin, centre of which demonstrates a column of parakeratotic cells. The keratinocytes in the epidermis below the parakeratotic column shows missed out granular layer, pyknosis and peri-nuclear hallowing. Occasionally dermis may show non-specific infiltrate comprising of chronic inflammatory cells, perivascularly.

Treatment comprises cryotherapy, usage of carbon dioxide and or pulsed dye laser, photodynamic therapy and topical keratolytics, topical 5-Fluorouracil and imiquimod [5]. The occurrence of this entity with classical histologic findings is rare to see, henceforth this case has been reported. Following up the patient for long durations is important as to look for early development of malignant changes.

Figure 1. Clinical picture of linear porokeratosis along the Blaschko's line. There are multiple, pigmented, annular, scaly plaques with elevated margins.



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CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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