CASE REPORT

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Disseminated Superficial Porokeratosis Mimicking Disseminated Discoid Lupus Erythematosus: An Unusual Presentation

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ABSTRACT

Porokeratosis is an acquired disorder of keratinization characterized by clonal expansion of keratinocytes which differentiate abnormally. All forms of porokeratosis have been reported to have familial clusters with autosomal dominant patterns of inheritance but with variable penetration. It is classified into the localized forms which include porokeratosis of Mibelli, linear porokeratosis (LP), and punctate palmoplantar porokeratosis, genital porokeratosis and perianal porokeratosis; the disseminated forms including disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis (DSP), and disseminated palmoplantar porokeratosis and systematized LP. Disseminated superficial porokeratosis presents with multiple pink or red-brown finely scaly macules with a well defined raised border which appears in early adult life predominantly on extremities. The histopathology is characterized by thin column of tightly packed parakeratotic keratinocytes within a keratin filled invagination of the epidermis through stratum corneum known as cornoid lamella. It is associated with immunodeficiency or may appear spontaneously in childhood. Here we describe a young man with hyperkeratotic, hyperpigmented annular plaques distributed over extremities, trunk and face, mimicking disseminated discoid lupus erythematosus.

Keywords: Porokeratosis, Disseminated, Disorder of keratinization

Introduction

Porokeratosis is a clonal disorder of keratinization characterized by lesions with an atrophic center, prominent peripheral ridge, and a histologic hallmark in the form of cornoid lamella. Genetics, immunosuppression, and sunlight are some of the factors blamed for its occurrence. Various morphological variants have been described. Here we report a case who presented with disseminated superficial porokeratosis (DSP) resembling discoid lupus erythematosus (DLE).

Case Report

A 23-year-old man presented with brown slightly raised skin lesions over both upper extremities for last 5 years. The lesions were small to start with and then gradually increased in size with central flattening. It first appeared on face and neck then progressed to involve trunk and upper extremities. There was no history of photosensitivity, joint pain, recurrent fever or Raynaud's phenomenon. Family history was positive. Annular hyperpigmented plaque with hyperkeratotic raised margins measuring 0.5x1 cm to 1.5x2.5 cm with central atrophy and scaling "Figure 1", "Figure 2". A biopsy from margin of annular plaque, stained with hematoxylin and eosin stain, showed cornoid lamella, a parakeratotic column of keratinocytes within a keratin-filled invagination of epidermis through the stratum corneum with absent underlying stratum granulosum, perivascular lymphocytic infiltrate "Figure 3". A Periodic acid-Schiff (PAS) stain was also performed which showed no thickening of basement membrane "Figure 4".

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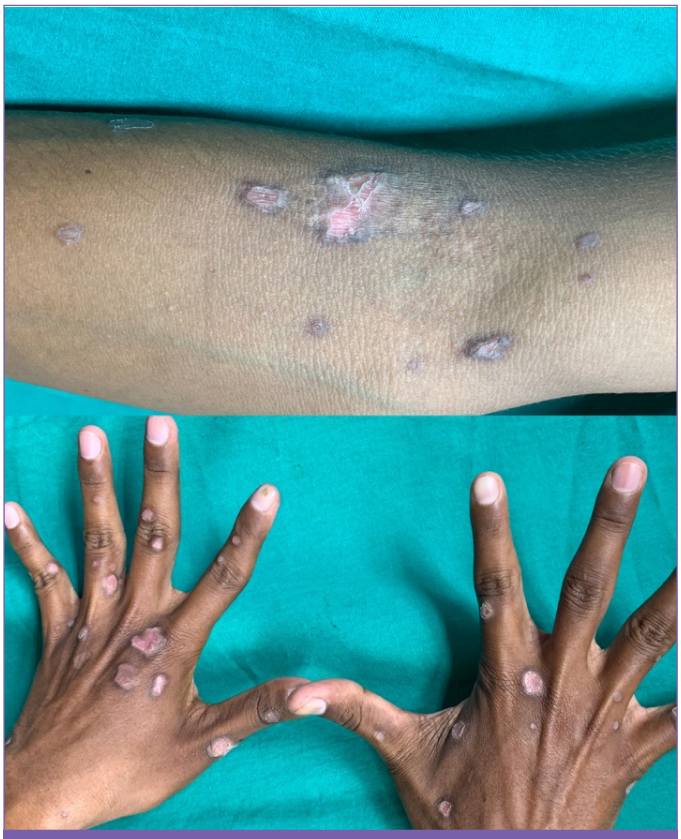

Figure 1. Annular hyperpigmented plaque with hyperkeratotic raised margins on extremities

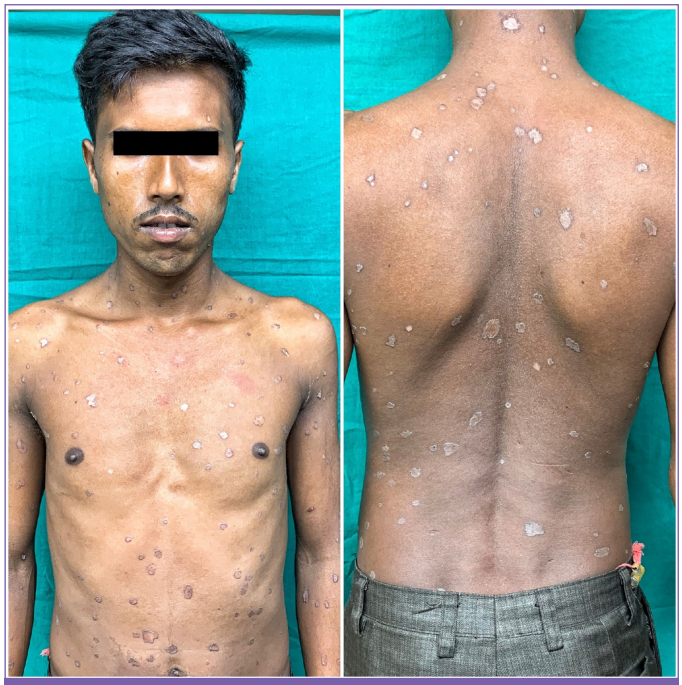

Figure 2. Annular hyperpigmented plaque with hyperkeratotic raised margins on anterior and posterior trunk

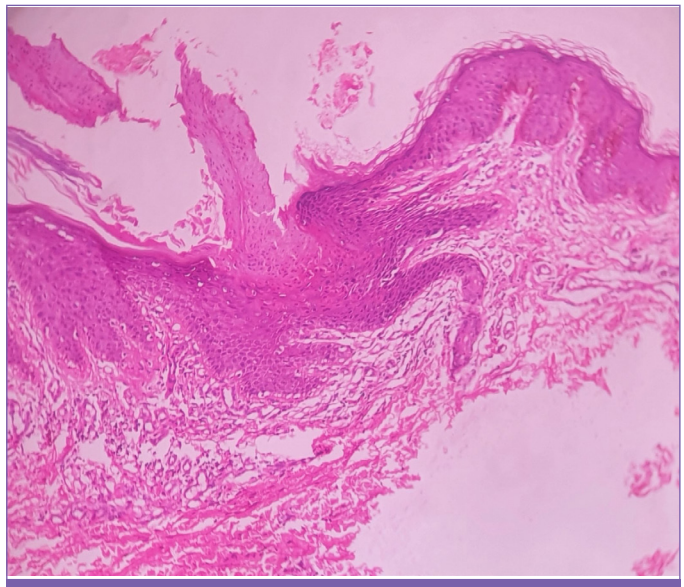

Figure 3. Biopsy from margin of annular plaque, stained with Hematoxylin and Eosin stain, showed cornoid lamella, a parakeratotic column of keratinocytes within a keratin&8208; filled invagination of epidermis with absent underlying stratum granulosum

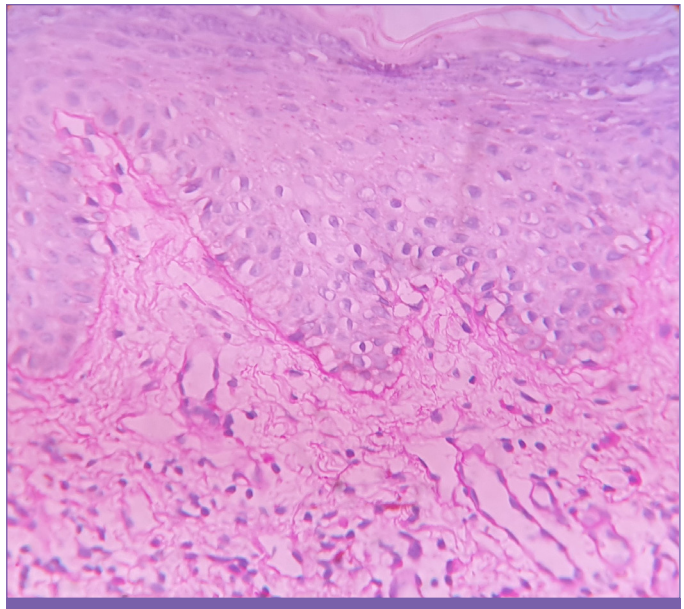

Figure 4. Periodic acid–Schiff stain was also performed which showed no thickening of basement membrane

Discussion

Porokeratosis is an acquired disorder of keratinization characterized by clonal expansion of keratinocytes which differentiate abnormally. One of its variants is DSP which presents with multiple pink or red-brown finely scaly macules with a well-defined raised border which appears in early adult life predominantly on extremities. The histopathology is

characterized by thin column of tightly packed parakeratotic keratinocytes within a keratin filled invagination of the epidermis through stratum corneum known as cornoid lamella [1]. All forms of porokeratosis have been reported to have familial clusters with autosomal dominant patterns of inheritance but with variable penetration [2]. It has been classified into the localized forms which include porokeratosis of Mibelli, linear porokeratosis (LP), punctate palmoplantar porokeratosis, genital porokeratosis, perianal porokeratosis and the disseminated forms including disseminated superficial actinic porokeratosis, DSP, and disseminated palmoplantar porokeratosis and systematized LP [3]. DSP is not necessarily related to sun exposure and will then present in both sun-exposed and sun-protected sites, including sometimes oral mucosa and genitalia. It may be associated with immunodeficiency (e.g. organ transplantation, malignancy, HIV infection) or may develop sporadically during childhood. Cutaneous malignancies particularly squamous cell carcinoma may occur as a complication of porokeratosis. All forms of porokeratosis are chronic with no tendency for spontaneous resolution. It can mimic DLE [4]. Its association with Gardner syndrome, Lichen planus, diabetes mellitus, CAP syndrome, Bloom syndrome and cystic fibrosis has also been reported. Various modalities of treatment include topical retinoids, cryotherapy, 5-fluorouracil, imiquimod, curettage and cautery, photodynamic therapy, CO_2 laser and topical vitamin D analogues such as calcipotriol has been used as first line therapy with varying degree of success. Oral retinoids such as isotretinoin and acitretin have been given to patients with porokeratosis who are immunosuppressed to reduce the risk for malignant transformation. Patient must be counselled regarding photoprotection and long term follow up. Our patient responded well to treatment.

Ethics

Informed Consent: Consent form was filled out by a participant.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.H., S.M., S.B., S.Ban., G.C., Concept: A.H., S.M., S.Ban., G.C., Design: A.H., S.M., S.B., S.Ban., G.C., Data Collection or Processing: A.H., S.M., S.B., S.Ban., G.C., Analysis or Interpretation: A.H., S.M., S.B., S.Ban., G.C., Literature Search: A.H., S.M., Writing: A.H., S.M.

Conflict of Interest: No conflict of interest was declared by the authors.

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