

Disseminated superficial actinic porokeratosis: A case report

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Abstract Disseminated superficial actinic porokeratosis (DSAP) is characterized by small, atrophic patches with distinctive keratin rims that occur on sun-exposed areas of the extremities, shoulders, and back. The diagnosis is based on the histopathologic finding of a cornoid lamella, absence of a granular layer, and often a thin epidermis. It is associated with exposure to ultraviolet radiation. We report a case of DSAP in our setting.

Key words

Porokeratosis, disseminated superficial actinic porokeratosis.

Introduction

Porokeratosis is a disorder of keratinization characterized by one or more atrophic macules or patches surrounded by a distinctive hyperkeratotic ridge-like border called a cornoid lamella. Multiple clinical variants of porokeratosis exist e.g. classic porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis, linear porokeratosis, porokeratosis plantaris palmaris et disseminate, punctate porokeratosis and giant porokeratosis. Malignant transformation occurs in a minority of cases.¹

Although clinical surveillance for malignant transformation is sufficient for the management of most patients with porokeratosis, patients who are concerned about the appearance of lesions or who have associated symptoms such as pruritus or pain may desire therapeutic intervention.

Various topical, excisional, destructive, and systemic therapies appear to be effective.

We herein present a case of disseminated superficial actinic porokeratosis along with review of literature.

Case Report

A 43-year-old woman, housewife, presented with 6-year history of bilateral, symmetrical, erythematous, papules gradually enlarging to form plaques on the dorsa of both hands. After 2 months, similar lesions appeared on the forearms, legs, trunk and forehead. These lesions enlarged to form plaques. The eruption was associated with mild pruritus and history of photosensitivity.

On examination, there were erythematous papules and plaques with well-demarcated, hyperpigmented, scaly border and central atrophy (**Figure 1**). Erythematous scaly papules were present on the dorsa of both hands with central atrophy and a well-demarcated, hyperpigmented and scaly border. There were no extracutaneous manifestations.

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Figure 1 An erythematous plaque with well-demarcated, hyperpigmented, scaly border and central atrophy.

A punch biopsy was taken and sent for histopathology. On histopathological examination of the specimen, cornoid lamella was found in the stratum corneum, the underlying epidermis showed vacuolar degeneration with a prominent granular layer at the edges. The case was diagnosed as disseminated superficial actinic porokeratosis. The patient was investigated for fasting lipid profile and liver function tests. Oral acitretin 20mg once a day, topical calcipotriol ointment twice a day while a sunblock SPF 60 was prescribed in the morning and afternoon. The patient was advised to have regular follow ups.

Discussion

Disseminated superficial actinic porokeratosis (DSAP) is the most common form of porokeratosis, and may account for almost half of all cases. Patients develop a few to several dozen tan, annular macules with raised peripheral ridges, developing predominantly on the distal extensor surfaces of the legs and the arms. Palms and soles are spared, and facial lesions may be seen in less than 15% of patients. Hyperkeratotic variants have been described. The lesions are usually asymptomatic, but they

may itch or sting slightly. Extensive exposure to natural or artificial ultraviolet radiation may trigger or worsen DSAP. The cornoid lamellae may be stained and accentuated by sunless tanning lotions containing dihydroxyacetone.

Patients are typically women in their third or fourth decade of life, with a history of ultraviolet light exposure. Patients may have a history of phototherapy for psoriasis. There is frequently a family history of DSAP, especially in other females in the family. Lesions of disseminated superficial porokeratosis (DSP), non-actinic, appear very similar except in a generalized distribution. Patients with DSP may be more likely to be immunosuppressed and to be less likely to have worsening with sun exposure than patients with DSAP.

The parakeratosis appears to be the result of faulty maturation of keratinocytes, rather than an increased rate of proliferation.⁵ Several risk factors for the development of porokeratosis have been identified; these factors include genetic inheritance, ultraviolet radiation, and immunosuppression. Sun exposure and/or artificial ultraviolet radiation exposure in a patient who is genetically predisposed cause DSAP. The formation of squamous or basal cell carcinomas has been reported in all forms of porokeratosis.⁶ Several medications have potential benefit like topical 5-fluorouracil, topical vitamin D analogues, topical immunomodulators like 5% imiquimod, systemic retinoids, photodynamic therapy, cryotherapy, electrodesiccation and curettage, CO2 laser and pulsed dye laser and surgical excision for lesions showing malignant changes.

The disease is common between 2nd to 4th decades, transmitted in an autosomal dominant fashion, and is more frequent in women.⁵ Though, porokeratosis is a known autosomal dominant genodermatosis, sporadic cases also

occur but sporadic cases without any family history have been reported in the literature.⁶ The skin lesions of disseminated superficial actinic porokeratosis are most pronounced on sun exposed areas and may aggravate after sun exposure.⁷ Histopathologically it is characterized by the presence of cornoid lamella.⁸

Clonal proliferation of atypical keratinocytes showing abnormal terminal keratinocyte differentiation leads to the formation of the cornoid lamella. Inherited or sporadic genetic defects, possibly creating a change in immune function and/or keratinocyte function, are thought to be responsible for several forms of porokeratosis. Familial cases of all forms of porokeratosis have been reported and appear to have an autosomal dominant inheritance pattern with incomplete penetrance.⁹ Genetic mutations in the *SART3* and *MVK* genes have been found in DSAP pedigrees. Diseases reported in association with porokeratosis include HIV infection,¹⁰ diabetes mellitus, liver disease,¹¹ and hematologic or solid organ malignancy. Immunosuppression may induce new lesions or cause preexisting lesions to flare.

Our female patient was in fourth decade of life. She had history of photosensitivity and classical skin lesions with hyperkeratotic ridge with central atrophy mainly distributed over sun exposed parts of the body. No other family members had similar skin lesions. Our patient had slightly dark complexion. Histopathological examination showed hallmark of porokeratosis the cornoid lamella.

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