Case Report

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Disseminated superficial actinic porokeratosis

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ABSTRACT

Disseminated Superficial Actinic Porokeratosis (DSAP) is a type of porokeratosis observed in parts of the skin exposed to intense sunlight and usually appears in the third or fourth decade of life. We are presenting a case report of a 30 year old male patient presenting with multiple lesions over the face and limbs.

Keywords: Porokeratosis, Skin, Multiple lesions, Face and limbs

INTRODUCTION

Porokeratosis is a rare keratinization disorder with a clinical appearance of papules surrounded by a welldemarcated peripheral keratotic wall, histologically corresponding to a 'cornoid lamella'. 1-3 Five clinical variants have been defined: Classical Porokeratosis (Mibelli), Porokeratosis Palmaris et Plantaris Disseminata (PPPD), Linear porokeratosis, punctate porokeratosis and Disseminated Superficial Actinic Porokeratosis (DSAP). The most common type is DSAP and it consists of brown to red superficial, dry, and anhydrotic annular lesions surrounded by a thin and slightly elevated keratotic wall.² Involvement of the face, mucosae, palms and soles is infrequent.⁴ Lesions generally develop in areas exposed to the sun or become more prominent in the summer months.² Most lesions are asymptomatic. Ulcerative lesions have been described. Giant porokeratosis in a facial or acral location may cause destruction of the underlying soft tissue or pseudoainhum with amputation.⁵ The formation of squamous cell carcinomas or, less commonly, basal cell carcinomas has been reported in all forms of porokeratosis.⁶⁻⁸ Lesions that are large, longstanding, or linear have the greatest risk of developing an associated malignancy.9 Chromosomal instability and reduced immune surveillance with overexpression of p53

are hypothesized to play a role in the development of porokeratosis. 10-14 cutaneous malignancies within Generally, no laboratory studies are required. Screening diseases causing immunosuppression hematologic malignancies) and/or renal failure is appropriate when new lesions of classic Porokeratosis of Mibelli (PM) or Disseminated Superficial Actinic Porokeratosis (DSAP) are seen or when sudden exacerbation of any form of porokeratosis develops. The cornoid lamella is the histopathologic hallmark of all forms of porokeratosis. It is essential that a biopsy specimen be taken from the peripheral, raised. hyperkeratotic ridge to demonstrate this finding.

CASE REPORT

A 30 year old male presented to our department with multiple atrophic macules on face, extensor aspect of both forearms and lesions on the dorsa of both feet since last 2 and a half years. The lesion first appeared behind the left ear and gradually progressed within a period of 6 months to involve the face (Figure 1 & 2) and later, the feet. The lesions became darker in colour over a period of time. The patient complained of itching over the lesions on exposure to sunlight. There were no other aggravating or relieving factors associated. On cutaneous

examination, multiple annular hyperpigmented macules were present over the sun exposed areas of the face, arms and dorsum of the foot. The macules were found to have multiple gutter-like furrows with elevated and hyperpigmented margins. The size of the lesions varied from 0.5 x 1 cm to 2 x 3 cm. The lesions were non-tender and the local temperature was not raised. The oral, genital and ocular mucosae were however spared. The scalp and nails were also not involved. Investigations revealed that the haematological and biochemical parameters were within normal limits. Biopsy of the lesion showed presence of a cornoid lamella (Figure 3), which typically manifests as a thin keratotic rim at the periphery of a slightly atrophic shows a column of homogenous skin lesion and parakeratotic cells. Stratum granulosum was absent below the cornoid lamella. The patient was given a sitting of carbon dioxide laser to which he responded well.



Figure 1: Multiple atrophic macules on face.



Figure 2: Multiple annular hyperpigmented macules surrounded by a well-demarcated keratotic wall.

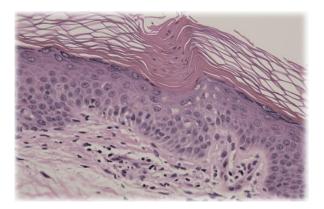


Figure 3: H & E staining showing cornoid lamella.

DISCUSSION

Disseminated superficial actinic porokeratosis is the most common porokeratosis type and was first defined by Chernosky, in 1966. 15 DSAP presents as irregular, threadlike raised rings surrounding discolored skin. The lesions most often present in the third decade of life in patients with chronic sun exposure; 15 a non-actinic form has been described, usually in the context of immunocompromised state. 16 Typically, the lesions are greater in number and are visible in areas with greater sun exposure. Superficial skin lesions bounded by a distinct peripheral ridge involving the palms and soles are the hallmark of Porokeratosis Plantaris, Palmaris et Disseminate (PPPD).¹⁷ Ionizing radiation of all types has also been linked to the development of DSAP, including natural¹⁸ or artificial¹⁹ ultraviolet radiation and excessive the rapeutic $\ensuremath{\text{photon}}^{20}$ or electron-beam $\ensuremath{^{21}}$ irradiation. The loss of heterozygosity model has been proposed to explain the link between ionizing radiation and porokeratosis.²² The prominent histopathological feature of all porokeratoses is the development of a "cornoid lamella". There is marked hyperkeratosis, parakeratosis, and acanthosis in the epidermis. The epidermis is atrophic in the central region. The cornoid lamella is less prominent in DSAP. Emollients and keratolytics can be used for treatment. Excision and grafting, cryotherapy, cauterization, dermabrasion, and carbon dioxide laser are the other treatments used. Oral retinoids have an inhibitory effects on cutaneous carcinogenesis. 23,24 We used carbon - dioxide laser in our case. DSAP, which is a rare disease, can often be misdiagnosed as other skin diseases such as actinic keratoses that appear in sunexposed areas. Therefore, in suspected cases a biopsy should be performed and protection from sunlight should be advised.

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