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CASE REPORT

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Disseminated superficial actinic porokeratosis: a case report

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. Since it is a rare disorder, herein we present our experience of a 57-year-old man with DSAP. Dermatological examination of the patient revealed pink/red multiple papular lesions, ranging in diameter from 0.5 to 1 cm, with an atrophic center and hyperkeratotic contours limited to sun-exposed skin parts especially in the extremities and rarely in trunk. Cryotherapy was effective in healing the lesions.

Key Words: Disseminated superficial actinic porokeratosis, porokeratoses, keratinization disorders

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Dissemine süperfisyel aktinik porokeratozis: vaka raporu

Özet: Dissemine süperfisyel aktinik porokeratoz, hayatın genellikle üçüncü ve dördüncü dekatında gözlenen ve vücudun güneş gören bölgelerinde ortaya çıkan bir porokeratoz tipidir. Nadir gözlenen bir olgu olduğu için, burada 57 yaşında DSAP'lı bir erkek hasta sunuyoruz. Hastanın dermatolojik incelemesinde çapları 0,5-1 cm arasında değişen, güneş gören bölgelerle sınırlı, özellikle ekstremitelerde ve nadiren gövdede, merkezi atrofik, kenarları hiperkeratotik, pembe-kırmızı çok sayıda papüler lezyonlar gözlendi. Kryoterapi lezyonların iyileşmesinde başarılı oldu.

Anahtar Sözcükler: Dissemine süperfisyel aktinik porokeratoz, porokeratoz, keratinizasyon hastalıkları

Introduction

Porokeratosis is a rare keratinization disorder with a clinical appearance of papules surrounded by a well-demarcated 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, 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). Patients with lesions in areas not exposed to the sun have also been reported. Immunosuppression is known to exacerbate the disease (5,6). DSAP has been reported in patients with AIDS, cirrhosis, and Crohn's disease, and those who have undergone organ transplants (7,8).

Case

A 57-year-old male patient presented to our outpatient department with lesions that had started a year ago and spread to the whole body. The

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dermatological examination revealed pink to red multiple papular lesions surrounded by a mildly hyperkeratotic wall, with an atrophic center, 0.5 to 1 cm in size, limited mostly to areas exposed to the sun, generally localized to the extremities with some on the trunk. There were no lesions on the face, mucosae, soles, or palms (Figure). It was notable that the lesions increased in summer. The histopathological examination of biopsy material from one of the lesions revealed hyperkeratosis, minimal papillomatosis, and minimal irregular acanthosis in an epidermis that showed invagination to the dermis in parts together with a decreased granular layer in these areas, and elevated parakeratotic layer in columns on the surface. There were mononuclear inflammatory cell infiltrates in perivascular areas of the upper dermis. These histopathological findings were consistent with porokeratosis. Cryotherapy was effective in healing the lesions.

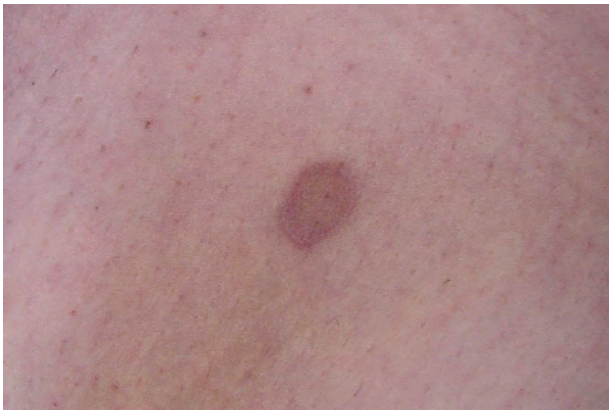

Figure. An erythematous plaque with hyperkeratotic border and mild central atrophy.

Discussion

DSAP is the most common porokeratosis type and was first defined by Chernosky, in 1966 (9). Atypical DSAP lesions have rarely been reported. Schuldenfrei et al. have reported a patient with exudative hyperkeratotic plaques (10). Kang et al. have reported a patient with lesions similar to prurigo nodularis (2). DSAP prevalence is not fully known and it is thought to be generally misdiagnosed as actinic keratosis in areas exposed to the sun (11).

The pathogenesis of porokeratosis has not been fully explained yet, but both epidermal and dermal abnormalities have been shown to play a role in the pathogenesis with DNA polyploidy, and keratinocyte and fibroblast sensitivity to ionizing radiation in the affected skin (12). Squamous cell carcinoma, basal cell carcinoma, or Bowen's disease may develop in approximately 7.5% of patients and the risk of malignancy is higher with linear porokeratosis (13).

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 (1). This case was consistent with DSAP as the patient was 57 years old and the lesions were mostly localized to areas exposed to the sun. The histopathological findings were also consistent with porokeratosis.

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 inhibitory effects on cutaneous carcinogenesis (14,15). We used cryotherapy in our case.

DSAP, which is a rare disease, can often be misdiagnosed as other skin diseases such as actinic keratoses that appear in sun-exposed areas. Therefore, in suspected cases a biopsy should be performed and protection from sunlight should be advised.

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