

Disseminated superficial porokeratosis involving the groin and genitalia in a 72-year-old immunocompetent man

Laura Gray Pruitt, BS, Ling-Lun Bob Hsia, MD, and William A. Burke, MD
Greenville, North Carolina

Key words: cornoid lamella; disseminated superficial actinic porokeratosis; disseminated superficial porokeratosis; porokeratosis; porokeratosis ptychotropica; topical diclofenac.

INTRODUCTION

The porokeratoses are a group of disorders of abnormal epidermal keratinization resulting in the characteristic histologic feature of the cornoid lamella. Several clinical variants of porokeratosis exist, which are unified by this common histologic feature but differ in morphology, distribution, and clinical course. The typical lesions of porokeratosis are characterized by an atrophic center surrounded by an elevated keratotic rim formed by the cornoid lamella. The lesions can be found almost anywhere on the body; however, the groin and genitalia are rarely involved. We report an unusual presentation of a rare clinical variant, disseminated superficial porokeratosis (DSP).

CASE REPORT

A 72-year-old white man with skin type II presented for evaluation of erythematous round to ovoid macules with collarettes of scale measuring approximately 0.5 to 2 cm in size, most prominently involving the lower extremities, buttocks, groin area, and penis that had been present for more than 13 years (Figs 1 and 2). He also had scattered lesions on his upper extremities and trunk. The lesions were associated with pruritus and paresthesias, especially in the groin. The patient was otherwise well, with hypertension, arthritis, and hypothyroidism being his only medical problems. His medications included acetaminophen, aspirin, levothyroxine, meloxicam, enalapril, and metoprolol. He denied any history of skin cancer, radiation, or heavy sun exposure or family history of a similar rash. He denied any changes in the lesions with sun exposure.

Abbreviations used:

DSAP: Disseminated superficial actinic porokeratosis
DSP: Disseminated superficial porokeratosis

On initial examination via interactive telemedicine, porokeratosis was suspected, and the patient was empirically treated with triamcinolone acetate 0.1% cream for the extremities and hydrocortisone valerate 0.2% cream for the groin region. His pruritic symptoms were moderately controlled with the topical steroids; however, there was no improvement in the number or appearance of lesions. Because of the atypical distribution, he was seen in the clinic for further evaluation and biopsy.

A shave biopsy of a lesion on the thigh was performed, and histology testing found well-formed cornoid lamellae with dyskeratotic cells at their base, confirming the diagnosis of porokeratosis (Fig 3). Based on the course and distribution of his disease, he was given a diagnosis of DSP.

DISCUSSION

Vittorio Mibelli first described porokeratosis and its characteristic histologic finding of cornoid lamellae in 1893.¹ Disseminated superficial actinic porokeratosis (DSAP) is the most common variant of porokeratosis. Its onset is usually in the third or fourth decade of life and involves the sun-exposed skin. It is often both induced and exacerbated by exposure to ultraviolet light, although the face is

East Carolina University Dermatology.

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Correspondence to: Ling-Lun Bob Hsia, MD, East Carolina University Dermatology, 517 Moye Blvd- 3rd Floor, Greenville, NC 27834. E-mail: HSIAL@ECU.EDU.

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Fig 1. Disseminated superficial porokeratosis. Multitude of erythematous round to ovoid macules with collarettes of scale on the thigh and leg.



Fig 2. Disseminated superficial porokeratosis. Similar lesions on the scrotum and penis.

involved in only 15% of cases.^{2,3} DSP, a rare variant, is distinguished from DSAP by its involvement of both sun-protected and sun-exposed areas of the body. Furthermore, DSP tends to present in children or immunocompromised patients.²

The lesions of porokeratosis may manifest on almost any area of the body; however, groin or genital involvement is rare, even in disseminated forms of the disease.⁴ Only 20 cases of porokeratosis confined to the genitalia have been reported in the literature, with lesions generally involving the scrotum but not the penile shaft as in our patient.⁵⁻⁷ Because of the location of the lesions, cases have been misdiagnosed early as condyloma, syphilis, granuloma annulare, and eczema.⁶ Porokeratosis ptychotropica is a variant that has been reported to involve the groin. It most commonly manifests in the flexural areas of the genitogluteal region as pruritic verrucous confluent plaques with satellite papules.¹ Linear porokeratosis may also appear on the buttocks but is characterized by large plaques in a linear distribution.⁸ Of note, our patient did have involvement of the buttocks, but the lesions had the same guttate morphology as in other areas of the body and were not consistent with the

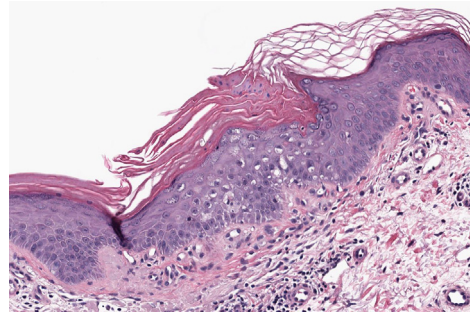


Fig 3. Porokeratosis. Shave biopsy section shows a characteristic cornoid lamella. (Hematoxylin-eosin stain; original magnification: $\times 200$.)

porokeratosis ptychotropica or linear porokeratosis variants.

Our patient represents a rare presentation of porokeratosis. DSP most commonly presents between the ages of 5 and 10 years^{1,2} but may develop later in life in the setting of immunocompromise.^{2,7} However, our patient had DSP in his sixth decade of life and has no immunocompromising conditions. Our clinical photographs capture a rare presentation involving the genitalia, including the penile shaft.

Although the specific etiology is unknown, the porokeratoses are thought to originate from clonal proliferation of abnormal keratinocytes.² Risk factors for porokeratosis include family history, light-colored skin, ultraviolet or ionizing radiation exposure, internal malignancies, and immunosuppression.^{2,3,9} The porokeratoses are typically asymptomatic but some patients may experience pain, pruritus, or cosmetic concerns, which are among the most common indications for treatment.² Several topical, systemic, and surgical treatment modalities have been described; however, treatment is challenging, as the lesions are often refractory to therapeutic intervention.^{9,10} Recently, diclofenac sodium 3% gel was found to be successful in stabilizing the lesions of DSAP and genital porokeratosis and providing symptomatic relief.⁹ Unfortunately, spontaneous regression is rare, and the lesions typically persist indefinitely without treatment.¹⁰ The most important indication for treatment is transformation into squamous cell carcinoma or basal cell carcinoma.² All of the clinical variants of porokeratosis have the potential for malignant transformation with an overall incidence of 7.5% to 11%.^{1,2,9}

We report this case to show that porokeratosis can affect the buttocks, groin, and genitalia. Because of the atypical location, it may be initially misdiagnosed, and a correct diagnosis is important to treat appropriately and monitor for the risk of malignant transformation.

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