Punctate porokeratosis—pruritic and hyperkeratotic papules on the palms and feet

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ABSTRACT

Porokeratosis is an uncommon cutaneous condition with multiple clinical variants that is defined by round patches with a raised, fine scaling border. Punctate porokeratosis is a rare variant of porokeratosis that is characterized by elevated, seed-like lesions of the palms and soles. While variants of porokeratosis may be associated with an increased risk of squamous cell carcinoma within the lesion, punctate porokeratosis does not increase the risk of squamous cell carcinoma. However, punctate porokeratosis may mimic other dermatoses affecting the palms and soles including spiny keratoderma, arsenical keratosis, and nevus comedonicus. Distinguishing punctate porokeratosis and spiny keratoderma is important, as spiny keratoderma may be associated with underlying solid organ malignancy or chronic medical conditions, including chronic kidney disease. Here, we present a case of punctate porokeratosis mimicking spiny keratoderma in order to distinguish the two conditions and aid in their differentiation from other dermatologic conditions involving the palms and soles.

KEYWORDS Porokeratosis; porokeratosis palmaris et plantaris disseminata; porokeratotic punctate keratoderma; punctate palmoplantar keratoderma type 2; punctate porokeratosis; spiny keratoderma

Punctate porokeratosis (PP), a rare form of porokeratosis initially described by Rahbari et al in 1977, is clinically characterized by linear or diffuse, tender, seed-like lesions with elevated borders on the palms and soles. Historically, PP has been referred to by varying names including porokeratosis puncta palmaris et plantaris and porokeratotic punctate keratoderma, which have been used interchangeably to refer to spiny keratoderma, a clinically distinct condition. While PP and spiny keratoderma may present similarly, their distinction is essential, as spiny keratoderma may be associated with underlying malignancy and chronic medical conditions, while PP is primarily of cosmetic concern. A case of PP is presented to aid in recognition of this rare condition and to highlight the distinguishing factors between PP, spiny keratoderma, and other dermatoses involving the palms and soles, including keratosis puncta of the palmar creases, arsenical keratosis, and nevus comedonicus.

CASE DESCRIPTION

An 89-year-old black woman with type 2 diabetes mellitus presented to the dermatology clinic for evaluation of hyperkeratotic and filiform papules on the bilateral palms and soles. These lesions had been present for approximately 15 years, were mildly

tender to pressure, and were occasionally pruritic with exposure to heat. Her family history was significant for similar lesions on the hands and feet of both her mother and sister, which were more linear in configuration and less diffuse. She had never pursued treatment but had been paring down the palmar lesions for many years.

Physical examination revealed diffuse hyperpigmented, hyperkeratotic papules with central pitting and raised borders on the bilateral palms and ventral surface of the digits with sparing of the dorsal aspect of the hands (*Figure 1a*). Hyperkeratotic papules were also present in the interdigital spaces and plantar surface of the toes of the bilateral feet (*Figure 1b*). Histopathologic examination of a 3 mm punch biopsy from the right palmar thenar eminence revealed columns of parakeratosis with underlying hypogranulosis, as well as scattered dyskeratotic keratinocytes, which was most consistent with a diagnosis of PP (*Figure 2*).¹

DISCUSSION

On histopathology, porokeratosis is distinguished by characteristic coronoid lamellae, a parakeratotic column with loss of the granular layer and dyskeratotic keratinocytes.² Diagnosis of PP is confirmed with biopsy; however, preliminary evaluation

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Figure 1. (a) Numerous hyperpigmented, hyperkeratotic papules with central pitting and raised borders across bilateral palms. (b) Hyperkeratotic papules of the interdigital spaces of the left foot.

Figure 2. (a) Histopathology displaying hyperkeratosis with columns of parakeratosis ($\times 10$). (b) Columns of parakeratosis with focal underlying hypogranulosis and rare dyskeratotic keratinocytes ($\times 100$).

with dermoscopy may reveal annular, keratotic papules with central tan-brown globules and a “white-track” periphery that accentuates with the application of Gentian violet.³

Porokeratosis palmaris et plantaris disseminata (PPPD) is a variant of PP that involves the palmoplantar and truncal body surfaces, although PPPD is also hypothesized to represent a progressive form of PP.^{4,5} PP and PPPD were previously referred as “porokeratosis puncta palmaris et plantaris” and “porokeratotic punctate keratoderma,” and accordingly, historical cases may carry varying nomenclature.⁶ However, these terms are best avoided, as they are also synonyms used to describe spiny keratoderma/punctate palmoplantar keratoderma type 2.⁶ Spiny keratoderma is an autosomal dominant condition clinically presenting with filiform, hyperkeratotic “music box” spines that may be associated with an underlying malignancy, type IV lipoproteinemia, type 2 diabetes, and polycystic kidney disease.⁷ While clinically and histologically comparable to PP, spiny keratoderma is a distinct diagnosis that exhibits a similarly diminished stratum granulosum and columns of parakeratosis but usually lacks the dyskeratotic keratinocytes of PP.^{6,8}

Other clinical entities may mimic PP, including keratosis puncta of the palmar creases, arsenical keratosis, and nevus comedonicus. Keratosis puncta of the palmar creases is a benign condition and variant of punctate keratoderma that occurs most commonly in African American patients. It is typified by hyperkeratotic pits limited to the palmar creases, as opposed to the

diffuse palmoplantar lesions in our patient. Arsenical keratosis occurs due to chronic ingestion of arsenic most commonly via contaminated well water and presents with pigmentary changes of truncal skin and mucous membranes, palmoplantar keratosis, and Mee’s lines of the nails.⁹ Arsenical palmoplantar keratoses range in clinical appearance from indurated, gritty millimeter palmoplantar papules to yellow, verrucous papules and plaques.⁹ On histology, arsenical keratosis is characterized by compact hyperkeratosis and a thickened stratum granulosum but lacks the columns of parakeratosis seen in our case.⁹ Nevus comedonicus is a hamartomatous growth of the pilosebaceous unit distinguished by dilated follicles with pigmented, keratinaceous plugs and histopathology displaying epidermal invagination, hyperkeratosis and follicular plugging, which is not seen in this case.

The management of porokeratosis is generally unsatisfactory, with lesions frequently recurring upon cessation of treatment.² Certain variants of porokeratosis may carry increased risk of cutaneous malignancy, but treatment of PP may be primarily of cosmetic value rather than therapeutic, as there is no documented risk of malignant transformation in PP.¹⁰ Treatment options for porokeratosis include ablative therapies (surgical excision, cryotherapy, laser ablation) and topical therapies (vitamin D analogs, photodynamic therapy, 5-fluorouracil, imiquimod, retinoids, tacrolimus).²

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