Symmetric Palmoplantar Papules With a Keratotic Border

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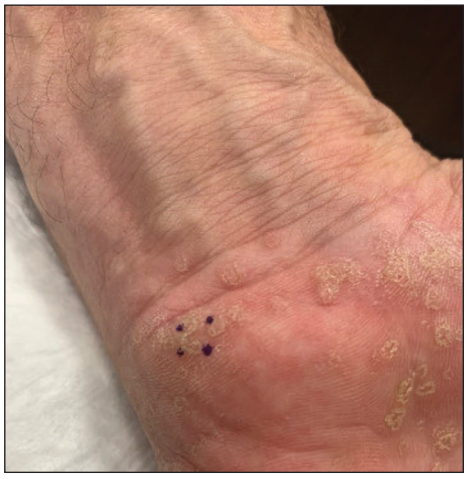

A 67-year-old man presented to our office with a rash on the hands, feet, and periungual skin that began with wartlike growths many years prior and recently had started to involve the proximal arms and legs up to the thighs as well as the trunk. He had a medical history of essential hypertension and chronic obstructive pulmonary disease. He had an 18-year smoking history and had quit more than 25 years prior, with tongue cancer diagnosed more than 5 years prior that was treated with surgery, chemotherapy, and radiation. The lesions occasionally were itchy but not painful. He also reported that his nails frequently split down the middle. He denied any oral lesions and was not using any treatments for the rash. He had no history of skin cancer or other skin conditions. His family history was unclear. Physical examination revealed annular red-pink scaling with a keratotic border on the soles of the feet, palms, and periungual skin. There also were small hyperpigmented papules on the arms, legs, thighs, and trunk over a background of dry and discolored skin, as well as dystrophy of all nails.

WHAT'S YOUR **DIAGNOSIS?**

a. Darier disease
b. disseminated superficial actinic porokeratosis
c. pityriasis rubra pilaris
d. porokeratosis plantaris palmaris et disseminata
e. psoriasis

PLEASE TURN TO PAGE 203 FOR THE DIAGNOSIS

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The authors report no conflict of interest.

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doi:10.12788/cutis.0747

THE **DIAGNOSIS:** Porokeratosis Plantaris Palmaris et Disseminata

A 3-mm punch biopsy of the right upper arm showed incipient cornoid lamellae formation, pigment incontinence, and sparse dermal lymphocytic inflammation (Figure), suggestive of porokeratosis plantaris palmaris et disseminata (PPPD). The dermatopathologist recommended a second biopsy to confirm the diagnosis and to confirm that the lesions on the palms and soles also were suggestive of porokeratosis. A second 4-mm punch biopsy of the left palm was consistent with PPPD.

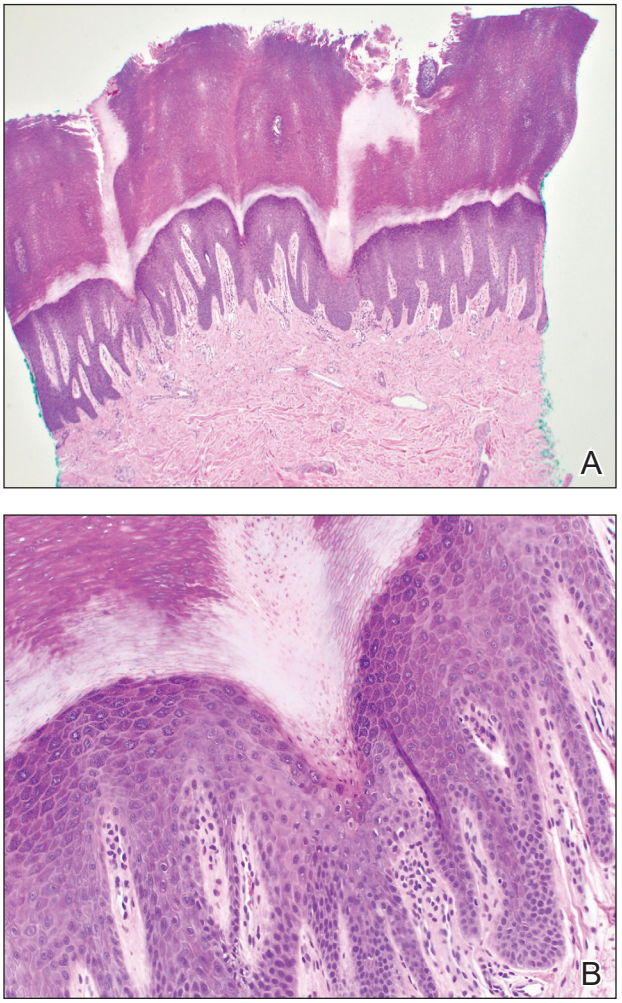
The risks of PPPD as a precancerous entity along with the benefits and side effects of the various management options were discussed with our patient. We recommended that he start low-dose isotretinoin (20 mg/d) due to the large body surface area affected, making focal and field treatments likely insufficient. However, our patient opted not to treat and did not return for follow-up.

Subtypes of porokeratosis, including disseminated superficial actinic porokeratosis (DSAP) and PPPD, are conditions that disrupt the normal maturation of keratin and present clinically with symmetric, crusted, annular papules.[1] The signature but nonspecific histopathologic feature shared among the subtypes is the presence of a cornoid lamellae.[2] Several triggers of porokeratosis have been proposed, including trauma and exposure to UV and ionizing radiation.[2,3] The clinical variants of porokeratosis are important conditions to diagnose correctly because they portend a risk for Bowen disease and invasive squamous cell carcinoma and may indicate the presence of an underlying hematologic and/or solid organ malignancy.[4] Management of porokeratosis is difficult, as treatments have shown limited efficacy and variable recurrence rates. Treatment options include focal, field, and systemic options, such as 5-fluorouracil, topical compound of cholesterol and lovastatin, isotretinoin, and acitretin.[1,2]

Porokeratoses may arise from gene mutations in the mevalonate pathway,[5] which is essential for the production of cholesterol.[6] Topical cholesterol alone has not been shown to improve porokeratosis, but the combination topical therapy of cholesterol and lovastatin is promising. It is theorized to deliver benefit by both providing the essential end product of the pathway and simultaneously reducing the number of potentially toxic intermediates.[6]

Porokeratosis plantaris palmaris et disseminata (also known as porokeratosis plantaris) is unique among the subtypes of porokeratosis in that its annular, red-pink, papular rash with scaling and a keratotic border tends to start distally, involving the palms and soles, and progresses proximally to the trunk with smaller lesions.[1,7] This centripetal progression can take years, as was seen in our patient.[1] The disease is uncommon, with a dearth of published reports on PPPD.[2] However, case reports have shown that PPPD is strongly linked to family history and may have an autosomal-dominant inheritance pattern. Penetrance is greater in men than in women, as PPPD is twice as common in men.[8] Most cases of PPPD have been diagnosed in patients in their 20s and 30s, but Hartman et al[9] reported a case wherein a patient was diagnosed with PPPD after 65 years of age, similar to our patient.

Although the lesions in DSAP can appear similar to those in PPPD, DSAP is more common among the family of porokeratotic conditions, affecting women twice as often as men, with a sporadic pattern of inheritance.[2] These same



A and B, Histopathology showed incipient cornoid lamellae formation, pigment incontinence, and sparse dermal lymphocytic inflammation, suggestive of porokeratosis (H&E, original magnifications ×40 and ×200, respectively).

features are present in some other types of porokeratosis but not PPPD. Furthermore, DSAP progresses proximally to distally but often with truncal sparing.[2]

Akin to PPPD, pityriasis rubra pilaris (PRP) often presents with palmoplantar keratoderma.[10] There are at least 6 types of PRP with varying degrees of similarity to PPPD. However, in many cases PRP is associated with a background of diffuse erythema on the body with islands of spared skin. In addition, cases of PRP have been linked to extracutaneous findings such as ectropion and joint pain.[11]

Darier disease, especially the acrokeratosis verruciformis of Hopf variant, is more common in men and involves younger populations, as in PPPD.[11] However, the crusted lesions seen in Darier disease frequently involve the skin folds. These intertriginous lesions may coalesce, mimicking warts in appearance, and are at risk for secondary infection. Nail findings in Darier disease also are distinct and include longitudinal white or red stripes running along the nail bed, in addition to *V*-shaped nicks at the nail tips.

Psoriasis can occur anywhere on the body and is associated with silver scaling atop a salmon-colored dermatitis.[12] It results from aberrant proliferation of keratinocytes. Some distinguishing features of psoriasis include a disease course that waxes and wanes as well as pitting of the nails.

Although PPPD typically affects young adults, we presented a case of PPPD in an older man. Porokeratosis plantaris palmaris et disseminata in older adults may represent a delayed diagnosis, imply a broader range for the age of onset, or suggest its manifestation secondary to radiation treatment or another phenomenon. For example, our patient received 35 radiotherapy cycles for tongue cancer more than 5 years prior to the onset of PPPD.

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