Concurrent Punctate Keratosis of the Palmar Creases and Focal Acral Hyperkeratosis

Pooja Khera, MD; Gennady Shiferman, BS; Joseph C. English III, MD

We report a patient with punctate keratosis of the palmar creases appearing concurrently with focal acral hyperkeratosis. This presentation is a unique coexistence of 2 simple and punctate hereditary palmoplantar keratodermas.

Cutis. 2008;81:348-350.

Case Report

A 35-year-old black woman presented to our dermatology clinic for atopic dermatitis. On physical examination, she was incidentally noted to have multiple, 2- to 5-mm round depressions filled with conical keratinous plugs along the creases of her palms (Figure 1). Additionally, she had numerous polygonal crateriform papules along the lateral borders of her wrists and feet (Figure 2). The patient denied associated pain, pruritus, and hyperhidrosis. The lesions had been present since early childhood and gradually increased in number over the years. Her mother and several cousins had similar lesions. The clinical findings were consistent with punctate keratosis of the palmar creases and focal acral hyperkeratosis. Because the lesions were asymptomatic, the patient declined treatment.

Comment

Hereditary palmoplantar keratodermas are characterized by an abnormal thickening of the skin on the palms and soles. They can be classified into 3 clinically distinct patterns: (1) diffuse, characterized by an even, thick, symmetric hyperkeratosis over the palms and soles; (2) focal, consisting of localized areas of hyperkeratosis over pressure points; and (3) punctate, characterized by tiny (1 mm to 1 cm) hyperkeratotic papules on the palms and soles. Additionally, these keratodermas can be classified as simple (only the skin is involved), complex (in association with lesions of nonvolar skin, hair, teeth, nails, and sweat glands), and syndromic (with other organ involvement, deafness, or cancer).[1] Punctate keratosis of the palmar creases and focal acral hyperkeratosis are both simple hereditary palmoplantar keratodermas (Table).

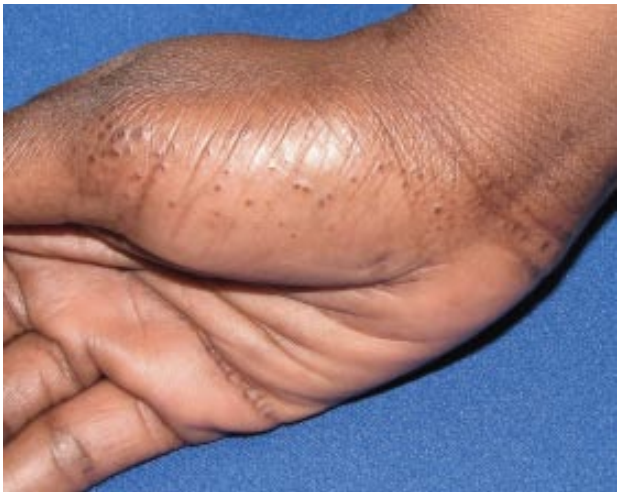

Figure 1. Concurrent punctate keratosis of the palmar creases and focal acral hyperkeratosis.

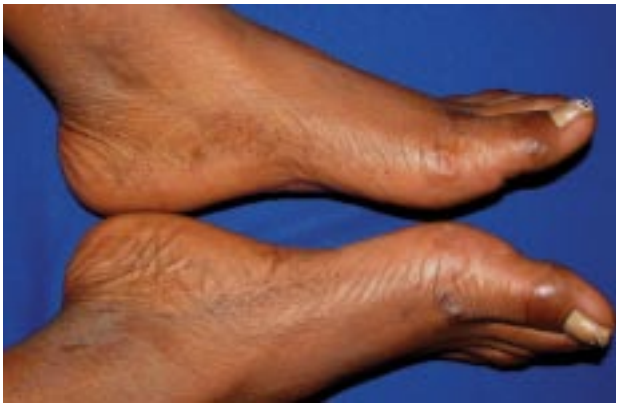

Figure 2. Polygonal crateriform papules of focal acral hyperkeratosis along the lateral borders of the feet.

Accepted for publication November 13, 2007.

From the Department of Dermatology, University of Pittsburgh, Pennsylvania.

The authors report no conflict of interest.

Correspondence not available.

Simple Hereditary Palmoplantar Keratodermas

Simple Hereditary PPKs	Inheritance	Gene	Clinical Features
Diffuse PPKs			
Unna-Thost syndrome (nonepidermolytic)	AD	*KRT1* (diffuse) *KRT16* (focal)	Thick fissured hyperkeratosis with erythematous border; nontransgrediens; hyperhidrosis; secondary dermatophyte infection
Vörner type (epidermolytic)	AD	*KRT1, KRT9*	Clinically identical to Unna-Thost type but histology shows EHK
Mal de Meleda	AR	*SLURP1*	PPK with transgrediens; pseudoainhum; hyperkeratotic plaques on elbows and knees
Focal PPKs			
Striate form	AD	*DSG1, DSP, KRT1*	Linear thickening on palms; islandlike areas of the soles
Punctate PPKs			
Punctate keratoses of the palms and soles	AD	Unknown	Tiny hyperkeratotic papules over entire palmar surface; pruritus is common
Spiny keratoderma	AD	Unknown	Multiple tiny keratotic plugs mimicking the spines on a music box
Punctate keratosis of the palmar creases	AD	Unknown	Round depressions filled with conical keratinous plugs along the creases of the palms, fingers, and soles; associated with atopic dermatitis or manual labor; most common in black individuals
Focal acral hyperkeratosis	Sporadic, AD	Unknown	Oval or polygonal, crateriform papules along the lines of transgredience of the palms, soles, and digits; most common in black individuals
Acrokeratoelastoidosis of Costa	Sporadic, AD	Unknown	Clinically identical to focal acral hyperkeratosis with elastorrhexis on histology

Abbreviations: PPKs, palmoplantar keratodermas; AD, autosomal dominant; *KRT1,* keratin 1; *KRT16,* keratin 16; *KRT9,* keratin 9; EHK, epidermolytic hyperkeratosis; AR, autosomal recessive; *SLURP1,* secreted leukocyte antigen-6/urokinase-type plasminogen activator urokinase receptor domain-containing protein 1; *DSG1,* desmoglein 1; *DSP,* desmoplakin.

Punctate keratosis of the palmar creases most commonly occurs in black individuals aged 19 to 49 years.[2,3] It has an autosomal dominant inheritance pattern and may be associated with atopic dermatitis[4] or manual labor.[5] Primary lesions are small round depressions filled with conical keratinous plugs along the creases of the palms, fingers, and soles. They may be aggravated by friction and occasionally present with pain. Histologically, there are hyperkeratotic plugs that cause depression of the epidermis.[3] Topical retinoids, keratolytic agents, and punch excisions have been used for treatment of symptomatic lesions with variable success.[6]

Focal acral hyperkeratosis is a term coined by Dowd et al[7] in 1983. Focal acral hyperkeratosis can be sporadic or inherited in an autosomal dominant pattern. It most commonly affects black individuals and manifests in childhood or adolescence. Clinically, the lesions are oval or polygonal, firm, yellowish crateriform papules along the lines of transgredience of the palms, soles, and digits.[7] Hyperhidrosis may be associated with this condition.[8] Histologically, there are acanthotic clavus-like depressions in the epidermis. It is thought to be a variant of acrokeratoelastoidosis of Costa, lacking the histologic finding of elastorrhexis (decrease or fragmentation of dermal elastic fibers).[8] Various treatments have been used, including salicylic acid, cryotherapy, tretinoin, oral etretinate, and prednisone, without long-term success.[9]

Dowd et al[7] described 15 patients with focal acral hyperkeratosis. Interestingly, a photograph of one of the patients also depicted lesions consistent with punctate keratosis of the palmar creases. Dowd et al[7] did not comment on this additional finding; however, the coexistence of the 2 entities in the photograph was subsequently noted.[10] To our knowledge, there are no other reports where both conditions are simultaneously noted. Both palmoplantar keratodermas can be familial (acquired palmoplantar keratoderma has been previously published by our institution),[11] have a similar racial distribution, and typically are asymptomatic. They share many clinical and histologic features and may represent 2 different manifestations of a similar underlying pathogenesis.

REFERENCES

1. Stevens HP, Kelsell DP, Leigh IM. The inherited keratodermas of palms and soles. In: Freedberg IM, Eisen AZ, Wolff K, et al, eds. *Fitzpatrick's Dermatology in General Medicine*. 6th ed. New York, NY: McGraw-Hill; 2003: 505-514.
2. Weiss RM, Rasmussen JE. Dermatosis punctata of the palmar creases. *Arch Dermatol*. 1980;116:669-671.
3. Penas PF, Rios-Buceta L, Sanchez-Perez J, et al. Keratosis punctata of the palmar creases: case report and prevalence study in Caucasians. *Dermatology*. 1994;188:200-202.
4. Anderson WA, Elam MD, Lambert WC. Keratosis punctata and atopy: report of 31 cases with a prospective study of prevalence. *Arch Dermatol*. 1984;120:884-890.
5. Smith EB, Jetton RL. Punctate pits and keratoses of the palmar creases. *South Med J*. 1970;63:1291-1293.
6. James WD, Berger TG, Elston DM. Parapsoriasis, pityriasis rosea, pityriasis rubra pilaris. In: James WD, Berger TG, Elston DM, eds. *Andrews' Diseases of the Skin: Clinical Dermatology*. 10th ed. Philadelphia, PA: WB Saunders; 2005:208-229.
7. Dowd PM, Harman RR, Black MM. Focal acral hyperkeratosis. *Br J Dermatol*. 1983;109:97-103.
8. Erkek E, Kocak M, Bozdogan O, et al. Focal acral hyperkeratosis: a rare cutaneous disorder within the spectrum of Costa acrokeratoelastoidosis. *Pediatr Dermatol*. 2004;21:128-130.
9. Natow S. Focal acral hyperkeratosis. *Dermatol Online J*. 2001;7(1):10.
10. Judge MR, McLean WHI, Munro CS. Disorders of keratinization. In: Burns T, Breathnach SM, Cox N, et al, eds. *Rook's Textbook of Dermatology*. Vol 2. 7th ed. Malden, MA: Blackwell Science Ltd; 2004:34.1-34.111.
11. Patel S, Zirwas M, English JC 3rd. Acquired palmoplantar keratoderma. *Am J Clin Dermatol*. 2007;8:1-11.