New-onset acrokeratoelastoidosis in an immunosuppressed patient

Aamir Hussain, BA,[a] Ashley Jenkins, MD,[b] Ashley Feneran, DO,[b] and Farah Abdulla, MD[b]
Chicago, Illinois

Key words: acrokeratoelastoidosis; asymptomatic papules; Crohn's disease; dermatopathology; focal acral hyperkeratosis; hyperkeratosis; immunosuppression; keratoderma; pathology; vedolizumab.

INTRODUCTION

Acrokeratoelastoidosis (AKE), is a rare keratoderma characterized by small, round, skin-colored papules on the palms and soles.[1] It was first described in 1952 by Brazilian dermatologist Oswaldo Costa and commonly presents before the age of 20.[2] The primary etiology is unknown, and both autosomal dominant and sporadic forms have been reported in the dermatology literature.[1,3] We present the case of an immunosuppressed patient who subsequently had papules on the hands, originally diagnosed as verruca palmoplantaris, that were later found to be consistent with AKE.

Abbreviation used:

AKE: acrokeratoelastoidosis

CASE REPORT

The patient is a 41-year-old woman with a history of Crohn's disease, taking vedolizumab, who presented to the dermatology clinic for evaluation of bilateral, asymptomatic papules that had been present on the palms of her hands for 4 years. The patient had previously been diagnosed clinically with verruca plana and started on topical imiquimod. Despite strict adherence to the treatment regimen of imiquimod, 3 times a week to affected areas, the patient stated that the papules had not changed.

On physical examination, the patient showed several 1- to 3-mm, smooth, flat-topped, barely raised, pinpoint skin-colored papules predominately concentrated near the periphery of the bilateral palms at the hypothenar eminences and some scattered near the digits (Fig 1).

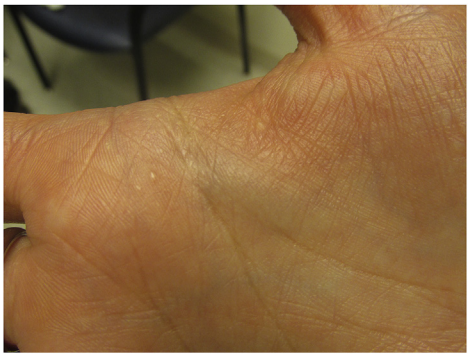

Fig 1. Smooth, flat-topped, skin-colored pinpoint papules located near the periphery of the palm.

The differential diagnosis included focal acral hyperkeratosis, keratosis punctata, and aberrant eccrine glands. A biopsy of one of the lesions found compact hyperkeratosis and a prominent granular layer. Focal areas of the epidermis showed invagination with hyperkeratotic plugs with clear relationships to the acrosyringia (Fig 2, *A* and *B*). Although the pathology report was nonspecific, the association with the acrosyringium favored the diagnosis of AKE. Verhoeff-van Gieson stains were significant for fragmented elastic fibers within the dermis (Fig 3).

DISCUSSION

AKE is part of a group of keratodermal disorders that affect the palmar, plantar, and marginal surfaces.[3] It is related to focal acral hyperkeratosis, a very similar disorder that typically presents earlier in life and typically lacks elastic changes.[1] The differential diagnosis of palmoplantar small, rhomboid, yellow to skin-colored papules also includes

From the University of Chicago, Pritzker School of Medicine[a] and the Department of Medicine, Section of Dermatology, University of Chicago.[b]

Funding sources: None.
Conflicts of interest: None declared.

Correspondence to: Aamir Hussain, BA, 5346 S Cornell Avenue, APT 305, Chicago, IL 60615. E-mail: aamirh@uchicago.edu.

JAAD Case Reports 2018;4:75-6.
2352-5126

http://dx.doi.org/10.1016/j.jdcr.2017.08.004

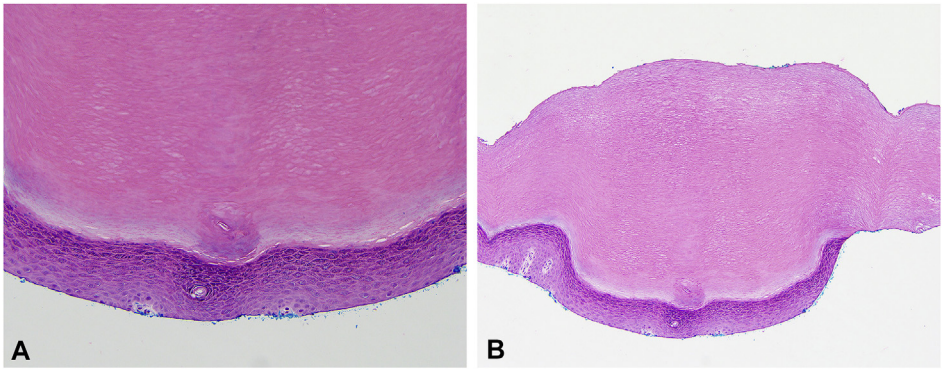

Fig 2. **A**, Sections of skin with compact hyperkeratosis, hypergranulosis, and a depression of the epidermis with a clear relationship to the acrosyringium. **B**, There is hyperkeratosis with a shallow depression of the underlying epidermis. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×20; **B**, ×10.)

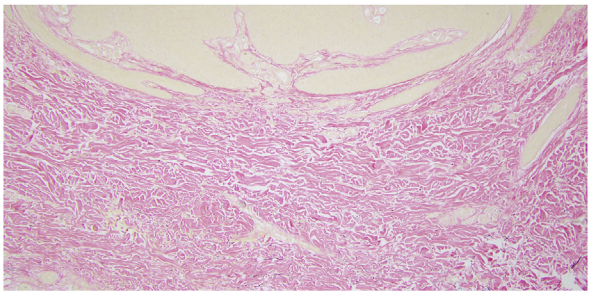

Fig 3. Elastic fibers are decreased in number, thin, and fragmented. (Hematoxylin-eosin with elastic stain; original magnification: ×10.)

degenerative collagenous plaques, keratoelastoidosis marginalis, and verruca palmoplantaris.[1,4] In case reports, AKE has been distinguished from degenerative collagenous plaques and keratoelastoidosis marginalis by distribution; the latter 2 disorders usually affect the index finger and opposing margin of the thumb.[5]

This patient originally had verruca vulgaris diagnosed because of the clinical appearance of the lesions and her immunocompromised state, which carries a higher risk for development of cutaneous HPV manifestations.[6] Indeed, palmoplantar wart flares have been observed specifically in patients immunosuppressed for Crohn's Disease.[7]

However, given that this patient's papules did not respond to imiquimod therapy, a biopsy was performed confirming our suspicion that the patient's papules were in fact not warts. This finding was significant, as, to our knowledge, this is the first instance of AKE arising after immunosuppression. A review of dermatoses in AIDS found that lower T-cell counts were associated with increasing prevalence of papulosquamous disorders, including keratoderma blenorrhagicum. Because AKE and keratoderma blenorrhagicum are both classified in the category of hyperkeratotic disorders, it is possible that AKE may also be associated with immunosuppression.[8] At this time, there is insufficient evidence to determine whether this patient has either the sporadic or autosomal dominant form of AKE. Although case reports have not found clinical distinctions between sporadic and hereditary forms of AKE, this patient's lack of family history indicates a sporadic etiology. In addition, this patient's age of presentation is atypical; most cases of AKE occur before age 30.[9] There is no effective treatment for AKE, although the condition is benign and does not seem to carry an increased risk for more serious diseases.[1]

REFERENCES

1. Hu W, Cook TF, Vicki GJ, Glaser DA. Acrokeratoelastoidosis. *Pediatr Dermatol*. 2002;19(4):320-322.
2. Costa OG. Akrokerato-elastoidosis; a hitherto undescribed skin disease. *Dermatologica*. 1953;107(3):164-168.
3. Greiner J, Krüger J, Palden L, Jung EG, Vogel F. A linkage study of acrokeratoelastoidosis. Possible mapping to chromosome 2. *Hum Genet*. 1983;63(3):222-227.
4. Shbaklo Z, Jamaleddine NF, Kibbi AG, Salman SM, Zaynoun ST. Acrokeratoelastoidosis. *Int J Dermatol*. 1990;29(5):333-336.
5. Handfield-Jones S, Kennedy CT. Acrokeratoelastoidosis treated with etretinate. *J Am Acad Dermatol*. 1987;17(5 Pt 2):881-882.
6. Wieland U, Kreuter A, Pfister H. Human papillomavirus and immunosuppression. *Curr Probl Dermatol*. 2014;45:154-165.
7. Timmer MR, van Ooteghem NAM. Recalcitrant plantar warts during azathioprine therapy for Crohn's disease. *Ann Gastroenterol*. 2013;26(2):173-174.
8. Sadick NS, McNutt NS, Kaplan MH. Papulosquamous dermatoses of AIDS. *J Am Acad Dermatol*. 1990;22(6):1270-1277.
9. Mervak JE, Lowe L, Cha KB. Chronic Translucent Papules of the Palms and Soles. *JAMA Dermatol*. 2014;150(9):1001-1002.