Atrophoderma Vermiculatum: A Case Report and Review of the Literature on Keratosis Pilaris Atrophicans

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GOAL

To understand atrophoderma vermiculatum (AV) to better manage patients with the condition

LEARNING OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical and histologic findings of AV.
2. Recognize the possible diseases associated with AV.
3. Differentiate AV from other variants of keratosis pilaris atrophicans.

INTENDED AUDIENCE

This CME activity is designed for dermatologists and generalists.

CME Test on page 89.

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Atrophoderma vermiculatum (AV) is a rare follicular disorder primarily affecting children with reticular or honeycomb atrophy of the cheeks and forehead. Along with keratosis pilaris atrophicans faciei (KPAF) and keratosis follicularis spinulosa decalvans (KFSD), AV falls within the broader spectrum of keratosis pilaris atrophicans (KPA). Although these 3 variants of KPA have unique presentations and associations, they can all be frustratingly difficult to treat. We describe a sporadic case of AV that presented in late adolescence, a relatively late age of onset.

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Keratosis pilaris atrophicans (KPA) is an uncommon and poorly understood group of disorders in which follicular keratotic papules (keratosis pilaris) result in scarring in the form of atrophy and/or alopecia. There are 3 entities of KPA: keratosis pilaris atrophicans faciei (KPAF), atrophoderma vermiculatum (AV), and keratosis follicularis spinulosa decalvans (KFSD).[1,2] All types of KPA tend to be associated with an atopic tendency and keratosis pilaris on the extensor surfaces of the extremities.[2,3]

Unlike the other types of KPA, patients with AV have atrophy without alopecia. Furthermore, AV is unique in that it typically presents in childhood or, more rarely, in adolescence or early adulthood, whereas onset of KPAF and KFSD is in infancy.

Case Report

A 20-year-old man presented with progressive atrophic scarring of both cheeks of 3 years' duration, associated with numerous follicular keratotic papules on the extensor surfaces of the arms and back (Figure 1). He was asymptomatic and otherwise healthy. He denied any history of acne or alopecia. He further denied a family history of any similar skin conditions.

A punch biopsy specimen of an atrophic keratotic papule on the upper back showed follicular hyperkeratosis with mild perifollicular fibrosis as well as perifollicular and perivascular chronic inflammation (Figure 2).

Comment

Atrophoderma vermiculatum was originally described as ulerythema acneform by Unna[4] in 1896 and was later coined "atrophodermie vermiculée" by Darier[5] in 1920. *Atrophoderma vermiculatum* has been the term used most frequently in the literature through the years, though AV also has been referred to by several other names, including acne vermoulante, atrophoderma reticulatum symmetricum faciei, folliculitis ulerythematosa reticulata, folliculitis atrophicans reticulata, atrophoderma reticulatum, and honeycomb atrophy.[6]

Atrophoderma vermiculatum typically presents in childhood with erythema and follicular keratotic papules that slowly progress to the characteristic reticular atrophy,[6,7] which has been described as worm-eaten, reticular, or honeycomb, and occurs on the cheeks, preauricular area, and forehead. More rarely, the atrophy may extend to the upper lip; helices; ear lobes; and, in some cases, the limbs. The degree of inflammation, the presence of milia, and the extent of follicular plugs are variable.[6] As seen in our patient, AV tends to be symmetric, but unilateral involvement has been reported.[8,9] Although AV typically presents before puberty, some patients have developed AV in late adolescence or early adulthood,[6] as seen in our patient.

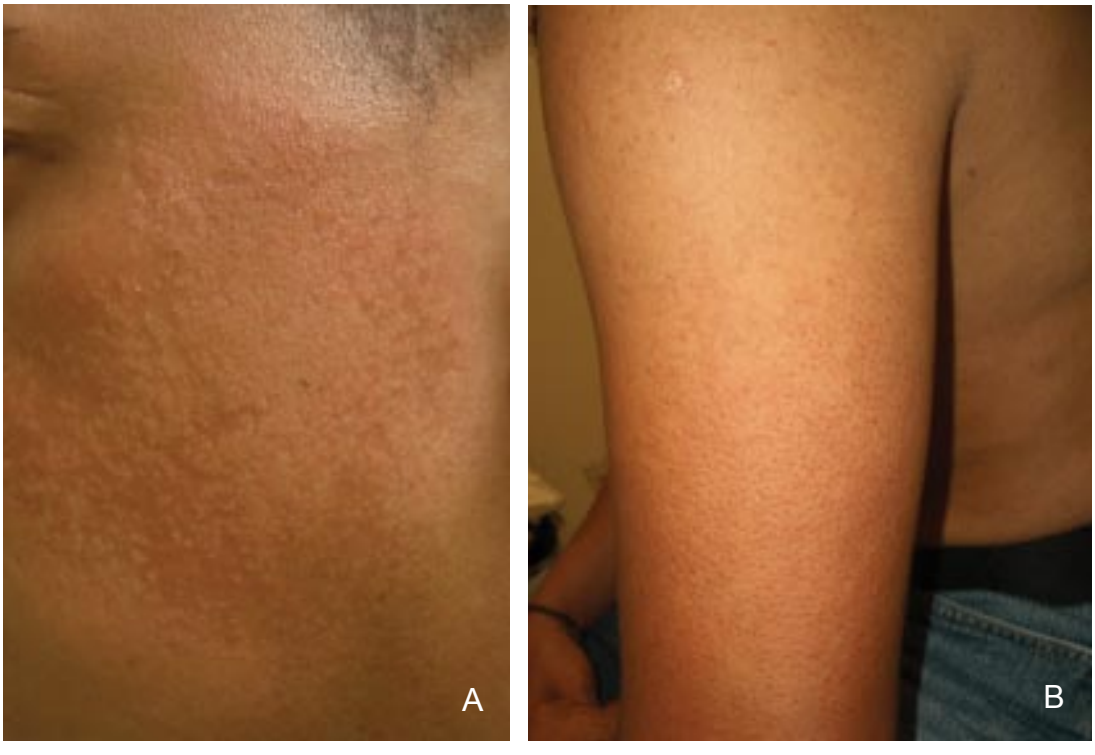

Figure 1. Honeycomb atrophy on the cheek (A). Extensive atrophic keratosis pilaris on the extensor surface of the left arm (B).

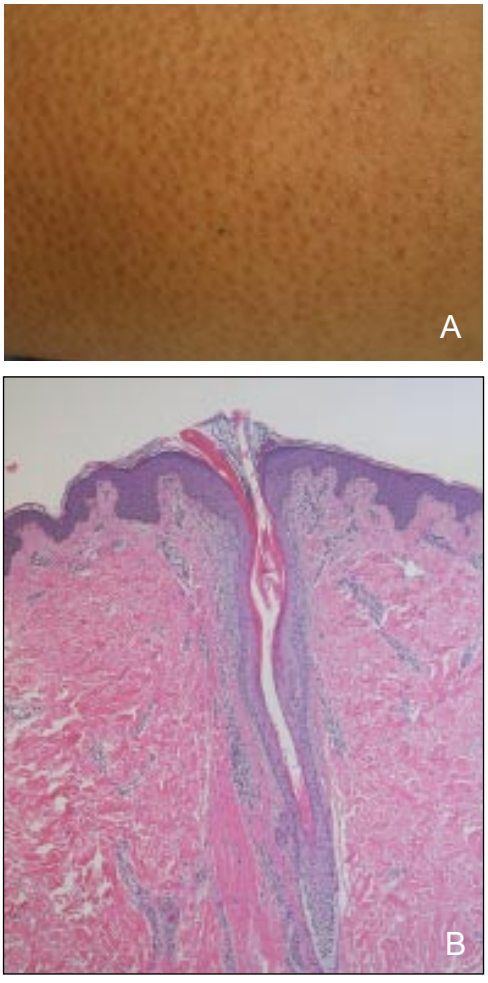

Figure 2. A punch biopsy specimen of an atrophic keratotic papule on the upper back (A) showed mild perifollicular fibrosis and chronic inflammation (B)(H&E, original magnification ×10).

Histologically, AV may display atrophic follicles with sclerosis of dermal collagen, loss of rete ridges, follicular plugs, and variable degrees of perifollicular inflammation.[6]

The pathogenesis of AV is unclear. The primary pathologic event seems to be the formation of follicular plugs. Baden and Byers[10] suggest that the keratinocytes in affected individuals mediate release of inflammatory cytokines in response to the plugs. This inflammation then leads to fibrosis and atrophy.[2,10]

The occurrence of AV typically is sporadic, though autosomal dominant inheritance has been reported.[1,6] Atrophoderma vermiculatum usually is an isolated skin finding, but it also has been reported in association with dioxin-induced chloracne,[11] steatocystoma multiplex,[2] epidermal cysts, leukokeratosis oris, leukonychia,[6] congenital heart block, atrial septal defect with associated Eisenmenger complex, neurofibromatosis, and Down syndrome.[2,6] Atrophoderma vermiculatum also is a key feature of Rombo syndrome, an autosomal dominant disorder characterized by AV, milia, hypotrichosis, trichoepitheliomas, basal cell carcinomas, and peripheral vasodilation with cyanosis.[12]

Atrophoderma vermiculatum differs from the other types of KPA because it lacks alopecia and typically has a later age of onset, usually between 5 and 12 years of age versus infancy for KPAF and KFSD. Keratosis pilaris atrophicans faciei typically presents in the first few months of life in boys with blond hair, with erythema and follicular keratotic papules on the lateral third of the eyebrows. Disease progresses to atrophy and alopecia, which may extend to the cheeks and forehead. Sometimes referred to as ulerythema ophryogenes, KPAF has been associated with Noonan syndrome,[13] woolly hair,[14] cardiofaciocutaneous syndrome,[15] Rubinstein-Taybi syndrome,[16] and other congenital anomalies.[17] The molecular defect in KPAF is unknown, but it may be related to a gene defect on chromosome arm 18p.[18,19]

Keratosis follicularis spinulosa decalvans also presents in infancy, but the erythema and follicular keratotic papules center on the malar area more than the eyebrows. Occasionally referred to as ichthyosis follicularis, KFSD is characterized by patchy scarring alopecia of the eyebrows and scalp. Thinning of axillary and pubic hair as well as palmoplantar keratoderma (especially over the heels) and ocular abnormalities may be seen in KFSD. Other associations include aminoaciduria and Noonan syndrome.[2,20]

All types of KPA tend to remit at puberty with the exception of the autosomal dominant pustular variant of KFSD, termed *folliculitis spinulosa decalvans*.[2] Treatment of KPA in general has been somewhat disappointing. Therapies for AV have included topical keratolytics, topical and intralesional corticosteroids, topical and oral retinoids, oral antibiotics, and UV therapy.[2,10,21] With the possible exception of prolonged, high-dose isotretinoin,[21] none of these therapies have been particularly helpful. Rarely, the lesions of AV may spontaneously regress[10]; more commonly, however, patients are left with permanent atrophic scarring after the inflammatory process remits. Dermabrasion, laser therapy (both carbon dioxide and pulsed dye lasers), and collagen implantation have been helpful in improving the appearance of atrophic scarring.[2,22]

Conclusion

Our patient presented with AV in late adolescence, which is unusual. More commonly, patients with AV present in childhood and experience resolution of inflammation at puberty. Although many associations have been reported with AV (notably autosomal

dominant inheritance, cardiac defects, and Rombo syndrome), the occurrence was sporadic in our patient, with no associated defects. Treatment of all types of KPA is challenging and often unrewarding. Our patient elected not to undergo any treatment for his AV.

REFERENCES

1. Rand R, Baden HP. Keratosis follicularis spinulosa decalvans—report of two cases and literature review. *Arch Dermatol*. 1983;199:22-26.
2. Callaway SR, Lesher JL. Keratosis pilaris atrophicans: case series and review. *Pediatr Dermatol*. 2004;21:14-17.
3. Davenport DD. Ulerythema ophryogenes; review and report of a case. discussion of relationship to certain other skin disorders and association with internal abnormalities. *Arch Dermatol*. 1964;89:74-80.
4. Unna PG. Ulerythema acneiforme. In: Unna PG, ed. *Histopathology of Diseases of the Skin*. New York, NY: Macmillan; 1896:1084.
5. Darier J. Atrophodermie vermiculée des joues avec keratoses folliculaires. *Bull Soc Fr Dermatol Syphiligr*. 1920;27:345-350.
6. Frosh PJ, Brumage MR, Schuster-Pavlovic C, et al. Atrophoderma vermiculatum: case reports and review. *J Am Acad Dermatol*. 1998;18:538-542.
7. MacKee GM, Parounagian MB. Folliculitis ulerythematosa reticulata. *J Cutan Dis*. 1918;36:337.
8. Hsu S, Nikko A. Unilateral atrophic skin lesion with features of atrophoderma vermiculatum: a variant of the epidermal nevus syndrome? *J Am Acad Dermatol*. 2000;43:310-312.
9. Nico MM, Valente NY, Sotto MN. Folliculitis ulerythematosa reticulata (atrophoderma vermiculata): early detection of a case with unilateral lesions. *Pediatr Dermatol*. 1998;15:285-286.
10. Baden HP, Byers HR. Clinical findings, cutaneous pathology, and response to therapy in 21 patients with keratosis pilaris atrophicans. *Arch Dermatol*. 1994;130:469-475.
11. Gianotti F. Chloracne due to tetrachloro-2,3,7,8,-dibenzo-p-dioxine in children [in French]. *Ann Dermatol Venereol*. 1977;104:825-829.
12. Michaëlsson G, Olsson E, Westermark P. The Rombo syndrome: a familiar disorder with vermiculate atrophoderma, milia, hypotrichosis, trichoepitheliomas, basal cell carcinomas and peripheral vasodilation with cyanosis. *Acta Derm Venereol*. 1981;61:497-503.
13. Snell JA, Mallory SB. Ulerythema ophryogenes in Noonan syndrome. *Pediatr Dermatol*. 1990;7:77-78.
14. McHenry PM, Nevin NC, Bingham EA. The association of keratosis pilaris atrophicans with hereditary woolly hair. *Pediatr Dermatol*. 1990;7:202-204.
15. Schepis C, Greco D, Romano C. Cardiofaciocutaneous (CFC) syndrome. *Australas J Dermatol*. 1999;40:111-113.
16. Gómez Centeno P, Rosón E, Peteiro C, et al. Rubinstein-Taybi syndrome and ulerythema ophryogenes in a 9-year-old boy. *Pediatr Dermatol*. 1999;16:134-136.
17. Burnett JW, Schwartz MF, Berberian BJ. Ulerythema ophryogenes with multiple congenital anomalies. *J Am Acad Dermatol*. 1988;18:437-440.
18. Zouboulis CC, Stratakis CA, Rinck G, et al. Ulerythema ophryogenes and keratosis pilaris in a child with monosomy 18p. *Pediatr Dermatol*. 1994;11:172-175.
19. Zouboulis CC, Stratakis CA, Gollnick HPM, et al. Keratosis pilaris/ulerythema ophryogenes and 18p deletion: is it possible that the *LAMA1* gene is involved? *J Med Genet*. 2001;38:127-128.
20. Oranje AP, van Osch LD, Oosterwijk JC. Keratosis pilaris atrophicans: one heterogeneous disease or a symptom in different clinical entities? *Arch Dermatol*. 1994;130:500-502.
21. Weightman W. A case of atrophoderma vermiculatum responding to isotretinoin. *Clin Exp Dermatol*. 1998;23:89-91.
22. Handrick C, Alster TS. Laser treatment of atrophoderma vermiculata. *J Am Acad Dermatol*. 2001;44:693-695.