

Case report

Atrophoderma vermiculata: A rare disfiguring condition

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Abstract Atrophoderma vermiculata is a rare genodermatosis with usual onset in childhood, characterized by a “honey-combed” reticular atrophy of the cheeks. The course is generally slow, with progressive worsening. We report a young male with multiple, ugly looking, worm eaten scars on his cheeks who partially responded to chemical peeling with trichloroacetic acid 35% solution and topical application of 0.05% tretinoin cream. A brief review of the disorder is also given.

Key words

Atrophoderma vermiculata, keratosis pilaris atrophicans, ulerythema ophryogenes

Introduction

Atrophoderma vermiculata is a rare genodermatosis that presents as an inflammatory follicular atrophy. The morphologic hallmark is a “worm-eaten” or “honey-combed” reticular atrophy of the skin typically localized at the cheeks, preauricular regions, and temples.^{1,2} Rarely, patients may experience the characteristic lesions on the extensor surfaces of the arms and legs. In addition to the follicular atrophic scars, generalized facial erythema, sparse open and closed comedones, and milia can be found. Associated cutaneous and visceral abnormalities can occur. The underlying pathologic defect appears to be an abnormal keratinization of the pilosebaceous unit.³ Possible genetic defect is supposed to be the deletion of chromosome 18p.⁴ This condition generally has its onset in childhood, although some cases arising during puberty or adulthood have been seen.⁵

Histologically, in the early phase, pilosebaceous follicles filled with keratotic plugs and a mild perifollicular inflammatory infiltrate are observed. Cystic dilatation of the hair follicles may be evident on the cheeks. Later, atrophy of both hair follicles and sebaceous glands, as well as dermal fibrosis, may appear. The course is generally one of slow progressive worsening; however, instances of spontaneous regression have also been reported. Primarily a cosmetic problem, therapy for this condition is aimed at reassurance, genetic counseling, and dermabrasion where appropriate. Other options include cryotherapy, ultraviolet light radiation, and several topical medications. Carbon dioxide (CO₂) and 585nm pulsed dye lasers have also recently been used with success.⁶⁻⁸ A case of successful induction of remission in the inflammatory component of the disease has also been reported in the literature, following a prolonged course of isotretinoin.⁹

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Case history

A 25-year-old male reported with history of multiple disfiguring scars over his right cheek since childhood. During last one year, he noticed two painless nodular swellings and a few small scars over left cheek. There was no history of any other preceding or concomitant cutaneous disorders like acne, folliculitis, small pox or chicken pox. None of his family members had similar lesions. On dermatological examination he was found to have multiple 1-3 mm sized pitted and ice picks scars with ridges producing a reticulated honeycomb appearance on right cheek (**Figure 1**). Sparse open and closed comedones were present but inflammatory acne lesions, milia and facial erythema were notably absent. Two nodulocystic swellings along with a few small scars and closed comedones were also seen over left cheek (**Figure 2**). Due to peculiar worm eaten appearance, typical site of involvement and characteristic morphology of the lesions he was clinically diagnosed as a case of atrophoderma vermiculatum. Nodular swellings on right side were excised and on histology these turned out to be sebaceous cysts. He was explained the nature of his disease and was advised fortnightly chemical peeling with trichloroacetic acid 35% solution and daily topical application of 0.05% tretinoin cream. He showed some response to this treatment regimen but then was lost to follow up.

Discussion

Atrophoderma vermiculatum is a condition that also has been called atrophoderma reticulatum, acne vermoulante, folliculitis ulerythematoso folliculitis reticulata, and honeycomb



Figure 1 Multiple pitted and ice picks scars along with few scattered comedones



Figure 2 Left side of the face showing two sebaceous cysts few scars and closed comedones

atrophy. It has been classified as one of the four conditions that present with keratosis pilaris atrophicans (KPA), which is characterized by follicular hyperkeratosis, inflammation, and scars. The other entities that show KPA are keratosis pilaris atrophicans faciei, ulerythema ophryogenes and folliculitis spinulosa decalvans.¹⁰ Some authors believe these entities are different presentations of the same condition. Atrophoderma vermiculata is characterized by erythema and reticulate atrophic scarring of the face and may also be called as a follicular syndrome with inflammation and atrophy. The other variants of this syndrome can be distinguished from atrophoderma vermiculata by location, degree of inflammation, mode of inheritance, and histologic pattern. Each of these diseases

usually manifests in infancy or childhood and runs a chronic course with only rare spontaneous regression seen. Their common pathologic features are follicular dilation, hyperkeratosis, and ultimate follicular destruction. Patients with atrophoderma vermiculata are often psychologically affected by their obvious facial lesions and thus are compelled to seek cosmetically effective treatment. Because there are no curative therapeutic modalities available, palliative treatment has been attempted with topical steroids, tretinoin creams, and systemic retinoids. More aggressive treatment modalities include cryotherapy, dermabrasion, and laser therapy. The patient we described here was a typical case of atrophoderma vermiculatum who was psychologically much disturbed due to obvious disfigurement of his face. Because of non-availability of suitable laser in the town and financial constraint of the patient, he was offered medium depth chemical peeling and topical tretinoin, to which, he partially responded. We think appropriate laser therapy or oral isotretinoin would have given him better results.

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