A Case of Bilateral Scarring of the Cheeks in a Child

An 11-year-old boy presented with scars on both cheeks. He gave a history of waxing and waning pruritic papules and papulovesicles on the face as well as on the dorsa of the hands since 3–4 years. There was no history of photosensitivity, other systemic complaints or family history of a similar condition.

On examination, pock-like and vermiculate scars were present on both cheeks, extending from the nasolabial folds to the pre-auricular region [Figure 1]. There were no comedones or milia.

A punch biopsy from the facial lesion revealed follicular plugging and dermal atrophy [Figure 2]. Some of the hair follicles were atrophic and accompanied by small, poorly developed sebaceous units [Figure 3]. The dermis showed a mild perivascular lymphocytic infiltrate. There were no vascular changes or dermal deposits.

WHAT IS YOUR DIAGNOSIS?

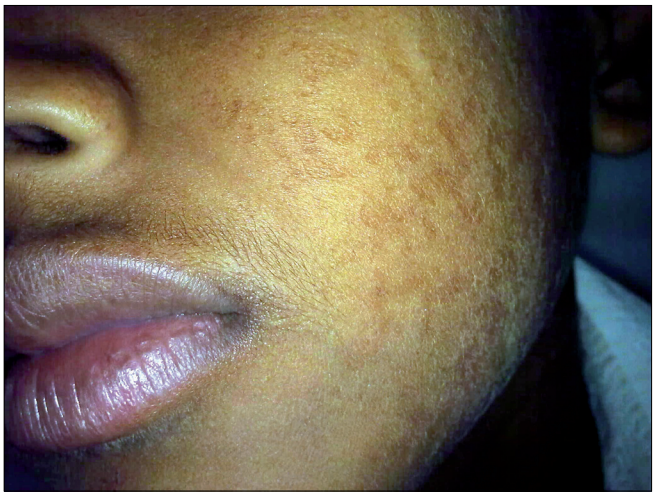

Figure 1: Pock-like, vermiculate scars on the cheek

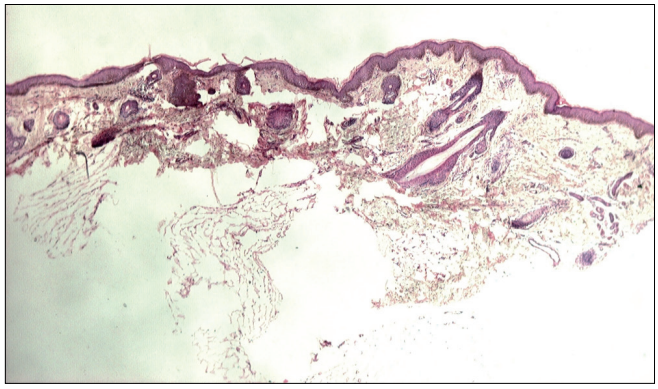

Figure 2: Thinned-out dermis (H and E, ×100)

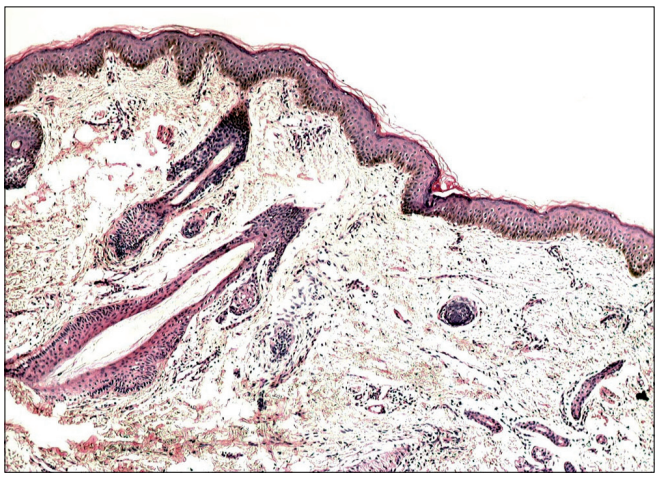

Figure 3: Atrophic hair follicles accompanied by ill-formed sebaceous units (H and E, ×400)

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YK Inchara, T Rajalakshmi, Mary Augustine[1]

Departments of Pathology and [1]Dermatology, St. John's Medical College & Hospital, Johnnagar, Bangalore, Karnataka, India

Address for correspondence:

Dr. Y. K. Inchara, Department of Pathology, St. John's Medical College & Hospital, Johnnagar, Bangalore - 560 034, India. E-mail: incharayk@yahoo.com

DIAGNOSIS

Atrophoderma vermiculata (AV)

DISCUSSION

AV is a rare, disfiguring dermatologic condition characterized by reticular atrophy of the cheeks. This is a consequence of abnormal keratinisation of the pilosebaceous unit.

AV (Folliculitis ulerythematosa reticulata) is one of the three related disorders categorised under Keratosis pilaris atrophicans (KPA), in which keratosis pilaris is associated with mild perifollicular inflammation and subsequent atrophy.[1] The other entities in this group are Keratosis pilaris atrophicans faciei and Keratosis follicularis spinulosa decalvans. Differences in location, degree of atrophy and mode of inheritance distinguish the three entities, which are detailed in Table 1.[2]

Table 1: Comparison of variants of Keratosis pilaris atrophicans

Variant	Age at onset	Site of involvement	Mode of inheritance
Keratosis pilaris atrophicans faciei	Soon after birth	Eyebrows, forehead, cheeks	Autosomal dominant
Keratosis follicularis spinulosa decalvans	Infancy	Cheeks, scarring alopecia	? Sex-linked
Atrophoderma vermiculata	Late childhood	Pre-auricular region, cheeks	Autosomal dominant

A typical lesion develops in late childhood, presents as "worm-eaten" or "honey combed" atrophy of the skin and usually affects the pre-auricular region and cheeks on both sides. Rarely, the lesion may be unilateral in distribution.[1,3] Erythema, comedones and follicular plugs may be present. Histologic findings are as described in the case. In addition, there may be comedones, milia and variable dermal fibrosis.

Less commonly, it may be part of a syndrome such as the Rombo syndrome, where there is a propensity to develop basal cell carcinoma.[4]

Clinically, the lesions mimic porphyria (distinguished by the presence of photosensitivity) or lipoid proteinosis and need a biopsy for confirmation.[2] Porphyria (the erythropoietic protoporphyria type) is typified by thickened, stiff vascular basement membranes owing to the deposition of porphyrins.[5] Lipoid proteinosis shows massive perivascular and peri-eccrine eosinophilic deposits that are periodic acid schiff (PAS) positive and diastase resistant. There is atrophy of sweat glands with increasing deposition.[2]

AV is difficult to treat and results are often disappointing. The suggested treatment options include topical application of keratolytics, steroids and ultraviolet irradiation. Dermabrasion and collagen implants can also be used.[6] There have been reports of cases treated with carbon dioxide and 585-nm pulsed dye lasers (PDL), with encouraging results.[7] According to one study, PDL was found to be effective in treating the erythema associated with KPA, but did not give significant improvement in associated skin roughness.[8] Systemic use of isotretinoin with beneficial effects has been reported in one case; however, the possible adverse effects of such treatment should be borne in mind.[6]

This is a rare lesion, and the Indian literature is limited to sporadic case reports.[9]

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