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ATROPHODERMA VERMICULARIS

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Three siblings of a family developed atrophoderma vermicularis, presenting as itchy, keratotic follicular papules all over the body mainly on the extensor aspect of the extremities and multiple atrophic pits involving the face and atrophic scars on other areas of the body.

Key words : Folliculitis ulerythematosus reticulata, Keratosis pilaris atrophicans.

There is a group of rare inflammatory follicular atrophic conditions, closely related to keratosis pilaris.[1] Minor differences in the regional distribution and morphological changes have been used to differentiate various syndromes viz keratosis pilaris decalvans, keratosis pilaris atrophicans faciei and atrophoderma vermicularis.[2] We saw a child and his younger brother and sister affected by such a disorder.

Case Report

Twelve-year old male child had itchy, keratotic, follicular papules all over the body, multiple atrophic pits involving the face and atrophic scars on other areas of the body for the last 9 years. The lesions were worse during winter and would heal spontaneously in a few weeks time leaving behind atrophic pits. His sister aged 8 years and brother aged 5 years had similar lesions beginning at the age of about 3 years but the course of the disease till date was milder. There was no past history of exanthematous fever. None of the other family members had night blindness, atopy or similar lesions.

Examination revealed a moderately built and well-nourished boy with multiple, superficial, irregular, atrophic pits, 5 to 10 mm in length, 1 to 2 mm in width and about 1 mm in depth predominantly on the central part of the face involving the forehead, nose and cheeks (Fig. 1), the distribution being almost symmetrical. The affected areas felt slightly rougher and firmer than the uninvolved skin. There were multiple keratotic follicular papules distributed all over the body mainly on the extensor aspects of the extremities. Some of the lesions revealed tendency to grouping and coalescence while most of these were isolated. Some papules were linearly arranged particularly on the anterior aspect of the right leg. A few 0.5 to 1.5 cm size, atrophic scars were present on the nape of the neck and dorsum of the left hand. There was hyperkeratosis of the palms and soles. The scalp, eyebrows, nails and mucous membranes were normal. Examination of the eyes as well as other systems was unremarkable.

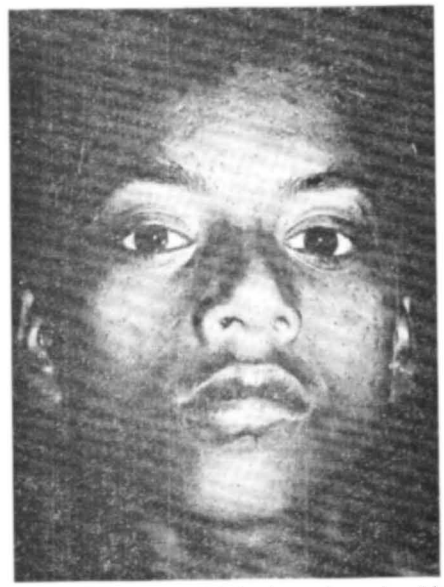

Fig. 1. Multiple, superficial, irregular, atrophic pits involving the face.

There was no evidence of keratosis pilaris in the parents. Routine investigations like blood

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Fig. 2. Thinned out epidermis with fibrosis and degeneration of the connective tissue (H and E ×100).

cell count, hemoglobin, sedimentation rate, liver function tests, urinalysis and stools examination were normal. Serum vitamin A and carotene levels were not done. Histopathological examination revealed thin to normal epidermis with atrophic or absent sebaceous glands (Fig. 2). There was fibrosis and degeneration of the connective tissue, with a chronic inflammatory cellular infiltrate. At places, a few hyperkeratotic, acanthotic and tortuous hair follicles were also seen.

Comments

Atrophoderma vermiculatum (syn folliculitis ulerythematosa reticulata) is a rare genodermatosis with autosomal transmission in most cases.[1] Since Unna in 1896 described for the first time this condition under the name ulerythema acneiforme, it has been reported under various names viz acne vermoulante, atrophoderma reticulata symmetrica faciei, folliculitis ulerythematosa reticulata, folliculitis atrophicans reticulata, atrophoderma vermicularis, atrophoderma vermiculatum, atrophoderma reticulata[3] and honeycomb atrophy.[4] This condition is characterized by the following clinical features : (1) appearance of the eruption in childhood with slow evolution, (2) symmetrical distribution of the lesions involving both cheeks, (3) erythema that may disappear later, (4) comedones or horny plugs that may disappear later, (5) reticulated atrophy that becomes less marked in adult life, and (6) absence of papules, pustules, crusts or hyperpigmentation.[5]

The condition could be confused with scarring due to acne vulgaris if the patients are first examined in adolescence, but its earlier onset is a distinguishing feature whereas in acne the scars are far deeper.

Familial occurrence with apparently dominant transmission, classical morphology of the lesions, almost symmetrical reticulate atrophy, milia and presence of keratosis pilaris along with compatible histopathological features as seen in our patient go in favour of the diagnosis of atrophoderma vermiculatum. The early onset of the disease, tendency of keratosis pilaris for Koebner phenomenon, bigger atrophic scars on areas other than the face were the unusual features in our cases.

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