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FAMILIAL GIANT REACTIVE PERFORATING COLLAGENOSIS

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A 21-year-old man presented with slowly growing, multiple, recurrent, discrete, asymmetrical hyperkeratotic and hyperpigmented papules of size varying between 10-15mm over face and upper limbs; usually after trauma. Histopathology confirmed the diagnosis of reactive perforating collagenosis. This case is reported because of unusual bigger size of the lesions.

Key Words : Reactive perforating collagenosis

Introduction

Reactive perforating collagenosis (RPC) is a form of transepidermal elimination in which altered collagen is extruded from the epidermis. The disease usually starts as papules in early childhood. The papules develop umbilications with keratinous plugs. We report a case of RPC with a positive family history. The size of umblicated papules was abnormally bigger, therefore we have named it giant RPC.

Case Report

A 21- year-old man presented with slowly growing, multiple, recurrent, discrete, asymmetric, hyperkeratotic and hyperpigmented papules of size varying between 10-15 mm, over the face and upper limbs. The eruption started at the age of 15 years. Initially a skin-coloured papule appeared on dorsum of right finger which soon developed central umbilication filled with a keratinous plug which was initially adherent but soon became loose and finally fell off. Involution of lesion occurred with hyperpigmentation and epidermal atrophy. Different stages of evolution and regression were seen at the same time, the lesions were showing Koębner's phenomenon. These lesions occurred at sites of acne, minor trauma because of shaving, and sometimes

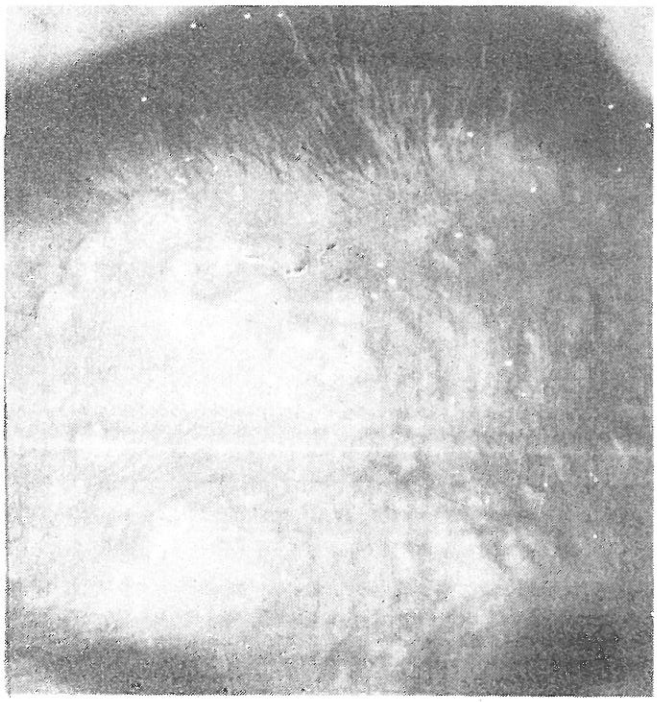

Fig.1. Multiple large hyperkeratotic lesions spontaneously.

Family history of such lesions was present in younger brother of 9 years who developed these lesions since early childhood. No history of diabetes, hepatic, renal or other systemic disorders was present.

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Routine investigations were normal. Biopsy of the initial non- umbilicated papule showed an area of necrobiotic, deeply basophilic collagen in the papillary dermis. The old umbilicated lesion in which the plug was eliminated showed a central area of depression, an accumulation of parakeratotic keratin, basophilic collagen, and numerous pyknotic nuclei of inflammatory cells. The epidermis at the base of the plug was atrophic and showed perforation through which basophilic bundles of collagen extending upwards were extruded from the underlying dermis. This was confirmed by staining with von Gieson method. The periphery of the cup-shaped lesion showed epidermal hyperplasia. The elastic fibres were normal and seen only in the dermis.

Comments

According to Mehregan et al[1] RPC is an abnormal response to superficial trauma in which collagen causes irritation and perforation of the epidermis with transepidermal elimination.

Formation of giant lesions in RPC may be associated with increasing age as the younger brother of our patient had smaller lesions. Familial occurrence of RPC has been recorded only in some cases. [2-5]

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