

REACTIVE PERFORATING COLLAGENOSIS

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Two brothers aged 9 and 7 years had asymptomatic, umbilicated papules on their extremities, buttocks and hair-line, since infancy. The lesions would heal spontaneously with atrophic scarring, but new lesions would keep appearing especially after trauma. Koebner phenomenon was observed on several body sites. Histopathology including van-Geison's stain confirmed the lesions to be reactive perforating collagenosis. There was no evidence of diabetes, renal or hepatic disease.

Key words : Reactive perforating collagenosis.

Reactive perforating collagenosis (RPC) has been recognised as a distinct clinical and histopathological condition.¹ The disease usually commences in early childhood as papular lesions which eventually become umbilicated with a keratinous plug. The lesions involute spontaneously in about 5 to 10 weeks, while new lesions may appear at the same or other sites. A distinct histopathological pattern characterises this condition as the altered collagen affected by an abnormal reaction of skin to superficial trauma, is extruded through the epidermis by transepithelial elimination.^{1,2} There are only a few cases reported in the literature.

Case Report

A male child aged 9 years was seen with multiple, recurrent, asymmetric, hyperkeratotic and hyperpigmented papules on areas of scratching and trauma since the age of 9 months. Eruptions appeared almost all over his body but were most prevalent on the face, arms, forearms, hands, fingers, buttocks, thighs, legs and scalp hairline. Lesions were skin-coloured to start with. Each lesion increased in size and developed a central umbilication filled with a keratinous plug in a period of 5-10 weeks. The keratinous plugs were adherent initially, later got loose and

finally fell off. By the end of ten weeks, the size of the lesions reached a maximum of 10-15 mm. Regression was associated with minimal atrophy at places and hyper- or hypo-pigmentation. The patient was never free of the lesions all through the course of the disease. There was no secondary infection in the lesions at any time and he was in excellent health, he had no history of diabetes, renal, hepatic or any other systemic disease.

The younger brother, aged 7 years too had similar lesions since his infancy, but was normal systemically. Other siblings (aged 11 and 13 years) had no lesions. Their parents were non-consanguineous.

The lesions showed evidence of Koebner's phenomenon especially on the trunk, extremities and hands (Fig. 1). Routine hematological examination, urine and stools were normal. Mantoux and VDRL tests were negative. Skiagram chest showed normal pattern. Biopsy from a lesion showed a conspicuous cup-shaped depression in the epidermis containing an accumulation of parakeratotic keratin, scant collagen material and pyknotic nuclei of inflammatory cells. The epidermis at the base showed marked atrophy and was interrupted at places. The collagen bundles closely juxtaposed to the epidermis were disposed vertically towards the epidermis. At places, these fibres extruded through the epidermis. This was confirmed by van-Geison's stain which stained the collagen

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Fig. 1. Lesions of RPC showing characteristic central umbilication filled with keratinous plug and Koebner's phenomenon.

red. Adjacent to the cup-shaped depression, the epidermis was hyperplastic. The fresh non-umbilicated lesion showed a prominent area of deeply basophilic collagen in the papillary dermis.

Comments

The genetic defect in this disease is ill-understood, it is inherited as an autosomal recessive character.⁵ Familial incidence has been recorded only in some case reports.^{2,6-8} Mehregan et al¹ considered that this is an abnormal response

to superficial trauma in which collagen causes irritation and perforation of the epidermis with transepidermal elimination.

For diagnosis, histopathological confirmation of invasion of the epidermis by collagen is essential, for which histochemical stains such as van-Geison's stain are essential.⁴

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