REACTIVE PERFORATING COLLAGENOSIS
(Case Reports)

C. GANGADHARAN, CYNTHIA PAPALI
P. U. ASOKAN AND K. ROBINS

Summary

The occurrence of reactive perforating Collagenosis in three members of one family is presented. A genetic abnormality of the collagen in the upper dermis is suggested as the probable cause for the disorder.

Mehregan et al in 1967[1] first described the entity reactive perforating collagenosis in a girl aged 6½ years, who had the disease from the age of nine months. It was characterised by the appearance of pin head sized papules which enlarged in size to about 4 mm to 6 mm and developed a keratinous plug in the centre. After reaching this stage, each lesion underwent spontaneous regression. The lesions were profuse over areas of trauma like the dorsa of the hands and feet, knees and elbows. Koebner phenomenon was also evident. Histologically the lesions exhibited an unique phenomenon characterised by necrobiosis of the collagen of the upper dermis which extruded through the epidermis.

The present paper is the report of a family in which three members are affected by this disorder.

Report of Cases

A 23 year old woman presented with multiple skin coloured papules varying in size from 2 mm to 5 mm distributed uniformly on the trunk, limbs, face and scalp. Each papule showed a central keratinous plug which was prominently seen in the bigger ones. On removal of the keratinous plug, a cup shaped depression was evident. Rounded varioliform scars of sizes 4 mm to 5 mm, left behind by healed lesions, were seen all over the body including the face. (Fig. 1 page No. 331) The patient was well till the age of 10 years, when the lesions first started appearing. They were in the form of skin coloured papules initially small in size which enlarged in the course of about 4 weeks, along with the formation of a central keratinous plug. These papules within another 3–4 weeks time regressed spontaneously with scar formation. The healing was hastened if the keratinous plug was removed. As earlier lesions subsided new ones appeared in crops. In areas of injury also new lesions were found to appear.

In the family, parents were free of the disease and there was no history of consanguinity. Out of the six siblings two brothers aged 32 years and 15 years were also affected by this disorder. They started manifesting the disease from the ages of 10 years and 6 years respectively. The clinical features in these cases were same as in the first case.

Department of Dermatology and Venereology
Medical College Hospital
Trivandrum 695011 S. India
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Laboratory investigations namely haematologic examination, urine analysis, blood VDRL, blood urea, serum proteins and x-ray chest were normal in all the three cases.

Histopathological findings :

From each patient one fully developed lesion was biopsied and serial sections studied after staining with haematoxylin eosin stain and Van Gieson's stain for collagen. (Fig. 2 page No. 331) The epidermis showed a cup shaped depression which was unrelated to hair follicle. This depression was filled by an eosinophilic parakeratotic plug which was intermingled with a basophilic degenerated mass which contained fragmented nuclei. The base of the depression showed thinning and rupture at one place where there was direct continuity between the basophilic material in the depression and the collagen in the upper dermis. The collagen in this region also showed a basophilic degeneration with inflammatory cells and fragmented nuclei. At the sides of the depression the epidermis was acanthotic. These findings seen with haematoxylin eosin stain was confirmed by the Van Gieson's stain. The keratinous material in the depression appeared yellow by this stain and the degenerated mass somewhat dark pink in colour. The collagen of the upper dermis also showed a dark pink colour with nuclei and nuclear fragments.

Discussion

After the original description of reactive perforating collagenosis by Mehregan et al[1] only few reports have appeared in the literature on this subject,[2,3,4]. Mehregan et al put forward the view that reactive perforating collagenosis is an abnormal response of the collagen to superficial trauma and this view has been supported by most other workers. [2,3,4]. The points given in support of this view are the occurrence of the lesions in areas exposed to trauma and the presence of Koebner phenomenon.

The three cases presented in this paper differed from the original case of Mehregan et al[1] in that the lesions were generalised in distribution. This distribution and the discrete nature of each lesion suggest that some aetiological mechanism other than trauma is initiating the disease. The role of trauma as evidenced by Koebner phenomenon remains unexplained as in psoriasis. The occurrence of the disease in three members of one family points to the fact that reactive perforating collagenosis may be a genetic disorder. It is possible that the collagen in the upper dermis is genetically abnormal and this abnormal collagen undergoes degeneration and is extruded through the epidermis.

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