Reactive perforating collagenosis in two siblings

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DESCRIPTION

A 5-year-old male child presented with multiple, asymptomatic, raised lesions over dorsum of bilateral hands, feet and face since the age of 1 year. The lesions used to aggravate in winters and heal with superficial scarring. His elder sister (7-years-old) also had a history of similar lesions since the age of 3 years. There was no history suggestive of systemic involvement. Their parents were concerned regarding the superficial scarring on the face. On examination, multiple, discrete, papular lesions with central adherent keratin plug, varying in size from 2 to 5 mm, were present over face, bilateral forearms and legs predominantly distributed over the elbow and knee in both siblings (figure 1A–D). Multiple atrophic scars were also visible over face. Few of the lesions were arranged in a linear distribution. Histopathological examination from a papule over forearm showed a shallow invagination of the papillary dermis containing a mixture of basophilic

material and degenerated collagen bundles. Adjacent epidermis showed moderate acanthosis. Haematological investigations including complete blood counts, liver and renal profile and serum blood glucose levels were within normal limits. The diagnosis of reactive perforating collagenosis (RPC) was thus established on the basis of clinical features aided by the histopathological findings.

RPC is a distinct transepithelial elimination (TEE) disorder characterised by umblicated skin coloured papules with central crust. RPC is considered to be a genetic disorder with no associations and presents at an early stage. Superficial trauma causes alteration of collagen in papillary dermis leading to its degeneration followed by perforation of epidermis with transepidermal elimination of necrobiotic material.¹ The four classical TEE disorders include RPC, elastosis perforans serpiginosa, perforating folliculitis and Kyrle's diseases. There has been an increasing number of reports of

Figure 1 (A, B) Multiple keratotic papules over chin and dorsum of left hand of the male child (arrows). Superficial scarring is present over the bilateral cheeks. (C, D) Similar keratotic papules present over dorsum of right hand in elder sister (arrow). Superficial scarring is present over forehead.

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perforating dermatoses in association with systemic diseases like diabetes mellitus, chronic renal failure, malignancies and liver disorders and hence labelled as acquired perforating collagenosis. Diagnosis is established by the classical morphology of papular lesions with central umbilication aided by characteristic histopathology.² Topical retinoids, corticosteroids, oral isotretinoin, methotrexate and Psoralens and Ultraviolet A phototherapy (PUVA) therapy have been shown to be useful in the treatment.^{2 3}

Learning points

- ▶ Reactive perforating collagenosis is a genetically determined rare cutaneous perforating disorder characterised by elimination of altered collagen through the epidermis.
- ▶ The disease usually starts in childhood and is characterised by umbilicated skin coloured papules with a keratinous plug which leaves a residual scar on healing.
- ▶ Diagnosis is established by the clinical presentation and characteristic histopathology.
- ▶ Topical retinoids, corticosteroids, oral retinoids, methotrexate and PUVA therapy are useful in the treatment.

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