A 7-year-old, Indian boy, born out of consanguineous parentage, presented with multiple scars on his face for the past 2 years and a few raised pruritic skin eruptions over different parts of his body for the same duration. His grandfather reportedly had similar kind of lesions. On examination, we found multiple well-defined, shallow, atrophic scars over his face. Some of these scars were arranged in a linear fashion. In addition, multiple skin-colored, discrete keratotic papules with a central adherent keratinous plug were located over the face, dorsum of hands, lower legs, and feet [Figures 1 and 2]. The skin was otherwise normal in appearance. Examination of the nails and mucosae showed no abnormality. Systemic examination was unremarkable. Routine laboratory investigations were within normal limits. We did 4-mm lesional punch biopsies from two representative lesions and histopathological examination was done subsequently.

Question
What is the diagnosis?
Answer
Familial reactive perforating collagenosis (RPC).

Discussion
The histopathological examination of the biopsy specimen showed a cup-shaped depression of the epidermis which was filled with a plug comprising of keratin, collagen, and inflammatory cell debris. The epidermis formed the base and was thinned out and showed fine vertical slits through which the collagen fibers were being extruded [Figure 3]. The dermis showed perivascular infiltration of lymphomononuclear cells. Van Gieson and Masson trichrome stains further confirmed that the collagen fibers (Masson trichrome: blue; Van Gieson: red) were being extruded out of the vertical slits in the epidermis [Figure 4]. Based on the clinical features and histopathological findings, a diagnosis of familial RPC was made.

Reactive perforating collagenosis is a relatively rare, benign perforating disorder clinically characterized by spontaneously involuting, skin-colored or slightly erythematous papules or nodules with a central adherent scab-like plug.\(^\text{[1-3]}\) Removal of the plug exposes a crateriform lesion. It leaves some faint scars during regression. The lesions usually occur on the extensor surfaces of the extremities and in areas that are accessible to scratching. The Koebner phenomenon is often seen.\(^\text{[3]}\)

Reactive perforating collagenosis can be familial or acquired, the former being the infrequent variety. The familial type often presents at an early age in the absence of any systemic disease, contrary to its acquired counterpart.\(^\text{[1,2]}\) Acquired RPC (ARPC) is a disease of multifactorial etiology.\(^\text{[1-3]}\) It may be associated with diseases such as diabetes mellitus, chronic renal failure, hypothyroidism, lymphoma, hyperparathyroidism, pulmonary fibrosis, scabies, and herpes zoster infection.

The precise pathogenesis of RPC remains unknown. In genetically susceptible patients, superficial trauma may lead to necrosis of the collagen in the dermis, followed by transepidermal elimination. According to other researchers, collagen change may be secondary to release of proteolytic enzymes from neutrophils following trauma. Grossly, a biochemical change in collagen has been proposed leading to extrusion, similar to an autoimmune phenomenon.\(^\text{[1]}\) The other primary perforating disorders of the skin that are characterized by transepidermal elimination, namely, elastosis perforans serpiginosa, perforating folliculitis, and Kyrie disease should be considered in the differential diagnosis of RPC. These major perforating diseases probably represent spectrum\(^\text{[4]}\) of related disorders which we are likely to see with increasing frequency, particularly in diabetes and population undergoing dialysis.\(^\text{[3]}\) There are many other disorders characterized by papules or nodules with central keratotic plugs and crusts. The deep penetration of the keratotic plugs, the size and irregularity, the age of onset, and the distribution of lesions should form the basis of the diagnosis. It is also essential to examine and test for any underlying conditions such as diabetes, liver, and renal disease. Histologically, the perforating disorders are characterized by transepidermal elimination of altered dermal substances. In RPC, the extruded material is predominantly collagen. The histological differential diagnosis includes the other perforating disorders.

Elastosis perforans serpiginosa shows increased numbers of thickened elastic fibers filling the perforation and dermis.

Perforating folliculitis by definition involves the hair follicles with infundibular perforation. In Kyrie disease, there is greater epidermal proliferation and a more intense inflammatory infiltrate.\(^\text{[3]}\)

Treatment of RPC is often challenging. The patient should be strongly advised to avoid injury, scratching, and photo-exposure. Although a large number of treatment modalities have been reported, there is a dearth of controlled trials. RPC may improve after treating the coexisting systemic disease.\(^\text{[5]}\) Lesions are usually self-limiting without any treatment but often recur. Individual lesions are initially treated with topical keratolytic agents such as salicylic acid in a concentration of 5–7% in Vaseline, urea 10–15%,\(^\text{[3]}\) and tretinoin (0.01–0.1%).\(^\text{[1]}\) Phototherapy is another

![Figure 3: Lesional skin biopsy showing: (a) A cup-shaped depression plugged with necrotic inflammatory debris (H and E, ×40). (b) Basophilic necrotic inflammatory debris extruding through a cup-shaped depression in the epidermis (H and E, ×100)](image1)

![Figure 4: (a) Van Gieson stain shows elimination of collagen fibers (stained red) through a cup-shaped depression in the epidermis (×100). (b) Masson’s trichrome stain shows elimination of collagen fibers (stained blue) through the epidermis (×100)](image2)
important treatment option; patients with ARPC may be treated with ultraviolet B (UVB) or narrow band UVB therapy.[6] After detachment of the crusts, and possibly curettage, treatment continues with class 2–3 corticosteroids.[5]

Intralesional injection of triamcinolone crystal suspension is another treatment option.[5] For systemic treatment, corticosteroids[5] and retinoids[7] may be used. The results for allopurinol are inconsistent.[3] Interestingly, biopsy itself may be equally effective (“inverse Koebner’s phenomenon”).[4]

**Learning points**

- Reactive perforating collagenosis is a relatively rare, benign perforating disorder clinically characterized by spontaneously involuting, skin-colored or slightly erythematous papules or nodules with a central adherent scab-like plug.
- It can be familial or acquired, the former being the uncommon variety.
- The Koebner phenomenon is often found in such cases.
- Histopathology with special stains for collagen confirms the diagnosis.
- The condition may be associated with diseases such as diabetes mellitus, chronic renal failure, hypothyroidism, lymphoma, hyperparathyroidism, pulmonary fibrosis, scabies, and herpes zoster infection among others.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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