Acquired Reactive Perforating Collagenosis of Diabetes Mellitus and Chronic Renal Failure

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We report, herein, a case of reactive perforating collagenosis in a 52-year-old man with diabetes mellitus and chronic renal failure on hemodialysis. He had multiple, pinhead to pea sized, round or oval shaped, central umbilicated papules on the face, trunk, and extremities for 3 months duration. The papules developed after scratching due to pruritus. Histopathologic examination showed a cup-shaped depression of the epidermis filled with parakeratotic material, necrobiotic collagen and inflammatory cells. At the base of the depression, the epidermis showed interruption through which basophilic bundles of collagen extend in a vertical direction from the dermis.

The skin lesions improved after treatment with oral antihistaminics.

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Reactive perforating collagenosis (RPC), originally described by Mehregan et al. in 1967, is believed to be a peculiar cutaneous response to superficial trauma characterized by the elimination of degenerated collagen fibers through the epidermis.1-4 In the past, it was thought that the disease usually affected children and had a hereditary tendency.5-6 Within the past decade, an increasing number of cases of RPC have been described in adults, especially those suffering from diabetes mellitus and chronic renal failure.7,8

In Korea, several cases of RPC which developed in children were reported9,12 and one of them had a family history.11 However, we could not find any reports of RPC in adult with diabetes mellitus and chronic renal failure in the Korean literature.

We report herein a case of RPC in adult with diabetes mellitus and chronic renal failure who was on hemodialysis.

REPORT OF A CASE

A 52-year-old man visited our outpatient clinic in February, 1989, with multiple skin lesions. His general health was poor. He had 16-year history of diabetes mellitus and 4-year history of diabetic nephropathy, and had been placed on chronic maintenance hemodialysis 2 years ago. Three months prior to the first visit, he developed multiple, pinhead to pea sized, round or oval shaped papules on the face, trunk and extremities (Fig. 1). Some lesions were in linear distribution suggesting a Koebner phenomenon (Fig. 2). Early lesions were pinhead to rice sized, erythematous to skin colored papules. As the papules enlarged they developed a central depression filled with a keratinous plug. The keratinous plugs were adherent and their removal caused bleeding. The whole body showed hyperpigmentation due to chronic renal failure. Funduscopic examination of eyes showed hyper-
Fig. 1. Multiple, discrete, pinhead to pea sized, round or oval shaped papules with central hyperkeratotic plug on the back.

Fig. 2. Linear distribution of the lesions suggesting a Koebner phenomenon.

tensive and sclerotic changes.

Laboratory tests revealed the following values: blood urea nitrogen, 49 mg/dl; serum creatinin, 8.5 mg/dl; urinalysis, negative except protein level of more than 300 mg/dl and glucose level of 500 mg/dl; fasting blood sugar, 210 mg/dl.

Histopathologic examination of a central umbilicated papule on the back showed a cup-shaped depression of the epidermis filled with a mixture of parakeratotic material, necrobiotic collagens and inflammatory cells (Fig. 3). At the lower border of the depression, the epidermis was disrupted, through which necrobiotic collagen fibers extended.

Fig. 3. Cup-shaped depression of the epidermis filled with parakeratotic cells and basophilic debris and surrounding epidermal hyperplasia (Masson’s trichrome stain, ×40)

Fig. 4. Perforating collagen bundles extending through the disrupted base of the epidermal crater (H & E stain, ×200)

Fig. 5. Vertical strands of collagen (Masson’s trichrome stain, ×200).
into the keratinous plug from the dermis (Fig. 4). Masson’s trichrome stain confirmed the perforating strands to be collagen (Fig. 5). Verhoeff stain showed no changes in the appearance or quantity of elastic tissue and no elastic tissue was found in the keratinous plug.

Diagnosis was confirmed by clinical appearance and typical histologic features.

We treated him with oral administration of antihistaminics for pruritus. The majority of the skin lesions healed within six to eight weeks when the patient was prevented from scratching, although new lesions developed after excoriation or trauma.

**DISCUSSION**

RPC is a skin disorder characterized by extrusion of collagen fibers through the epidermis.1,4 This rare form of cutaneous perforating disease, first described by Mehregan et al.1 in 1967, appears clinically as recurrent, umbilicated, Koebnerizing papules that were resolved spontaneously in six to eight weeks.1,4,7,11,13 In 1970, Bovenmeyer14 induced lesions in a patient with RPC by scratching the patient with a No. 25 needle.

Recently, RPC was classified into two clinical variants, classic and acquired.2 The two types are identical in histologic features, but different in clinical features.2,3 The histologic pattern of the disease is characterized by hematoxylinophilic changes of the collagen fibers and transepidermal elimination of them.1,3,5,7,13,15 These features were demonstrated in the present case also and confirmed by Masson’s trichrome stain.

The classic type, which occurs during childhood and has a genetic predisposition (probably autosomal recessive),6 is very rare and not associated with pruritus.2 The acquired type occurs during adulthood and is associated with significant pruritus and severe systemic disease particularly diabetes mellitus and chronic renal failure.2,3 RPC in association with lymphoma was also reported recently.15 Trauma such as scratching probably plays a great role in development of the skin lesions.2,3,7,16 The trauma transforms collagen into foreign material. In the present case, the trauma was scratching secondary to pruritus and the skin lesions resolved when he was prevented from scratching. As he has been suffered from long standing diabetes mellitus and chronic renal failure, we considered that he was of the acquired type.

Perforating disorders such as RPC, perforating folliculitis (PF) and Kyrle’s disease (KD), have developed in patients with chronic renal failure, usually the result of diabetic nephropathy.3,4 There are striking similarities among them and they are characterized by keratotic papules and nodules on the trunk and/or extremities which showed perforation on biopsy.4 The presence or absence of Koebnerization alone appear to be an unreliable discriminator among the above perforating disorders, but Koebnerization is usually seen in RPC.4,7,14 Nonfollicular involvement would appear to favor RPC or possibly KD.4 The penetration of the epidermis by collagen support the diagnosis of RPC and absence of this feature is used to support the diagnosis of KD.3,4 PF and KD are closely related and KD may represent an exaggerated form of PF.4

Based upon the aforementioned differential points, we considered that the present case is RPC both clinically and histopathologically. However, the clinical and histological similarities among the perforating diseases associated with chronic renal failure appears to outweigh their differences. Patterson4 proposed that a more acceptable term was “acquired perforating disease”.

Many treatment modalities have been tried including antibiotics, vitamin A orally and intramuscularly, intralesional and topical steroids, and keratolytics. But most of them have been ineffective.7,9,13,14 In 1979 Cullen17 reported successful treatment of RPC with 0.1% tretinoin cream. But Kim et al.9 reported that topical application of 0.1% tretinoin acid solution and vitamin A and D were not effective. Cochran et al.7 reported two cases of RPC which showed successful clearing with UVB treatment. In the present case, only antihistaminics were used for pruritus and skin lesions were almost resolved in six to eight weeks. However, new lesions developed after excoriation or trauma. Therefore, we believed that the present case in a man with diabetes mellitus and chronic renal failure who was on hemodialysis was acquired RPC and the perforation was initiated by scratching.
REFERENCES


