Acquired Reactive Perforating Collagenosis : Report of a Case and Review of the Literature†

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Abstract

A case of acquired reactive perforating collagenosis is reported in a 57-year-old Thai woman, with a history of diabetes mellitus, chronic renal insufficiency needing hemodialysis, and dry gangrene of the right fourth toe. Physical examination revealed multiple scattered erythematous hyperkeratotic nodules and plaques and some showed ulceration. Histopathology showed vertical strands of collagen perforating from the ulcerated lesions. The authors also reviewed the literature on this subject.

Key word : Reactive Perforating Collagenosis, Diabetes Mellitus, Chronic Renal Insufficiency

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Kyrle originally described hyperkeratosis follicularis and parafollicularis in cutem penetrans in a 22-year-old woman with diabetes in 1916[1,2]. In 1967, Mehregan et al first used the term perforating collagenosis to describe the disorder beginning early in childhood[1-3]. Poliak et al[2,4] in 1982, described an acquired form of perforating collagenosis beginning in adulthood[4,5] and associated with diabetes mellitus and renal failure[4].

Reactive perforating collagenosis (RPC) is an uncommon condition[6]. Two forms of this condition have been reported, first an inherited autosomal recessive form and second a sporadic acquired form. The inherited form occurs in childhood while the sporadic form occurs in adulthood and is associated with systemic diseases especially diabetes mellitus. This lesion also arises in patients undergoing hemodialysis[1,4,5,7-11]. Both forms of RPC

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are believed to be a cutaneous response to superficial trauma[3,12,13].

The authors report a patient with acquired reactive perforating collagenosis associated with diabetes mellitus and its complications. To the best of our knowledge, this is the first reported case in Thailand.

CASE REPORT

A 57- year-old Thai woman had had diabetes mellitus for about ten years. In 1997 she progressed to end stage renal failure and has been maintained on hemodialysis since then. At this admission, she had dermatologic complaints. The physical examination revealed generalized multiple discrete erythematous papules (Fig. 1). Some showed central umbilication and ulceration with a firmly adherent keratotic plug particularly on the extensor surface of the extremities (Fig. 2). Some were in linear distribution and others were excoriated and inflamed. The eruptions were itchy. Koebner's phenomenon was evident. She also had dry gangrene of the right fourth toe. The roentgenogram of the soft tissue on the right foot and leg failed to reveal calcification. She was treated with cefpirome (fourth generation cephalosporins), metronidazole, hydroxyzine HCL, ketosteril (a-ketoanalogue to essential amino acids) and pentoxifylline without any improvement. A biopsy taken from a lesion on the right thigh showed a dome-shaped lesion with a central crater that extended from the epidermis to the reticular dermis and contained basophilic debris, horny material, neutrophils and degenerated collagen in vertical strands. The epidermis adjacent to the crater was hyperplastic, thin near the base of the crater and completely absent at its base where the collagen was extruded. A dense accumulation of neutrophils, lymphocytes, histiocytes and cellular debris was demonstrated where the dermis was in direct contact with the contents of the crater (Fig. 3 and 4). Masson's trichrome stain revealed collagen within the epithelium - lined crater. Most collagen bundles were degenerated and fragmented and were not blue. The diagnoses of acquired reactive perforating collagenosis and progressive ischemic gangrene were established. Allopurinol was prescribed orally 100 mg a day without any improvement. She developed congestive heart failure, cerebrovascular accident and gastrointestinal bleeding three weeks after the amputation of the right fourth toe.

DISCUSSION

RPC, described by Mehregan et al in 1967, is one of the four diseases that have been considered the essential perforating disorders: elastosis perforans serpiginosa, RPC, perforating folliculitis, and Kyrle's disease[6,14]. In the inherited or childhood form, the lesions appear early in life, mainly on exposed surfaces. A genetic predisposition has been suggested by a positive family history in at least two thirds of the patients[3,6,15-17]. The male-to-female ratio is

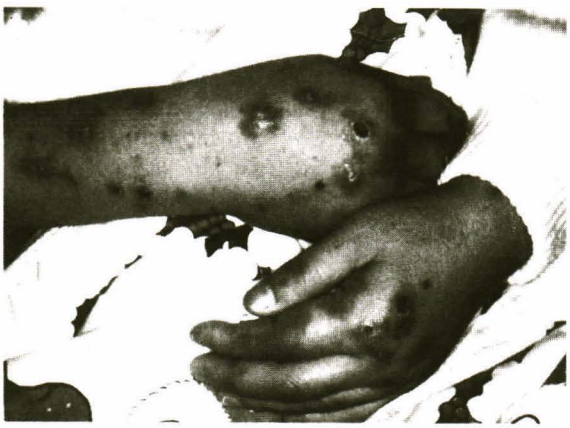

Fig. 1. Clinical photograph shows multiple scattered hyperkeratotic nodules and plaques on both hands.

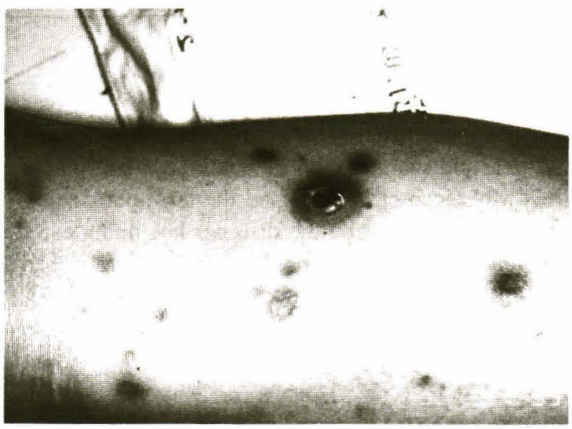

Fig. 2. Close-up view of a hyperkeratotic umbilicated nodule on the right leg.

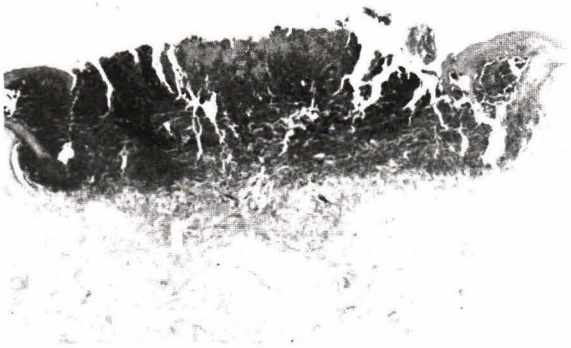

Fig. 3. Photomicrograph shows a cup-shaped umbilicated depression that was plugged by collagen and necrotic inflammatory cells. (H&E; 40X)

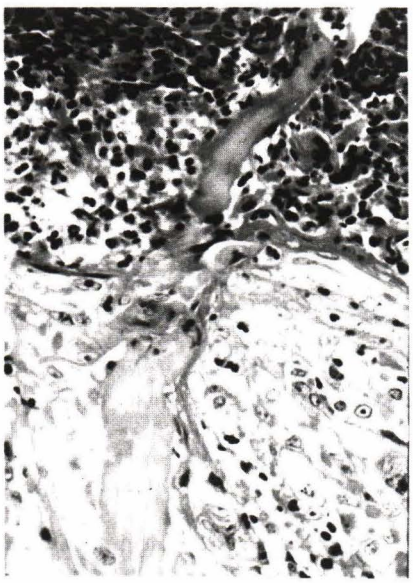

Fig. 4. Photomicrograph shows vertical strands of collagen perforating through the epidermis. (H&E; 200X)

2 : 1[6]. The acquired or adult form occurs in adulthood, is not familial and is usually accompanied by scratching, excoriation and pruritus[4,6,18]. There is no predilection to sex or race. Both forms are believed to be a cutaneous response to superficial trauma[6,19-21].

In both forms, the lesions are multiple, usually recurrent, umbilicated, hyperpigmented papules and nodules with a central adhering keratotic plug that may resolve spontaneously in 6 to 8 weeks [6]. The lesions are located mostly on the extensor surfaces of the extremities and on areas that can be reached easily by hands[6,22] and related to pruritus, since it can occur with diabetes mellitus, lymphomas, hyperparathyroidism, and hypoparathyroidism[6].

The pathogenesis is still unknown[6]. Mehregan et al originally postulated that mild superficial trauma in genetically susceptible persons leads to necrobiosis of the collagen in the dermal papillae, which is subsequently eliminated from the dermis by means of "transepidermal elimination" of altered collagen[3,6,18]. Others consider trauma to be an insufficient factor[6]. Koebner's phenomenon (linear development of the lesions along the scratch marks) is seen in almost all patients[23]. Bovenmyer succeeded in producing such a lesion by a needle scratch [19]. Cochrane et al suggested that diabetic vasculopathy accompanied by trauma from scratching caused by diabetes and renal insufficiency may be the underlying factor[1,5]. Kawakami and Saito concluded that trauma and microvasculopathy are triggers of transepidermal elimination and degeneration of collagen bundles. Major structural alterations in collagen are not considered to be the triggers of the transepidermal elimination process[24]. Ultrastructurally, the collagen fibers being eliminated showed normal periodicity [24,25]. Yanagihara et al proposed the mechanism in RPC as follows: 1) in the developing stage, the regeneration of epidermis progresses between the necrotic mass and the reticular dermis and among the collagen bundles; the collagen bundles remain in the channels of epidermis and 2) the regenerated epidermis makes the thick horny layer; the necrotic masses are lifted up and the collagen bundles are pulled up from the dermis through the epidermal channels[23]. The extruded collagens show immunoreactively against type IV collagen, providing an evidence that they may be derived from the basement membrane zone[26]. However, little is known about the altered collagen and its extrusion through the epidermis. It is possible that the altered collagen is rejected in a way similar to a foreign body reaction[2].

Faver et al proposed the diagnostic criteria for the acquired form of RPC: 1) histopathological findings of transepidermal elimination of necrotic

basophilic collagen bundles into a cup-shaped epidermal depression; 2) umbilicated papules or nodules with a central adherent keratotic plug; and 3) onset of the lesions after 18 years of age[6].

The acquired form of RPC is associated with many systemic diseases, such as diabetes mellitus or its complications (retinopathy, peripheral vascular disease, or cardiomyopathy)[6,27], chronic renal failure with or without dialysis[1,4,5,6,12,18], lepromatous leprosy[27,28], Hodgkin's lymphoma[18,20], acquired immune deficiency syndrome (AIDS)[6, 29], and endocrinopathies[6].

The authors believe that this case of acquired type of RPC is confirmed histologically and fulfills the criteria proposed by Faver et al. In this patient the trauma was scratching secondary to pruritus and dermal necrosis resulting from a poor blood supply associated with diabetic vasculopathy.

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รายงานผู้ป่วย 1 รายที่เป็นโรค acquired reactive perforating collagenosis และการทบทวนวารสาร†

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ได้รายงานผู้ป่วยหญิงไทยหนึ่งรายอายุ 57 ปี ที่ป่วยเป็นโรคเบาหวานร่วมกับภาวะไตวายที่ต้องได้รับการฟอกเลือด และมีการเน่าตายแบบแห้งของนิ้วนางของเท้าขวา ผลการตรวจร่างกายพบมีตุ่มนูนและผื่นนูนสีแดงขนาดต่าง ๆ กัน บนผิวหนังกระจายทั่วไป บางรอยโรคมีแผลเกิดขึ้น ลักษณะทางจุลพยาธิวิทยาพบมีเส้นใยคอลลาเจนแทงทะลุรอยโรคที่เป็นแผลขึ้นมา ผู้เสนอรายงานได้ทบทวนวารสารที่เกี่ยวข้องกับโรคนี้ด้วย

คำสำคัญ : เส้นใยคอลลาเจนแทงทะลุแผลที่ผิวหนัง, เบาหวาน, ไตวายเรื้อรัง

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