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Acquired Reactive Perforating Collagenosis

Akkiz Reaktif Perforan Kollajenoz

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Abstract

Acquired reactive perforating collagenosis (ARPC) is commonly recognized as an unusual skin reaction to superficial trauma that is observed in patients with a certain genetic predisposition or underlying diseases, such as diabetes mellitus or renal diseases. We present the unusual case of a 55-year-old female diabetic patient with numerous characteristic dome-shaped nodules, which consisted of central umbilication containing firm keratotic plugs.

Key Words: Acquired reactive perforating collagenosis, Diabetes mellitus.

Özet

Akkiz reaktif perforan kollajenoz sıklıkla diabetes mellitus veya renal hastalıklar gibi altta yatan hastalıklar veya belli bir genetik yatkınlığı olan hastalarda görülen yüzeyel travmaya nadir bir cilt reaksiyonu olarak tanımlanır. Biz burada 55 yaşında diabetik karakteristik çok sayıda kubbe şekilli noduller, merkezi umblikasyon gösteren sert keratotik tıkaçları bulunan nadir kadın vakayı sunuyoruz.

Anahtar Kelimeler: Akkiz reaktif perforan kollajenoz, Diabetes mellitus.

Introduction

Acquired reactive perforating collagenosis (ARPC) is a skin disorder that is characterized by the transepidermal elimination of altered collagen through the epidermis [1]. An inherited form was first described by Mehregan et al. [2] in 1967. The adult form, which commonly manifests in diabetics, has been reported in 10% of patients undergoing dialysis and has been mostly ascribed to diabetic nephropathy [3]. We hereby describe a case of acquired reactive perforating collagenosis in a patient with diabetes mellitus.

Case Report

A 55-year-old woman presented with a 4 month history of skin eruption and pruritus on her lower extremities. She had an 18 year history of type 2 diabetes mellitus that was treated with subcutaneous insulin injections. The skin eruptions developed on both legs. At the time of presentation, there were numerous dome-shaped nodules, which were characterized by central umbilications containing firm keratotic plugs, and erythema was observed around the nodules (Figure 1). Comprehensive laboratory investigations revealed no signs of renal dysfunction, including a urea level of 52 mg/dL and a creatinine level of 0.8 mg/dL; however, the mean fasting blood glucose level was 155 mg/dL, and there was an elevated glycosylated hemoglobin of 8.6%. The patient did not have a renal disorder due to the diabetes mellitus. Histopathological evaluation of a representative skin lesion showed transepidermal elimination of necrotic collagen bundles into a cup-shaped epidermal depression. Numerous neutrophil polymorphonuclears were observed. Marked neutrophilia, in addition to mononuclear cell infiltration particularly of macrophages and giant cells, was observed under the dome-shaped lesions and debris (Figure 2). Van Gieson simple staining showed degenerated collagen fibers that had perforated through the epidermis (Figure 3). Therefore, an ARPC diagnosis was verified, and the patient was referred to an endocrinologist for the management of her diabetes mellitus and underwent narrow UVB treatment, without significant amelioration.

Discussion

Perforating disorders are uncommon diseases that are histopathologically characterized by transepidermal elimination and include reactive perforating collagenosis, elastosis perforans serpiginosa, Kyrle's disease and perforating folliculitis [4]. This classification was made according to the type of epidermal damage and the characteristics of

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Figure 1. *Umblicated and erythematous papules with adherent central keratotic plug on lower extremities.*

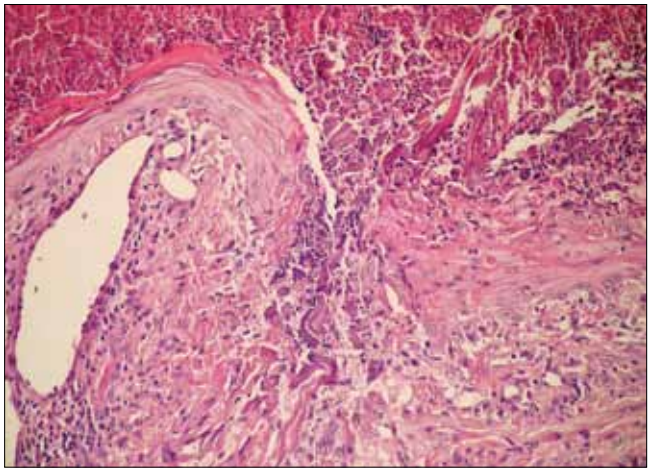

Figure 2. *Transepidermal elimination of necrotic collagen bundles into a cup-shaped epidermal depression.*

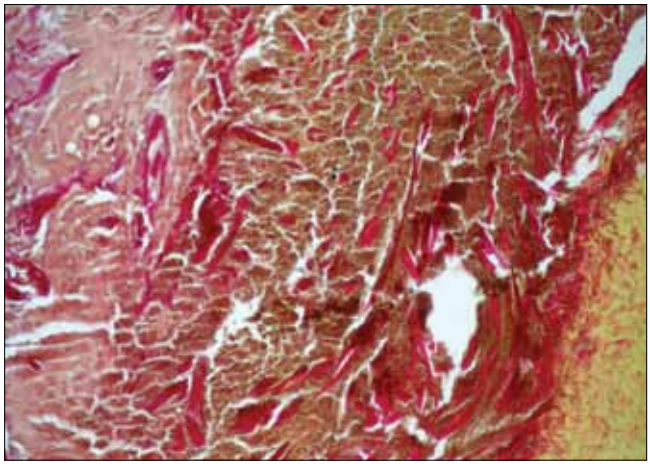

Figure 3. *Van Gieson simple staining showed degenerated collagen fibres.*

the eliminated material [5]. The pathogenesis of ARPC is unknown. As originally postulated by Mehregan et al. [2], "mild superficial trauma in genetically susceptible persons leads to necrobiosis of collagen in dermal papillae, which is subsequently eliminated from the dermis by means of transepithelial elimination," still seems to be correct. In ARPC, the defect occurs in the papillary dermis where histochemically altered but ultrastructurally intact-type IV collagen is present; the collagen is surrounded and engulfed by focal epidermal proliferation. A central crater containing inflammatory cells, keratinous material and altered collagen then develops and the altered collagen is subsequently expelled by transepithelial migration [6]. ARPC has been observed in association with multiple disorders, including diabetes mellitus, renal failure, hyperparathyroidism, liver disease, neurodermatitis, IgA nephropathy, periampullary carcinoma with jaundice, adenocarcinoma and liver neoplasms [7].

The disease presents clinically as umbilicated papules with a central adherent keratotic plug [8]. The hands and legs are frequently involved. In most cases of ARPC, patients experience severe pruritus and Köbner's phenomenon. Thus, it is thought that traumatic stimulation, such as scratching, induces the transepidermal elimination of the degenerated collagen fibers [9[. Severe pruritus and Köbner's phenomenon were present in our patient.

The possible biochemical or immunological mechanisms of the systemic diseases that are potentially responsible for the development and appearance of ARPC are still under investigation [10]. There is no specific treatment for ARPC. Topical and oral retinoids and topical and intralesional glucocorticoids have been reported to be effective in some patients in a case series [11]. In addition, individual lesions can be excised. Tretinoin 0.1% cream may also be effective. ARPC is usually refractory to treatment; however, there have been reports of ARPC that has been successfully treated with phototherapy. The course of this disease is chronic, and if the associated diseases are treated, patients may have a better prognosis [12].

In conclusion ARPC is a rare disease in terms of the clinical and histological findings. In the presence of diabetes mellitus and severe pruritus, ARPC must be considered in the differential diagnosis.

Conflict of interest statement: The authors declare that they have no conflict of interest to the publication of this article.

References

1. Khan Y, Swain JP. Acquired reactive perforating collagenosis in an insulin dependent diabetes mellitus patient. J Indian Med Assoc 2009; 107: 825-6.
2. Mehregan AH, Schwartz OD, Livingood CS. Reactive perforating collagenosis. Arch Dermatol 1967; 96: 277-82. **[CrossRef]**

3. Brinkmeier T, Herbst RA, Frosch PJ. Reactive perforating collagenosis associated with scabies in a diabetic. J Eur Acad Dermatol Venereol 2004; 18: 588-90. **[CrossRef]**
4. Lee SJ, Jang JW, Lee WC, et al. Perforating disorder caused by salt-water application and its experimental induction. Int J Dermatol 2005; 44: 210-4. **[CrossRef]**
5. Gambichler T, Birkner L, Stücker M, Othlinghaus N, Altmeyer P, Kreuter A. Up-regulation of transforming growth factor-beta3 and extracellular matrix proteins in acquired reactive perforating collagenosis. J Am Acad Dermatol 2009; 60: 463-9. **[CrossRef]**
6. Yadav MK, Sangal BC, Bhargav P, Jai PR, Goval M. Reactive perforating collagenosis. Indian J Pathol Microbiol 2009; 52: 106-7. **[CrossRef]**
7. Schmults CA. Acquired reactive perforating collagenosis. Dermatol Online J 2002; 8: 8.
8. Weiss SC, Moschella SL, Kwan T, Craven DE. Cutaneous mucormycosis secondary to acquired reactive perforating collagenosis. Cutis 2003; 72: 119-23.
9. Yazdi S, Saadat P, Young S, Hamidi R, Vadmal MS. Acquired reactive perforating collagenosis with papillary thyroid carcinoma: a paraneoplastic phenomenon? Clin Exp Dermatol 2010; 35: 152-5. **[CrossRef]**
10. Karpouzis A, Giatromanolaki A, Sivridis E, Kouskoukis C. Acquired reactive perforating collagenosis: current status. J Dermatol 2010; 37: 585-92. **[CrossRef]**
11. Yancovitz M, Johnson H, Wang N, Pomeranz MK. Perforating collagenosis. Dermatol Online J 2008; 14: 14.
12. Golusin Z, Poljacki M, Matovic L, Tasic S, Vuckovic N. Kyrle's disease. Med Pregl 2002; 55: 47-50. **[CrossRef]**