Acquired reactive perforating collagenosis in Skin of Color

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Abstract

Acquired reactive perforating collagenosis is an uncommon disorder that present as keratotic plugs on the skin owing to transepidermal perforation of dermal connective tissue. Etiology is unclear, although the condition has been associated with renal disease, diabetes, and HIV. It can frequently be misdiagnosed and lead to significant impact on quality of life, particularly in patients with skin of color. Herein, we present an skin of color patient with this condition who experienced severe pruritus over years before receiving a definitive diagnosis. Clobetasol improved her condition and quality of life over several months.

Keywords: acquired, perforating, collagenosis

Introduction

Acquired reactive perforating collagenosis (ARPC) is a rare dermatosis that can be misdiagnosed for extended periods of time and may continue undiagnosed, particularly in skin of color, It impacts quality of life owing to associated pruritus. We present a case of longstanding disease in a skin of color patient.

Case Synopsis

A 61-year-old woman with a history of type two diabetes and rheumatoid arthritis presented with a several-year history of recurrent pruritus and papules on the right lower leg. Examination revealed violaceous to skin-colored papules with central thick crusting on the right lateral lower leg and hyperpigmented macules on the right lower medial leg at sites of previous involvement (**Figure 1**). Herpes simplex virus PCR testing was negative.

Tangential biopsy was performed for H&E (**Figure 2**) along with Verhoeff-Van Gieson (**Figure 3**) and trichrome staining (**Figure 4**). The diagnosis of acquired reactive proliferating collagenosis was made. Application of clobetasol ointment to the affected areas resulted in stabilization of lesions noted at follow-up appointment 5 months later.

Case Discussion

Acquired reactive proliferating collagenosis is a rare subtype of proliferating dermatoses. This condition results from transepidermal removal of damaged collagen fibers from the dermis [1]. Excoriation may be a trigger for the development of ARPC, similar to the development of prurigo nodularis [2]. However,

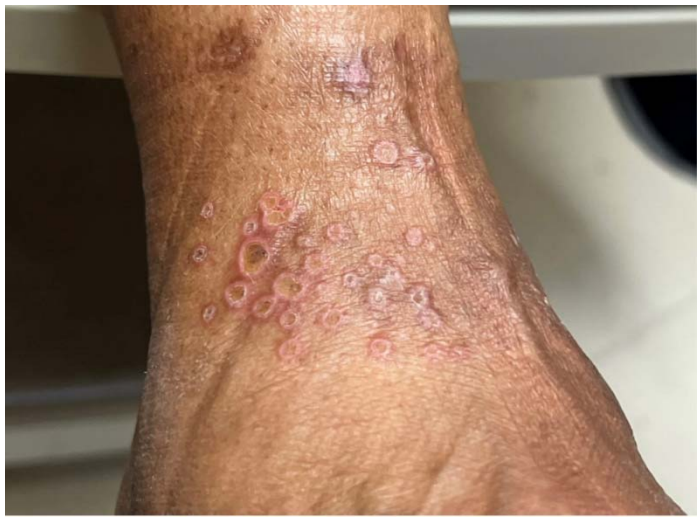

Figure 1. *Active round papules with central hyperkeratotic crust and hyperpigmented macules from prior lesions.*

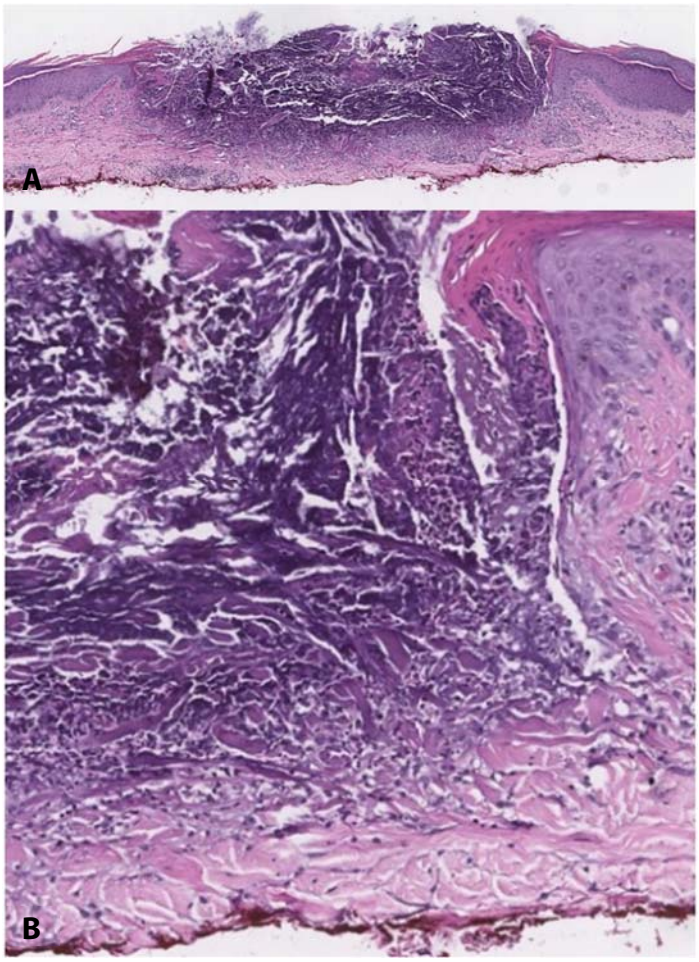

Figure 2. *H&E histopathology **A)** demonstrates cup-shaped invagination in the epidermis filled with central basophilic plug of keratin, collagen and inflammatory debris surrounded by epidermal hyperplasia, hyperkeratosis and parakeratosis, 4×. **B)** Higher magnification demonstrating similar findings, 20×.*

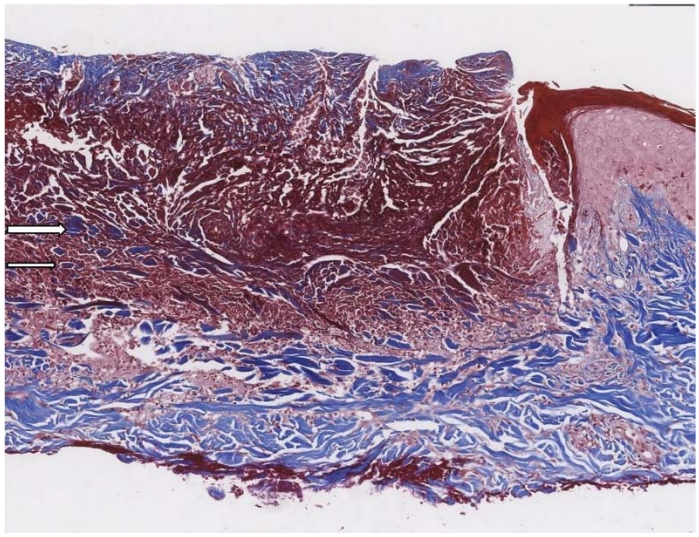

Figure 3. *Trichrome stain highlights the perforating collagen fibers that are stained blue as indicated by the arrows, 10×.*

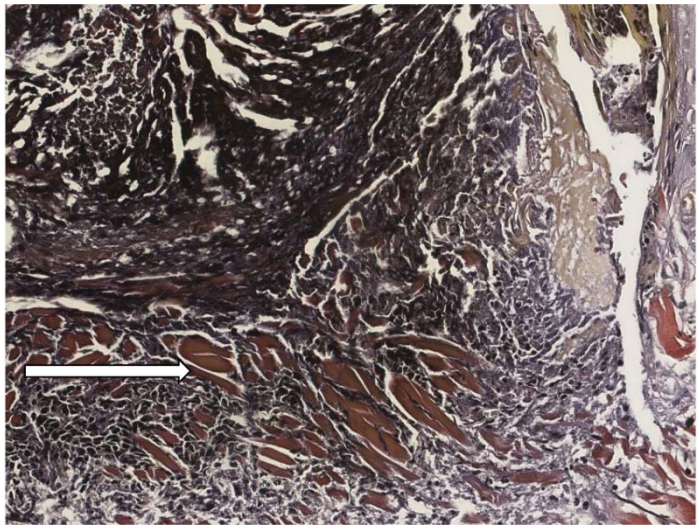

Figure 4. *Verhoeff Van Gieson stain highlights perforating collagen fibers (as indicated by the arrow) without substantial elastin perforation, 20×.*

the pathogenesis of ARPC is not fully understood. Both acquired and hereditary types exist with the hereditary form presenting in childhood and associated with trauma to the skin around the arms and hands. The acquired form more often presents in adulthood and frequently is associated with type two diabetes mellitus [3].

No official diagnostic criteria exist for ARPC. However, Faver et al. proposed the following criteria for APRC: "1) histopathologic findings of elimination of necrotic basophilic collagen tissue into a cup-shaped epidermal depression, 2) clinical presentation of umbilicated papules or nodules with a central adherent keratotic plug, and 3) onset of skin lesions after the age of 18 years" [4]. Our patient case meets the diagnosis under these criteria. In a study reviewing 101 cases of ARPC, Karpouzis et al. found the most common age of onset in the fifth decade of life with a majority of cases having diabetes mellitus. Acquired reactive proliferating collagenosis also has been found to be associated with chronic kidney disease, HIV, malignancy, and several other inflammatory conditions [5,6]. Some authors have found that controlling underlying hyperglycemia and treatment of concomitant malignancy resulted in improvement of lesions without other intervention.

Treatment of ARPC can be challenging owing to its recurrent nature and incomplete elucidation of

pathophysiology. Although the condition is often self-resolving in two to three months, it often recurs and can resolve with atrophic scars. Symptomatic management is usually necessary owing to the pruritus and itch-scratch cycle that can perpetuate the disease. Treatments that have been reported include topical corticosteroids, topical and systemic retinoids, oral antihistamines, allopurinol, and UVB phototherapy therapy, with varying degrees of success [3]. In our patient's case, she was treated with multiple rounds of topical clobetasol owing to recurrence with gradual sustained resolution of disease after several months.

Conclusion

We report a case of ARPC associated with diabetes mellitus and rheumatoid arthritis in a patient with skin of color. Although relatively uncommon, this condition can be important to recognize in this patient demographic and treat early on given impact of pruritus on quality of life.

Potential conflicts of interest

The authors declare no conflicts of interest.

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