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Punched out Ulcers with Eschar Like Floor – A Rare Presentation of Acquired Reactive Perforating Collagenosis without Systemic Associations

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Abstract

Observation: Acquired perforating collagenosis is a type of transepidermal elimination disorder which is usually associated with systemic illness like diabetes mellitus and renal disorders. Here we present a case of acquired perforating collagenosis without any systemic associations and therapeutic response to systemic steroids which is not yet reported in the literature.

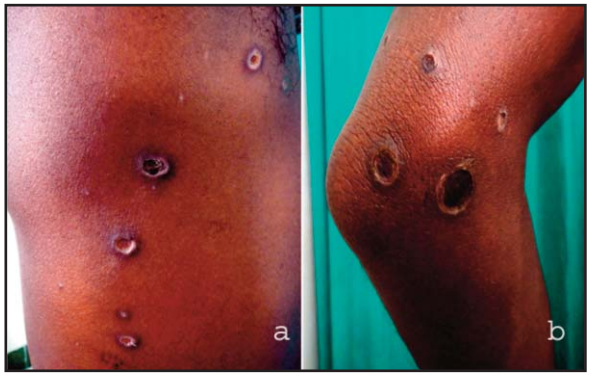
Introduction

Reactive perforating collagenosis (RPC) is a type of trans epidermal elimination disorder with extrusion of altered collagen through the epidermis. It exists in 2 forms, inherited and acquired [1]. Here we present a case of acquired RPC with unusual morphology, without any systemic associations and rapid response to systemic steroids.

Case Report

A 43 year old male, presented to dermatology OPD with asymptomatic multiple ulcers over the trunk and extremities of 4 months duration. There was no past history of systemic illness like diabetes mellitus or renal disease. Family history was also negative. Lesions started as papules with central umbilication which gradually increased in size with central keratotic plug.

Dermatological examination revealed multiple discrete well defined punched out ulcers with eschar like floor distributed over the lateral aspect of trunk and extensor aspect of both upper and lower limbs (**Figures 1a and b**). A few papular lesions with central umbilication were seen over the lateral aspect of trunk. Palms and soles, nails, mucosa and hair were normal. No significant findings were revealed in systemic examination. Investigations including complete blood count, fasting and post prandial blood sugar, urine routine, peripheral smear, chest X-Ray, liver, renal and thyroid function tests were within normal limits. HIV screening, VDRL and mantoux test were done and was found to be negative. With the above history and clinical examination, differential diagnosis of Cutaneous vasculitis, perforating dermatosis and pa-



Figures 1a and b. Clinical photographs showing circular punched out ulcers with eschar like floor

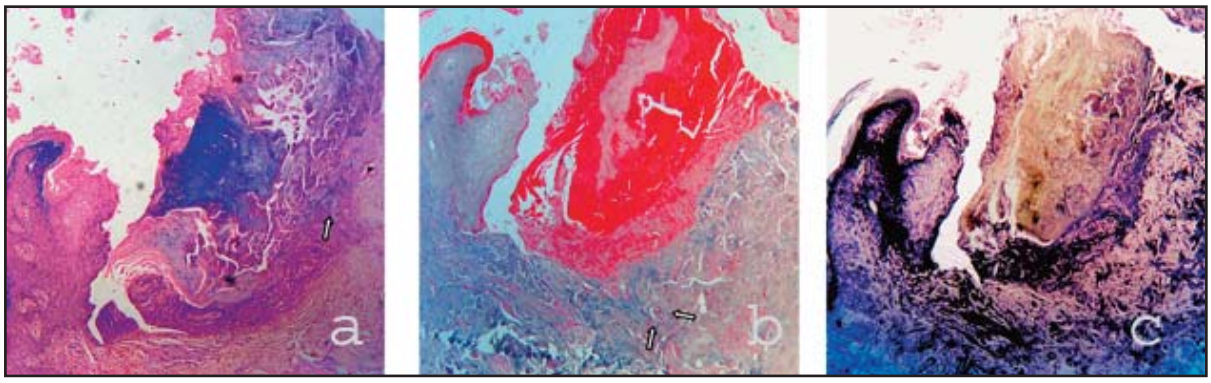


Figure 2a, b and c. a: Epidermis with cup shaped invagination and surface ulceration filled with keratotic and degenerated basophilic material and neutrophils showing perforation into the dermis (H&E, original magnification 10x)
b: Dermis shows haphazardly and vertically arranged collagen bundles impinging the perforated invagination. (Masson Trichrome, original magnification 10X)
c: Thick and haphazardly oriented elastic fibers in the dermis (Verhoeff -van Gieson, original magnification 10X)

pulonecrotic tuberculid were considered. Histopathological examination of the lesions showed the following findings. Epidermis showed cup shaped invagination surrounded by acanthosis and surface ulceration. The invagination is filled with keratotic and degenerated basophilic material with neutrophills. Base of invagination showed perforation into the dermis. Dermis showed acute and chronic inflammatory infiltrate along with thick and haphazardly oriented elastic fibers. Special staining (**Figures 2a, b and c**) with Masson Trichrome and Von Gieson showed vertically oriented degenerated collagen bundles impinging the perforated invagination.

Based on clinical and histo-pathological findings, the diagnosis of acquired reactive perforating collagenosis (ARPC) was made. Patient was put on systemic steroids for a period of 2 weeks. Lesions started resolving after 2 weeks and complete resolution of lesions with post inflammatory hyper-pigmentation was seen in a period of 2 months. Patient was followed up for six months with no recurrence.

Discussion

Acquired perforating dermatosis (APC) can be divided into four types depending on the epidermal disruption and nature of materials extruded through the epidermis. They include acquired reactive perforating collagenosis, Kyrle's disease, elastosis perforans serpiginosa and perforating folliculitis [**2**]. APC is usually seen associated with diabetes mellitus [**1**] and renal diseases [**3**]. There are other reports of ARPC associated with hepatocellular carcinoma [**4**], Hodgkin's lymphoma [**5**] chronic myeloid leukemia [**6**], acquired immunodeficiency syndrome [**7**], atopic dermatitis [**8**], pulmonary aspergillosis [**9**] and pregnancy [**10**]. In contrast our case has none of the reported or other systemic illness. The asymptomatic nature of the ulcers, normal haemogram and ESR with a negative culture report ruled out vasculitic and infective etiology. Papulonecrotic tuberculid was ruled out with the help of histopathology findings, negative mantoux test and normal ESR. The histopathological differential diagnoses considered were perforating folliculitis , Kyrle's disease and ARPC. As majority of the lesions were nonfollicular and invagination of epidermis was not involving hair follicle, perforating folliculitis was ruled out. The points against Kyrle's disease were presence of collagen impinging on the perforation and elastic fibers in the dermis as evidenced by special staining.

Reactive perforating collagenosis was first described by *Mehergan* et al in the year 1967 [**11**]. Our case has met the diagnostic criteria of Faver's [**1**] which include: 1) histopathological findings of transepidermal elimination of necrotic basophilic collagen bundles into a cup shaped epidermal depression. 2) umbilicated papules with a central adherent keratotic plug and 3) onset of the lesions after 18 years of age. The recommended treatment modalities for APC includes allopurinol, [**11**] topical or intralesional steroids, PUVA [**12**], NBUVB [**12**], tacalcitol, tazarotene [**13**] and doxycycline [**14**]. In our case there was complete resolution to systemic steroids which is not yet reported in the literature.

Conclusion

Our case was a clinical diagnostic challenge due to its peculiar morphology as punched out ulcer with eschar like floor. Absence of any associated systemic illness and complete subsidence of lesions with short course of systemic steroids worth a special mention here.

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