ORIGINAL ARTICLE

Perforating disorders of the skin

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ABSTRACT

Background: Perforating disorders of the skin, is an often overlooked entity characterized by transepidermal elimination of material from the upper dermis and are classified histopathologically according to the type of epidermal disruption and the nature of the eliminated material. They include Kyrle's disease, perforating folliculitis, reactive perforating collagenosis, and elastosis perforans serpiginosa. **Aim:** The aim of this study was to delineate the clinical and histopathological features of perforating disorders of the skin. **Materials and Methods:** In our study, we reviewed last 2 years skin biopsies received by us. Hematoxylin and eosin sections were re-examined and histochemical stainings (elastic van Gieson and Masson trichrome stains) were also used for histopathological evaluation. **Results:** We reviewed five cases of perforating disorders of skin which included two cases of Kyrle's disease, two cases of reactive perforating collagenosis and a single case of perforating folliculitis. Two patients had family history of perforating dermatosis in their siblings and three had associated systemic disease. **Conclusion:** Perforating disorders of the skin should be considered when ulcer with keratotic plugs is found.

KEY WORDS: Kyrle's disease, perforating disorders, perforating folliculitis, reactive perforative collagenosis

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INTRODUCTION

The perforating disorders of the skin are a group of diseases sharing the common characteristic of transepidermal elimination and are classified histopathologically according to the type of epidermal disruption and the nature of the eliminated material.[1,2] They have classically been divided into four types: Kyrle's disease, perforating folliculitis, reactive perforating collagenosis, and elastosis perforans serpiginosa.[1,2]

In this study, we aimed to delineate the clinical and histopathological features of perforating disorders. We report five cases with perforating disorders of the skin.

MATERIALS AND METHODS

This is a retrospective study, conducted during a period of 2 years, from January 2010 to December 2012. During this period, we received 425 non-neoplastic skin biopsies, out of these five were perforating dermatoses. Data were obtained regarding the clinical presentation from the indoor case sheets. Perforating dermatoses was diagnosed on the basis of following clinical and histopathologic setting: Histopathological findings of transepidermal elimination of basophilic cellular debris into invagination of the epidermis and pruritic keratoticumblicated papules or nodules. The relevant slides were retrieved and reviewed. All were stained with hematoxylin and eosin, Elastic-van Gieson and Masson trichrome stains.

RESULTS

Our study included five patients; four women and one man. The clinical features of our patients are shown in Table 1. Their age at the time of initial examination ranged from 17 to 63 years. Also disease duration ranged from 1 month to 24 months. Two patients had family history of perforating dermatosis in their siblings; these patients presented at young age. Two patients had insulin-dependent diabetes mellitus and one patient had both diabetes mellitus along with hypertension.

All patients had an eruption of generally pruritic keratotic papules and nodules, primarily on the extensor surface of the extremities and the trunk [Figure 1a, 2a, 3a]. The lesions typically had a central keratin-filled crater and might be grouped to form small plaques or arranged in a linear fashion. Extensor surfaces of the lower extremities were the most commonly involved site, followed by the upper extremities and trunk.

Histopathological evaluations revealed three types of lesions:

1. Histopathological features of two cases showed a shallow cup shaped epidermal invagination filled with compact orthokeratosis. The

epidermis showed irregular hyperplasia, acanthosis and dyskeratotic cells [Figure 1 b, c]. There was dense lymphoplasmacytic infilterate in upper dermis. Also, Masson trichrome and elastic van Gieson stains were negative in the epidermis and in the crater. Hence, the overall histological appearance in both cases was of Kyrle's disease.

2. Histopathological features of two cases showed vertically oriented epidermal invagination densely packed with degenerated basophilic staining material and altered collagen bundles [Figure 2b]. The base was lined by attenuated epithelium with foci of erosion containing vertically oriented bundles of collagen [Figure 2c]. Adjacent epithelium shows acanthosis. The superficial and deep dermis showed mixed inflammatory infiltrate. There was no evidence of follicular involvement. Masson trichrome stain showed the presence of collagen in both the cases [Figure 2c] while Elastic van Gieson stain was negative in the epidermis and in the crater. The overall histological appearance was of Reactive perforating collagenosis.
3. Histopathological examination of a single case showed parakeratotic plug with neutrophils and hair shafts above dermis. Underlying epidermis showed small cup shaped invagination filled with keratin [Figure 3 b, c]. Serial sections show dense granulomatous inflammation composed of foreign body giant cells in dermis. Masson trichrome stain showed no collagen and diagnosis of perforating folliculitis was made.

DISCUSSION

The perforating disorders comprise a group of pathologic abnormalities sharing the common characteristic of transepidermal elimination.[1,2] This phenomenon is characterized by the elimination or extrusion of altered dermal substances. They have classically been divided into four types: Kyrle's disease, perforating folliculitis, reactive perforating collagenosis, and elastosis perforans serpiginosa. This classification is primarily based on the nature of the eliminated material and the type of epidermal disruption.[3] In

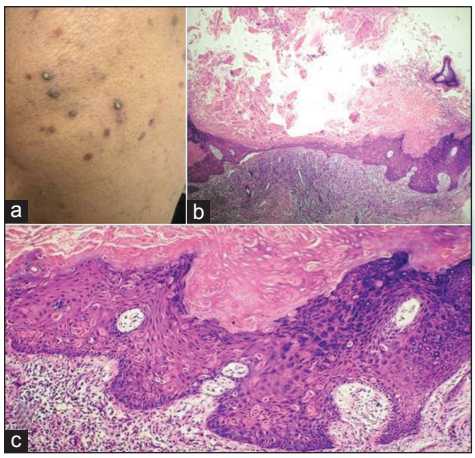

Figure 1: (a) Kyrle's disease: Multiple hyperpigmented plaques with central keratotic plugs on the leg of 45-year-old woman. (b) Photomicrograph showing shallow extrafollicular cup shaped epidermal invagination filled with compact orthokeratosis, dermis showing dense inflammatory infiltrate (H and E, 40x). (c) Photomicrograph showing dyskeratotic keratinocytes going upto the basal layer (H and E, 400x)

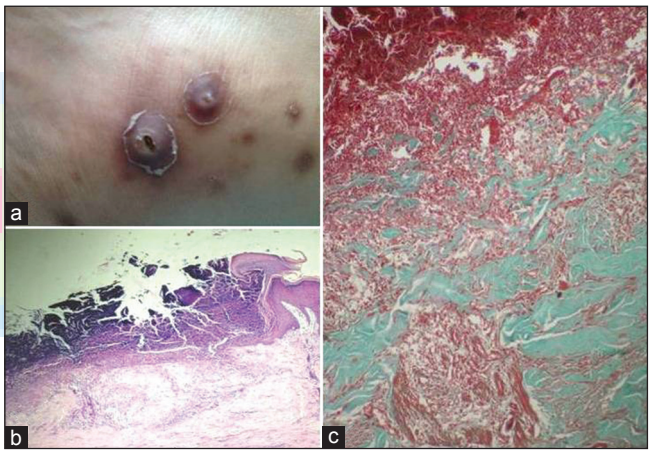

Figure 2: (a) Reactive perforating collagenosis: Multiple erythematous plaques with umbilication on the leg of 50 year old woman. (b) Photomicrograph showing shallow extrafollicular cup shaped epidermal invagination filled with compact orthokeratosis, dermis showing dense inflammatory infilterate (H and E, 40x). (c) Photomicrograph showing green stained collagen bundles present at the invagination (Masson trichrome, 100x)

Table 1: Clinical features of patients with perforating disorders of skin

Age/ Sex	*Distribution*	*Clinical appearance*	*Symptom*	*Past history*	*Diagnosis*
17/F	Extensor surface of upper and lower limbs	Hyperkeratotic nodules with crusted crateriform centre	Pruritus with excoriation and crusting	h/o similar lesions in sister	Kyrle's disease
45/F	Hands, feet, trunk and buttock	Hyperkeratotic macules, papules and nodules	Pain and pruritus	Uncontrolled DM since 5 yrs	Kyrle's disease
20/F	Forearm, legs and heels	Papules with umbilication	Pruritus	h/o similar lesions in brother	Reactive perforating collagenosis
50/F	Both extremities	Erythematous papules with umbilication	Severe pruritus	DM since 16 yrs on insulin; Hypertension since 2 yrs	Reactive perforating collagenosis
63/M	Abdomen, both extremities	Erythemaous papules and nodules with central keratotic plug	Pruritus	DM since 5 yrs	Perforating folliculitis

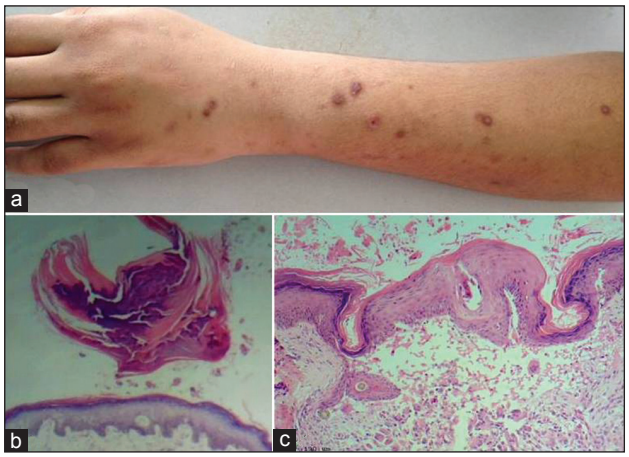

Figure 3: (a) Perforating folliculitis: Multiple erythematous papules on extensor surface of 63-year-old man. (b) Photomicrograph showing parakeratotic plug with neutrophils and hair shafts above epidermis (H and E, 40x). (c) Photomicrograph showing underlying epidermis showed small cup shaped invagination filled with keratin and dense granulomatous inflammation composed of foreign body giant cells in dermis (H and E, 100x)

our study, we reviewed five cases of perforating disorders of skin which included two cases of Kyrle's disease, two cases of reactive perforating collagenosis and a single case of perforating folliculitis. However, we did not observe any case of elastosis perforans serpiginosa.

Kyrle's disease was first described in 1916 by Kyrle as "hyperkeratosis follicularis et follicularis in cutem penetrans."[4] The primary event is claimed to be a disturbance of epidermal keratinization characterized by the formation of dyskeratotic foci and acceleration of the process of keratinisation.[1] This rapid production of abnormal keratin results in the formation of an overlying parakeratotic column, which perforates into the dermis, eliciting a granulomatous inflammatory reaction.[1,5] Subsequent re-epithelization from the adjacent epidermis covers this entire process from the base. The dermal connective tissue, inflammation and the keratotic debris degenerate to form the basophilic debris, which corresponds to the keratotic plug.[4] This is exuded from the invagination seen in the fully evolved form of the lesion.

Kyrle's disease has two distinct forms: An inherited form that presents in childhood and an acquired form that usually develops in adulthood, more commonly in women between 30 and 50 years of age associated with some underlying systemic disorder.[4,5,6] In our first case the patient developed the lesions at 3 years of age, she had no other systemic complaints and there was no history of similar complaints in the family; this was a case of primary Kyrle's disease. In the second case, the patient was a known diabetic since 5 years; this was secondary Kyrle's disease.

Kyrle's disease should be differentiated from other primary perforating dermatosis.[1] In perforating folliculitis, where the epidermal invagination is seen in relation to a vellus hair, which were absent in our cases.[7,8] In Elastosis perforans serpiginosa, there are thickened elastic fibers around epidermal invagination. Reactive perforating collagenosis was also ruled out due to the lack of degenerated collagen at the base of the perforation.[9]

Similar to Kyrle's disease, reactive perforative collagenosis is also seen in two forms: Inherited and secondary. We had our third case of primary reactive perforative collagenosis who had similar disease in her brother and fourth case of secondary reactive perforative collagenosis who had both diabetes and hypertension. The pathogenesis of this disorder is not known, but it is thought to be an abnormal response to scratching or other trauma, which leads to epidermal hyperplasia and transepithelial extrusion of collagen.[7,8]

Reactive perforating collagenosis should be differentiated from other primary perforating dermatosis.[1] In perforating folliculitis, the epidermal invagination is seen in relation to a vellus hair. In our case there was no vellus hair in relation to the epidermal invagination. In Elastosis perforans serpiginosa, there are thickened elastic fibers are seen around epidermal invagination. Elastic Von Geison stain did not reveal any elastic fibers.

Perforating folliculitis has overlapping features with Kyrle's disease. As described by Mehregan and Coskey, this is a relatively uncommon disorder usually observed in the second to fourth decade.[9] It is the end result of the abnormal follicular keratinization most likely caused by irritation, either chemical or physical and even chronic rubbing. A portion of a curled up hair is often seen close to or within the area of perforation or even in the dermis, surrounded by a foreign body granuloma.

Perforating folliculitis has to be distinguished from other perforating diseases of the skin. In Kyrle's disease, the keratinous plug may be extra follicular, the perforation usually is present deep in the invagination at the bottom of the keratinous plug and no eosinophilic degeneration of elastic fibers is found. Also, in Kyrle's disease, epithelial hyperplasia is seen. In elastosis perforans serpiginosa there is increase in elastic tissue in the dermis and especially in dermal papillae on staining with elastic tissue stains.

Perforating disorders of the skin should be considered when ulcer with keratotic plugs is found. A good interdisciplinary cooperation is crucial for the early recognition by histopathology.

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