CASE REPORT

Kyrle's Disease in a 57-year-old patient.

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ABSTRACT

Kyrle's disease is an acquired perforating dermatosis characterized by the presence of multiple hyperkeratotic papules, plaques and nodules commonly on the lower extremities. We report the case of a 57-year-old patient with papules and nodules on legs and abdomen whose histopathologic examination was compatible with Kyrle's disease.

INTRODUCTION

Kyrle's disease (lat. hyperkeratosis follicularis et follicularis in cutempenetrans) or follicular hyperkeratosis is an acquired perforating dermatosis that most commonly manifests in adulthood, usually associated with chronic kidney disease, diabetes mellitus and rarely with liver disease.[1] Skin lesions are characterized by erythematous and hyperpigmented papules and nodules with a crusty center and a keratotic cap, preferentially located on extensor surfaces of the extremities, trunk and rarely on the face and scalp.[2]

CLINICAL CASE

A 57-year-old male patient with a history of diabetes and renal insufficiency II, consulted the "Centro Dermatológico Dr. Uraga" for presenting hyperpigmented, eroded, multiple nodular papules with keratotic center located on the arms, legs and abdomen of 3 months of evolution, accompanied by intense pruritus (Figure 1). It was decided to take a biopsy sample from a lesion on the arm.

Histopathology showed the epidermis with acanthotic lateral margins, flattening and invagination in the center with a large amount of parakeratotic keratin and cellular remains of neutrophils. The dermis presents a large number of dilated capillaries and moderate inflammatory infiltrate, diagnostic compatible with perforating folliculitis (Kyrle's disease) (Figure 2). The patient was treated with topical corticosteroids and antihistamines, presenting improvement of the lesions.

DISCUSSION

Kyrle's disease was first diagnosed in the 20th century.[3] It was J. Kyrle who described it in 1916 in a woman with diabetes.[4]

The etiology of the disease is unknown but several hypotheses were proposed such as defective differentiation of the epidermis and dermoepidermal junction secondary to glycosylation processes, others may have infectious etiology by anaerobic bacteria and among others those of genetic etiology.[5]

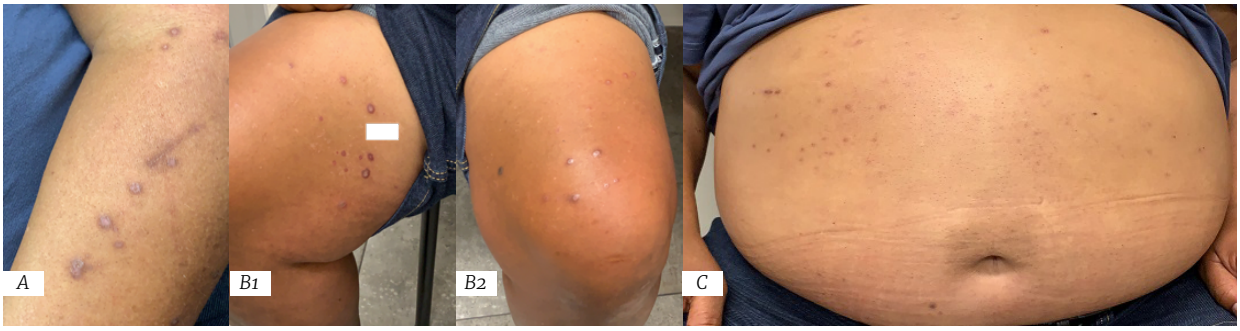

Figure 1. A) brownish papules on the arm. B1 and B2) keratotic center papules on legs. C) papules abdomen.

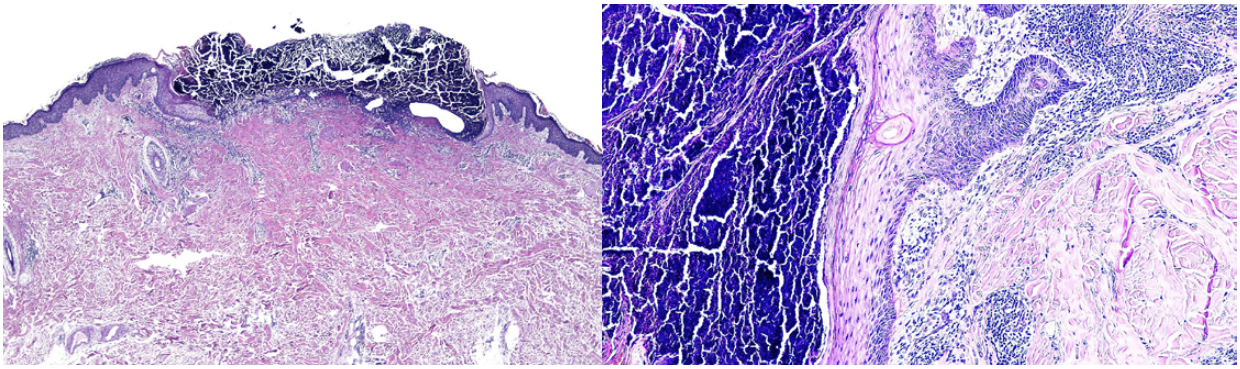

Figure 2. A) Acanthotic epidermis towards the lateral margins, keratin plug with parakeratosis and cellular debris mainly of neutrophils. B) In the adjacent dermis we observe a large number of dilated capillaries and moderate inflammatory infiltrate.

The pathogenesis of the disease is unknown, but superficial trauma is thought to be an inciter in patients susceptible to developing acquired perforating disorders, while vasculopathy and deposits of dermal exogenous materials appear to be predisposing factors.[5]

Clinically, the disease is described by the presence of papules or nodules with a central keratotic cap, the lesions may confluence and form plaques, mainly located on the extremities and less frequently on the trunk, pruritic in most cases.[1,2]

On dermoscopy there is no characteristic pattern that defines the disease, however there are two publications in the literature with suggestive patterns, describing a concentric pattern of 3 zones, characterized by bright whitish-brown scales in the center, a whitish-gray area without structure surrounding the central crusts and a peripheral brown pigmentation.[6,7]

Histopathology shows a keratotic plug in an epithelial invagination, keratinization may affect the entire thickness of the epidermis, there are basophilic cells, there may be lymphocytic and histiocytic infiltrate.[1,8]

Kyrle's disease must be differentiated from other disorders that present central hyperkeratotic plug among these: perforating folliculitis, elastosis perforans serpiginosa, prurigo nodularis, lichen planus, porokeratosis.[2,5]

Treatment options for Kyrle's disease include topical corticosteroids and retinoids, keratolytic nb-UVB and PUVA phototherapy, doxycycline, minocycline and acitretin; CO2 laser sessions, surgery and cryotherapy have also been performed. Maurelli et al. report the case of a 69-year-old patient with Kyrle's disease effectively treated with low doses of isotretinoin and complete remission 4 months after its use.[9]

The therapeutic combination of antihistamines to modulate pruritus together with topical steroids are the most widely used. Recent clinical trials support new immunomodulatory antipruritic drugs targeting IL4 receptors and intracellular signaling such as Janus kinase, these may have a potential role in the treatment of Kyrle's disease and show promise as emerging therapies in pruritic disorders as in other diseases.[10]

CONCLUSION

Kyrle's disease often develops as a consequence of previously diagnosed systemic diseases. As it is a recurrent disease, it can affect the patient's quality of life, so it is necessary to have an adequate diagnosis and treatment.

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