Kyrle disease: report of a Tunisian case

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INTRODUCTION

Kyrle disease (KD) was first described in 1916 by Kyrle under the name of hyperkeratosis follicularis and parafollicularis in cutem penetrans [1]. As he described, this is a disorder characterized by the formation multiple hyperkeratotic follicular and parafollicular papular lesions with a central keratotic plug. It is considered as a variant of primary perforating dermatoses which share the same pathophysiology: a transepidermal elimination of dermal substances [2]. The prognosis depends essentially on the association with some systemic disorders such as diabetes mellitus, renal failure, liver disease, congestive cardiac failure hyperlipidemia, etc [2].

CASE REPORT

A 60-year-old man, consulted for papulo-nodular lesions mainly located on the extremities. He had no medical past history particularly of renal failure or diabetes mellitus. On examination, there were multiple erythematous, purpuric, infiltrated and crusted lesions of extremities, buttocks and scalp. A punch biopsy showed a hyperkeratotic horny plug invaginating the epidermis. The diagnosis of Kyrle disease was retained. The clinicopathological features of this disease and its therapeutic problems are discussed.

DISCUSSION

Primary perforating disorders include Kyrle disease (KD) elastosis perforans serpiginosa, perforating folliculitis and reactive perforating collagenosis [1]. KD is a rare disorder of keratinization particularly observed in the setting of chronic renal failure and occurs in 10% of hemodialysed patients [2]. It affects both men and women throughout life, with a mean age of 30 years at onset [1]. However, cases in children have been reported [3]. KD seems to affect more frequently African Americans. The incidence of diabetes mellitus...
and renal failure is high in this population. Perforating dermatoses are quite rare. Thus, their pathogenesis is still misunderstood [4]. Some cases seem to be idiopathic inherited, but in other cases, KD occurs as cutaneous manifestation of a systemic disorder (diabetes mellitus, hepatic abnormalities, congestive heart failure, renal disease). The role of infectious agents, probably anaerobic bacteria, was suggested by some authors [4, 5]. Clinically, KD is characterized by silvery or red-brown papules or nodules centered by keratin plug or crusts. Some lesions appear to be follicular. They are not typically painful but may be very pruritic. Koebner’s phenomenon is exceptional. The lower extremities are mostly affected. Lesions may also develop in the arms and in the head and neck region. Keratotic lesions of conjunctiva and cornea were described in a single case report.

Histologically, Constantine and Carter suggested the presence of some criteria to diagnose KD: keratotic plug filling an epithelial invagination, parakeratosis in parts of the plug, basophilic cellular debris which does not stain with elastin stains and parakeratotic keratinised cells in at least one area deep to the plug. The clinical and histological differential diagnosis can be difficult between KD and perforating folliculitis because they are quite similar. The elastosis perforans serpiginosa should also be evoked [1].

Rapid improvement of lesions is often seen once the underlying disease is treated. Treatments that have been used to treat and reduce lesions include Isotretinoin, high dose vitamin A, Tretinoin cream, emollients and oral antihistamines.

**CONCLUSION**

The evolution of KD is unpredictable. The absence of a therapeutic consensus is due to the little number of reported cases. Therefore more cases are needed to understand the underlying pathogenesis and to improve the management.

**CONSENT**

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article.
REFERENCES