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Kyrle's disease (KD): "An Update with review of literature" A Spongebob Skin pores simulation

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ABSTRACT

Kyrle's disease (KD) is a Chronic skin condition first described by Austrian pathologist Josef Kyrle in 1916. Kyrle referred to this condition as hyperkeratosis follicularis & parafollicularis in cutem penetrans. These diseases are characterized by the phenomenon of transepidermal elimination of denatured dermis an acquired form of perforating dermatosis [14]. It is characterized by keratotic crater plugs that develop in hair follicles penetrating the epidermis and extending into the dermis [4]. This trans epithelial migration and elimination of proteinaceous components is associated with systemic disorders like renal, liver diseases, chronic heart failure and diabetes mellitus. We present two case scenarios of a young Males with multiple chronic papular eruptions along with a review of literature for Kyrle's disease (KD).

Keywords: Kyrle's disease (KD), metabolic disease, perforating dermatoses, primary disorder

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Introduction. Systemic disorders are extremely common in hospital wards. Skin disorders are a reflection of underlying diseases and the association between the two are often missed. Kyrle's disease (KD) is an example of such chronic painless skin disorder with pruritis characterised by migration of either keratin, collagen materials giving rise to intense pruritis. With Obesity and diabetes on the rise worldwide It is this intense itching on a myriad of rashes that every treating clinician needs to be aware which make Kryles disease merits worth of discussion.

CASE PRESENTATION

The patient was a 50-year-old male with a one year history of gradually progressive pruritic papules with central dark plugs that started on the lower legs and spread over to cover his neck, face and torso. He had chronic renal failure for 5 years and was on regular haemodialysis. He had no similar episode in the past and had no family history with a similar dermatological disorder. The early skin lesions started as hyperkeratotic papules of 2-5 mm in the lower limbs that had gradually spread to the trunk and face. In a few months these lesions enlarged to 10 mm in diameter. His Laboratory test were as follow serum creatinine was 15.45 mg/dl, serum uric acid was 9.1 mg/dl, and HBA1c was 7.8%. Urine routine and microscopy showed glucose 2 + and protein 3+, fasting blood sugar 180 mg/dl, Bacterial and fungi culture were negative. A Dermatological referral revealed depression with keratotic plugging over his torso and lower limbs (figure 1A, B). A histopathological examination revealed epidermal invagination into the dermis, epidermal hyperplasia, and plug formation that were consistent with Kyrle's disease (KD). He who had received treated with different topical and systemic regimens and had a partial resolution of symptoms.

The second patient was a 35 year old male with a history of popular rashes all over the trunk and back he had a long history of dermatological consultation and was referred to us for the evaluation of a focal abscess collection over his right lower back.

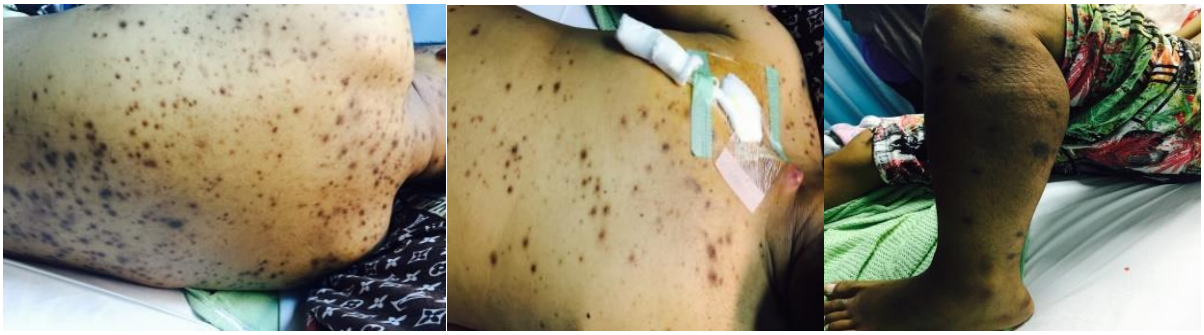

Figure 1 (A) Erythematous papules exhibiting a central depression with keratotic craters. (B) A close-up image of papules over front of chest & Abdomen. (C) Hyperpigmented papules lower limb.

Fig A.

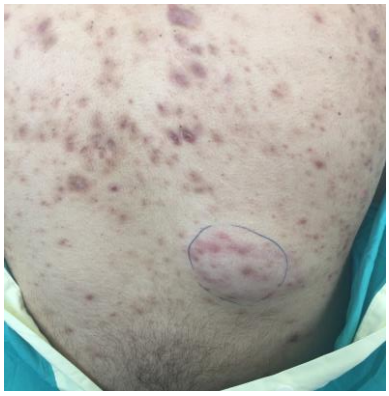

Fig B.

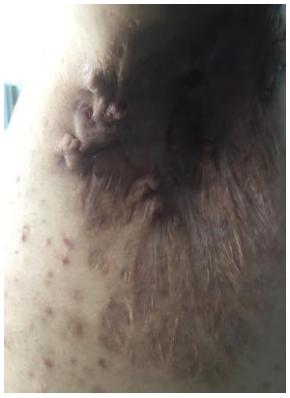

Figure 2 (A) Papules with keratotic craters, localised abscess (B) Axillary and truncal distribution

Discussion- The incidence of Kyrle's disease is reported higher in women than men. The common age at presentation is the third decade. Apart from adults It is reported that it might also exist in children [2] KD can be a primary or a secondary disorder. It can have to have a genetic inheritance [12,4] and is clinically characterised by hyperkeratotic parafollicular papules with a centrally localised plug, It usually involves the extensor surface of the extremities [10] The face torso and limbs may be involved but the mucous membranes and palms & soles are spared [9]. These plugs of keratin may fuse to form coil shaped plaques. The pathophysiology of Kryles disease is Chronic Itching induces microtrauma the combination of chronic inflammatory mediator's tissue hypoxia and altered wound healing, macro and microangiopathy coupled with oxidative stress in chronic metabolic disease states like diabetes, and liver disorders all set the stage for an acquired dermatosis [16].

There are currently two views on the reason for this trans epidermal migration, Pruritis induced repeated microtrauma that injures collagen and elastic tissue that induces transmigration [18]. Metabolic derangements induced micro deposition of calcium salts [19]. The plugs incite an inflammatory response and foreign-body giant cell reaction in the dermis adjacent to the lower end of the keratin plug, increased epithelial migration and proliferation, culminating in perforation is as a result of increased fibronectin concentration [17] Eventually there is a transcutaneous migration of elastic fibres, collagen and keratin plugs. Acquired KD is found frequently associated with systemic diseases like diabetes [6], chronic renal failure [1,11], liver and Chronic heart conditions.

A diagnostic criteria called Favers Criteria is described as meeting all of the following three criteria: (i) age of 18 years (ii) clinical presentation of umbilicated papules or nodules with a central adherent keratotic plug; and (iii) histopathological elimination of necrotic collagen [15]. Trans epidermal elimination is characterised with large keratotic and parakeratotic plugs penetrating from the epidermis through the dermis. At times there is a dislocation of the level of keratinization toward the dermal-epidermal junction [5]. It has been proposed that in Kyrle disease (KD) keratinization focally occurs at the basilar layer of the epidermal skin [3,1] There is a report on the role of an Infectious aetiology as there appears a regression of small lesions with antibiotics like clindamycin [7]. These lesions regress over time leaving behind scars and pigmented spots.

Treatment Itching is relieved with Local treatment with soothing agents, antipruritic lotions containing menthol and Systemic anti histaminic for severe cases. The recommended duration for use of topical retinoic acid (0.1%) as treatment is six weeks. Tacalcitol is used its for is action on the process of keratinization [8].

The chronic recalcitrant lesions need to be considered for surgical removal of the lesions using Laser, cryotherapy, electrotherapy [13]. Evidence based preference of one modality of treatment over the other are unfortunately lacking.

Conclusion Chronic dermatological conditions like Kryles disease are mirror reflection of the underlying body metabolic disease states that every Clinician ought to be familiar with. More studies are required before drawing conclusions on the most effective long term treatment modalities for chronic recalcitrant cases.

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