CASE REPORT

Kyrle's disease

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SUMMARY

Kyrle's disease (KD) is a dermatosis which was first described by Kyrle as "hyperkeratosis follicularis et para-follicularis in cutem penetrans" in 1916. Perforating dermatoses are a heterogeneous disorder group characterised by transepithelial elimination. KD has been seen in association with multiple disorders, including diabetes mellitus, renal and liver diseases, congestive heart failure, hyperlipidaemia, infective diseases and abnormal metabolism of vitamin A. This case report presents two patients with KD with associated systemic disease.

BACKGROUND

Kyrle's disease is clinically important because it is often associated with a systemic disorder and these cases were chosen since they were rare with interesting features.^{1–3}

CASE PRESENTATION

Case 1: A 38-year-old female patient presented with a 4-month history of a pruritic eruption of multiple, keratotic papules. The patient had a history of chronic renal failure and total blindness of the left eye. Dermatological examination revealed erythematous papules exhibiting a central depression with keratotic plugging over the extensor surfaces of knees and shins (figure 1A, B). Histopathological examination revealed epidermal invagination into the dermis, epidermal hyperplasia, compact hyperkeratosis, basal vacuolisation, degenerative basophilic material and plug formation consistent with KD. The presence of collagen fibres was not detected with Masson's trichrome histochemical staining in the epidermis (figure 1C). The patient was diagnosed with KD in the light of clinical and histopathological findings.

Case 2: A 62-year-old female patient presented with a 3-month history of multiple, itchy, keratotic papules over her shins. There was a medical history of diabetes mellitus, hypertension, asthma and hypertriglyceridaemia. Dermatological examination revealed that erythematous papules, which exhibited a central depression with keratotic plug, were located on the extensor surfaces of shins (figure 2A, B). Epidermal invagination into the dermis, compact hyperkeratosis of the epidermis, parakeratosis and plug formation were significant findings in the lesions. Dyskeratotic keratinocytes in the epidermis, vacuolar degeneration in the basal layer, dermal vascular proliferation and chronic inflammatory cell infiltration especially in neutrophils were also observed. The presence of collagen fibres was not detected with Masson's trichrome histochemical staining in the epidermis (figure 2C). The patient was diagnosed with KD in the light of clinical and histopathological findings.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of Kyrle's disease

- Primary perforating dermatoses
 - Acquired perforating collagenosis
 - Perforating folliculitis
 - Elastosis perforans serpiginosa
- Secondary perforation
 - Granuloma annulare
 - Pseudoxanthoma elasticum
 - Chondrodermatitis nodularis chronica helices⁴

TREATMENT

Topical retinoic acid (0.1%) treatment was recommended for 6 weeks. Furthermore, the patients were referred to consulting physicians for the treatment of their underlying diseases.

Figure 1 (A) Erythematous papules exhibiting a central depression with keratotic plug are located on the extensor surfaces of the knees and shins. (B) A close-up image of papules. (C) Epidermal invagination into the dermis, degenerated basophilic material and plug formation were noteworthy in the specimen. H&E staining $\times 40$.

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Figure 2 (A) Erythematous papules exhibiting a central depression with keratotic plug are located on the extensor surfaces of the shins. (B) A close-up image of papules. (C) Vacuolar degeneration of the epidermis and suppurated inflammation. H&E staining $\times 200$.

DISCUSSION

KD is one of the rare variants of primary perforating dermatosis.² Perforating disorders, including KD, reactive perforating collagenosis, elastosis perforans serpiginosa and perforating folliculitis, are a group of uncommon diseases, which are histopathologically characterised by transepidermal elimination.^{5–6} Although rare, its incidence is significantly increased in patients with chronic renal failure.^{7–9} This classification was established based on the type of epidermal damage and the characteristics of eliminated material.¹⁰ The common pathophysiological finding in these disorders is apparent transepidermal elimination. The eliminated material is predominantly collagen in the acquired perforating collagenosis, keratin in KD and perforating folliculitis, and thickened elastic fibres in elastosis perforans serpiginosa.⁴ KD is clinically characterised by hyperkeratotic parafollicular or follicular papules with a centrally localised cone-shaped plug, which can be removed easily. It usually involves the extensor surface of the extremities.¹¹ In general, the papules are discrete, but they may coalesce to form circinate plaques. The lower extremities are involved more easily, but the upper extremities, head and neck may also be affected. The patients experience pruritus and Koebner's phenomenon, which were also noted in our patients. KD is histologically characterised with large keratotic and parakeratotic plugs penetrating from the epidermis through the dermis. The plugs cause inflam-

matory response and foreign-body giant cell reaction in the dermis adjacent to the lower end of the plug. Mild degenerative changes in the connective tissue with no increase in the elastic tissue may also be noted. KD remains stationary for years, with possible clearing of lesions when the associated illness is under control. Treatment options are topical retinoic acid, isotretinoin and acitretin.¹² There are also some studies suggesting that phototherapy is an appropriate choice of treatment.¹³ Our patients were treated with 0.1% topical retinoic acid. They were also referred to consulting physicians for their underlying pathologies.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- If a patient presents with signs of Kyrle's disease, one has to investigate for underlying systemic disorders. However, Kyrle's disease often develops secondary to previously diagnosed systemic diseases, most commonly following chronic renal failure.
- Kyrle's disease should be considered in the differential diagnosis of erythematous papules exhibiting a central depression with keratotic plug.

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