

Case Report:

A case of Kyrle's disease with diabetes and renal insufficiency

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Abstract

A 58 year old male patient with type 2 diabetes and systemic hypertension presented with complaints of severe pruritic lesions all over the body; sparing face, palms, soles and mucosae. After control of diabetes, the skin lesions started to heal. By 8 weeks, the lesions had completely healed leaving behind hyperpigmented new lesions and the size of the existing lesions increased whenever his blood glucose levels were high. The patient also had renal insufficiency and significant proteinuria. He also had coronary artery disease (CAD) and New York Heart Association (NYHA) Class IV cardiac failure. Patient responded well to topical anti-inflammatory salicylic acid. The case is of interest as with better glycemic control, the skin lesions healed, leaving behind hyperpigmented scars.

Key words: Kyrle's disease, type 2 diabetes, renal insufficiency

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Introduction

Kyrle's disease is a rare skin disorder characterized by transepidermal elimination of abnormal keratin. Despite being described nearly a century ago (1), the etiology of the disease is still not known. However, it has been shown to be strongly associated with diabetes, chronic renal failure and hepatic dysfunction (2, 3). We describe here a case of Kyrle's disease in a patient with type 2 diabetes and renal insufficiency.

Case study

A 58 year old male patient with type 2 diabetes and systemic hypertension presented to us with complaints of severe pruritic lesions all over the

body. The lesions started over the back and then progressed to the legs, hands and scalp. The patient mentioned that new lesions appeared and the size of the existing lesions increased whenever his blood glucose levels were high. Itching was aggravated after intake of food, was more severe during night time and was relieved after a shower. The patient had a strong family history of diabetes but no one else in the family had similar skin lesions. Physical examination revealed multiple, well demarcated, papular lesions of variable sizes (0.5 to 2 cm), most prominent on the scalp, back and the extensor surface of the legs. Lesions were also present on knees, thighs, forearms and elbows, but the face, palms, soles and mucosae were spared. Lesions were asymmetrically distributed and hyperpigmented with central keratotic plaques which were round or irregular in shape. The base was filled with a necrotic plug. Hyperpigmentation was also observed around the papule and the consistency of the skin was hard to touch. New lesions showed signs of inflammation.

The patient also had renal insufficiency with a serum creatinine of 1.6 mg/dl and significant proteinuria (1 g/day). He also had coronary artery disease (CAD) and New York Heart Association (NYHA) Class IV cardiac failure.

At the time of the index visit, the patient had fasting plasma glucose level of 21.53 mmol/l, post-prandial plasma glucose level of 26.14 mmol/l and HbA1c level of 11.2%, indicating very poor glycemic

control. C-peptide assay revealed fairly good pancreatic beta cell reserve (fasting C-peptide 1.9 pmol/ml and stimulated C-peptide 2.4 pmol/ml) suggestive of type 2 diabetes. Hemogram showed leucocytosis. Liver function tests revealed elevated bilirubin, reversal of albumin: globulin ratio and elevation of alkaline phosphatase and gamma glutamyl transferase (GGT) levels. Hyponatremia (Na^+ 127 meq/l) was also present.

Ultrasound abdomen showed normal sized kidneys with renal calculi, right cortical cyst and bilateral pyelonephritis, with a renal stent in situ. The bladder wall was thickened. Echocardiogram results showed an ejection fraction (EF) of 28% with severe left ventricular (LV) systolic dysfunction suggestive of ischemic cardiomyopathy.

The diagnosis of Kyrle's disease was confirmed by a dermatologist (Figure 1). The patient was started on topical anti-inflammatory salicylic acid and the dose of insulin and oral antidiabetic agents were adjusted so as to normalize blood glucose levels. It was noted that as the degree of glycemic control improved, the skin lesions also started to heal. By 8 weeks, the lesions had completely healed leaving behind hyperpigmented scars. Eight months after the index visit, the patient's fasting and post-prandial plasma glucose were 4.16 mmol/l and 7.27 mmol/l and HbA1c was 7.5%. The patient continues to do well with no recurrence of skin lesions.

Discussion

Kyrle's disease is an uncommon skin disorder, representing a category of perforating dermatosis (PD) which involves the transepithelial elimination (TEE) of dermal structures. PD is a heterogeneous group of disorders which are categorized into four types based on physical and pathological investigations (4);

- 1) Elastosis perforans serpiginosa (EPS) involving TEE of eosinophilic elastic fibers.
- 2) Reactive perforating collagenosis (RPC) characterized by elimination of necrobiotic basophilic collagen bundles.
- 3) Perforating folliculitis (PF) illustrated by TEE of basophilic debris and eosinophilic elastic fibres into the hair follicles.

4) Kyrle's disease diagnosed based on the TEE of a granulomatous focus of basophilic debris into an epidermal invagination (4).



Figure 1: Kyrle's disease

Although the exact etiology of PD remains unknown, it has been found to be associated with renal failure in diabetes as shown by the report of Saray et al., (5) who showed that among 22 patients with PD, 72% had chronic renal failure and 50% had diabetes. Among patients with diabetes and PD, 90% had chronic renal failure. Among patients with PD, Kyrle's disease is a rare variant that has been shown to be associated with diabetes, renal disease and a spectrum of other disorders. This association has been confirmed in a report involving 21 cases of Kyrle's disease, where 19 cases presented with a renal disorder and 12 cases had diabetes (6).

Kyrle's disease, though known to affect adults in the third decade, occurs among children as well. It does not commonly involve mucous membranes, plantar or palmar surfaces, but one report has shown a rare case of Kyrle's disease with mucosal and palmo-plantar involvement in multiple family members (7). Although the hereditary nature of the disease is not clearly understood, it has been reported in siblings in a family of three generations (8, 9) suggesting possible genetic transmission and the mode of inheritance may be autosomal dominant or recessive. However, in our case, the disease was not found in any other family member, even those who had diabetes.

The pathogenesis of the disease and the mechanistic links that connect Kyrle's disease to diabetes mellitus are unknown. However, one hypothesis suggests that it may be an outcome of changes in the epidermis or dermis leading to metabolic derangements and a cutaneous response to the superficial trauma and vasculopathy associated with diabetes. Derangements in epidermal or dermal structure in diabetes might arise from products of oxidative damage or endoplasmic stress like advanced glycation end products, oxidized low density lipoprotein (LDL) etc. Other infectious and chemical agents have also been shown to cause Kyrle's disease and the infectious origin of the disease is supported by the

fact that anti-microbial therapy has resulted in successful regression of the disease (10 - 12).

The first line therapy for treatment of Kyrle's disease is keratolytics (salicylic acid and urea), followed by electrocautery or radiocautery. Surgical excision is reserved as the last option. In our case, anti-inflammatory salicylic acid was used. Our patient also had poorly controlled diabetes with nephropathy, systemic hypertension and CAD. Administration of insulin and stabilizing the glycemia resulted in disease regression. The hypoglycemic effect and anti-inflammatory nature of insulin might have resulted in improvement of the disease symptoms.

Conclusion

We report here a case of Kyrle's disease in a patient with type 2 diabetes and renal insufficiency. While Kyrle's disease is a rare entity, it has the potential to adversely affect the quality of life. Physicians should therefore maintain a high index of suspicion for Kyrle's disease in patients with diabetes, particularly in those with concomitant renal insufficiency, so that an accurate diagnosis can be made and appropriate management started at the earliest.

Conflict of interest

None declared.

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