Kyrle’s Disease: A Rare Skin Manifestation of Diabetes Mellitus

A Pandey¹, Keshri S Yadav², Gaurav Singh², M Chaturvedi³

Abstract

Kyrle’s disease is a rare skin disorder which is characterized by hyperkeratotic papules and nodules with a central keratotic plug mostly located in lower limbs. Exact etiology of Kyrle’s disease is unknown, but its association has been reported sparsely with renal disorders, uremic patients on dialysis, diabetes mellitus, liver disease and paraneoplastic syndromes, tuberculosis and some fungal diseases. We report Kyrle’s disease in a middle aged female suffering from diabetes mellitus with diabetic nephropathy on hemodialysis.

Introduction

Kyrle’s disease or hyperkeratosis follicularis et parafollicularis et cuten penetrance was first described in 1916 by J. Kyrle as hyperkeratotic verrucous papules and nodules in diabetic women.¹ The disease is most closely associated with renal failure and diabetes mellitus. The common pathophysiological principle in these disorders is transepidermal elimination of dermal substances.

Case Report

We report a case of Kyrle’s disease involving lower extremities (gluteal region, back of thighs and lateral aspect of both legs) for the last 1 month. The patient was a 50 year old diabetic female and was on irregular treatment since 5 years. She presented to us with breathlessness and generalized anasarca, her blood pressure was 170/90 mmHg and she had undergone hemodialysis 1 month back in view of chronic kidney disease.

She developed severe pruritic papulonodular lesions on her legs which progressed to involve both the thighs and the gluteal region. The patient had no family history of diabetes and no other member of family had similar complaints.

Physical examination revealed multiple well demarcated papular lesions of variable sizes (2-5 mm); lesions were asymmetrically distributed over gluteal region lateral side of legs and back of thighs. The lesions were papulonodular and hyper pigmented (purplish blue in colour) with central keratin plug (Figure 1). They were non tender and firm in consistency. Investigations (blood sugar, blood urea, serum creatinine, serum Na+, and K+, USG abdomen) were done and she was found to have developed diabetic nephropathy (CDk stage 5). Her HbA1C was 10.5% indicating very poor glycemic control. Viral markers (HIV, HBsAg, anti HCV) were negative.

Diagnosis of Kyrle’s disease was confirmed on punch biopsy which showed keratotic and partly parakeratotic plug invaginating the dermis.

Discussion

Kyrle’s disease is a rare skin disorder representing a subtype of perforating disorders dermatosis which involves transepithelial elimination of dermal structures. The perforating dermatosis has been classified on the basis of nature of substance eliminated.¹ Transepidermal elimination of necrobiotic basophilic collagen, eosinophilic elastic fibres and degenerated follicular contents with or without collagen or elastic fibres is seen in reactive perforating collagrosis, elastosis perforans serpiginosa and perforating follicularis respectively.¹ In Kyrle’s disease there is transdermal elimination of keratin with no collagen or elastic fibres.¹

Kyrle’s disease has been proposed to be genetically determined disease with onset during adulthood mostly during 30-50 years of age but cases as early as 5 years and as late as 75 years have been reported. The mode of inheritance is not clearly understood, it can be autosomal dominant or recessive.² A case of familial Kyrle’s disease has been reported in a 30 year old male with onset at 5 years of age with asymptomatic lesions on both upper and lower extremity, upper back, palms and soles. Involvement of cornea, conjunctiva, palms and soles along with dental anomalies was noted in all the affected family members in an autosomal dominant pattern.³

Although the exact etiology of Kyrle’s disease is unknown, it has been found to be associated with renal disorders, uremic patients on dialysis, diabetes mellitus, liver disease and paraneoplastic syndrome in multiple myeloma. It has also been associated with other conditions including tuberculosis, pulmonary aspergillosis, scabies, atopic dermatitis, AIDS, neurodermatitis and malignancy and endocrinial disorders.¹,³

Kyrle’s disease is characterized by hyperkeratotic papules and nodules with a central keratotic plug mostly located in lower limbs.² Pruritis may or may not be associated with the lesion. These lesions have marked propensity to involve calf and tibial region and posterior thigh. The exact pathogenesis of Kyrle’s disease is not clear. However it has been proposed that abnormal keratinization is responsible for the

Fig 1: Papulonodular and hyper-pigmented Kyrle’s lesion with central keratin plug on lateral and posterior aspect of thigh and gluteal region

¹Assistant Professor, ²Junior Resident, ³Professor, P.G. Department of Medicine, S.N. Medical College, Agra, Uttar Pradesh

Received: 13.06.2015; Revised: 19.08.2015; Accepted: 27.08.2015
Process, keratinization occurs faster than epidermal proliferation. It is suggested by Detmar et al that defective differentiation of the epidermis and dermoeipidermal junction owing to the altered glycosylation may be responsible. The elevated levels of extracellular matrix protein fibronectin may be responsible for the increased epidermal migration and proliferations, resulting in perforation. Kaiskou et al have suggested an infective etiology as they found improvement on oral clindamycin therapy in a patient.

Treatment of the underlying associated disease improves the lesion and prognosis becomes better. Other reported effective treatment options include cryotherapy, systemic and topical retinoids and narrow band UVB and keratolytics (salicylic acid and urea). Antimicrobial clindamycin 300 mg three times a day have also shown regression in lesions combination of oral retinoids and psoralen plus UVA have also been used. Surgery (CO2 laser surgery) is used as the last resort. Cessation of therapy usually leads to relapse of the lesion.

References