Nekam's disease

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ABSTRACT

Keratosis lichenoides chronica also known as Nekam's disease is a rare mucocutaneous disorder, characterized clinically by asymptomatic violaceous keratotic papules arranged in linear, reticular, or plaque form usually on the trunk and extremities and histologically by interface dermatitis. The disease is considered rare with only 128 cases being reported in the literature till date and very few from India. We report a case of a 40-year-old man who presented with constellation of features of lichen planus, seborrheic dermatitis, and apthous ulcers, which upon workup was found to be Nekam's disease.

Key words: Interface dermatitis, Keratosis lichenoids chronica, Nekam's disease

INTRODUCTION

Keratosis lichenoids chronica (KLC) was first described by Kaposi in 1895, who named the disease as lichen ruber acuminatus morbilliform disease.[1] In 1938, Nekam called the disease porokeratosis striate lichen, after seeing the acrosyringeal hyperkeratosis in the case published by Kaposi, despite the absence of coronoid lamella.[2] The widely used term keratosis lichenoides chronica was introduced by Margolis *et al*. in 1972.[3] The following report highlights the features of this rare disease along with a brief review of literature.

CASE REPORT

A 40-year-old man, farmer, presented with 4 months history of widespread asymptomatic scaly lesions, which initially appeared over flexor aspects of forearms and subsequently progressed to involve legs, thighs, and genitalia. He also complained of burning sensation in mouth on taking food and voluntarily disclosed dyspareunia. He did not receive any medication prior to, or after the onset of the lesions. There was no history of comorbidities or similar complaints in the family. The general and systemic examination was unremarkable. Cutaneous examination revealed multiple, discrete, well-defined, violaceous, hyperkeratotic, scaly, papules and plaques of sizes ranging from 0.2 × 0.1 cm to 2 × 1 cm over flexor aspects of both the forearms, extensor aspects of both legs, and inner aspects of thigh. Similar papules without scaling were present on the scrotum and few lesions over the forearms were arranged in a linear fashion [Figure 1]. Seborrheic dermatitis-like picture was present over face. Auspitz sign was negative. Multiple aphthous ulcers were noted on inner surface of both the lips and buccal mucosa. Erosions with violaceous margin were noted over inner aspect of prepuce [Figure 2]. Subungual hyperkeratosis of right thumb, onycholysis, and black discoloration of toe nails were present.

Differential diagnosis of hypertropic lichen planus, keratosis lichenoides chronica, lichen planus-psoriasis overlap were considered and investigated accordingly.

Routine blood tests were within normal range. Histopathology of the papule revealed parakeratosis, hyperkeratosis, alternate areas of acanthosis and atrophy, follicular plugging, vacuolar alteration of cells of dermoepidermal junction along with plasma cell infiltrates and dilated dermal capillaries confirming the diagnosis of KLC [Figures 3 and 4].

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.193923

Quick Response Code:

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Cite this article as: Aruna C, Ramamurthy D, Neelima T, Bandaru H. Nekam's disease. Indian Dermatol Online J 2016;7:520-2.

The patient was started on systemic steroids (prednisolone 30 mg) and dapsone 100 mg. After 8 weeks of follow up, there was only slight improvement. We planned to put him on acitretin but unfortunately the patient was lost to follow up.

DISCUSSION

KLC, also known as Nekam's disease, is a rare mucocutaneous skin disease of unknown etiology with 128 cases reported in the literature till date[4] and very few from India.

Clinically the disease is characterized by asymptomatic violaceous papules and nodules with reticular or linear arrangement, distributed symmetrically over trunk and extremities.[5] Additional features include rosacea or seborrheic dermatitis like rash of the face, recurrent aphthous-like ulcers, palmoplantar keratoderma, and ungual dystrophy in 75%, 50%, 40%, and 30% of cases, respectively.[1,6] In one case the disease mimicked Darier's disease.[7] Böer studied over 60 cases of KLC available in the literature and proposed clinical and histopathological criteria, which help in diagnosis of the disease.[8] A recent review by Paravina *et al*. concluded KLC as a chronic, progressive disease, easy to diagnose despite its variations in presentation but very difficult to treat.[4] Ruiz-Maldonado *et al*. compared 8 cases of pediatric onset KLC with that of adult onset and opined that the former represent a subset of the latter with special genetic and clinical characteristics such as facial erythematous and purpuric macules, eyebrow, and eyelash alopecia.[9]

Histopathologically KLC is characterized by vacuolar degeneration of keratinocytes at dermoepidermal junction,

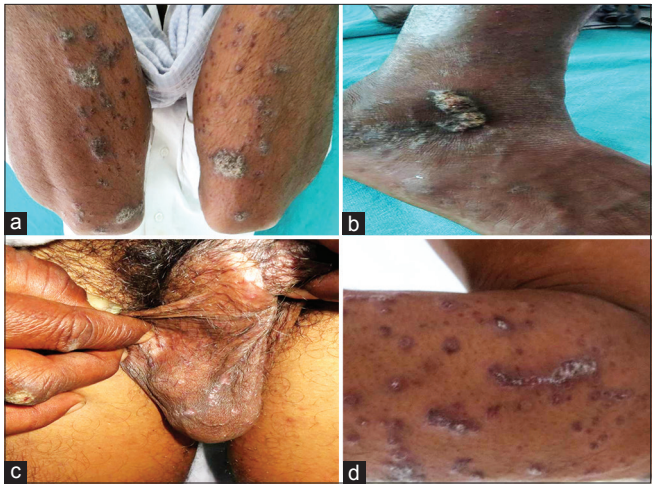

Figure 1: Multiple, violaceous, hyperkeratotic, scaly papules and plaques over extensor aspects of forearms (a), over lateral aspect of ankle (b), scrotum (c). Lesions over forearm arranged in a linear fashion (d)

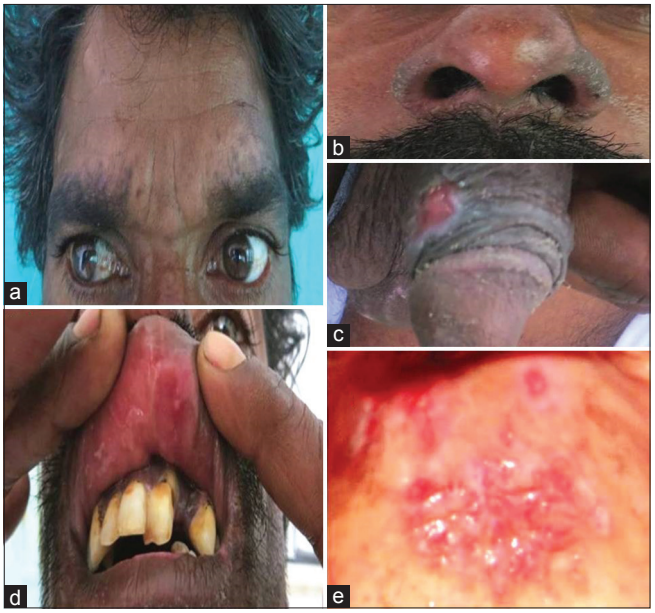

Figure 2: Seborrheic dermatitis like picture over medial aspect of eyebrows (a), ala nasi (b), erosions with violaceous margin over inner aspect of prepuce (c), apthous ulcers over inner aspect of upper lip (d), erosions over hard palate (e)

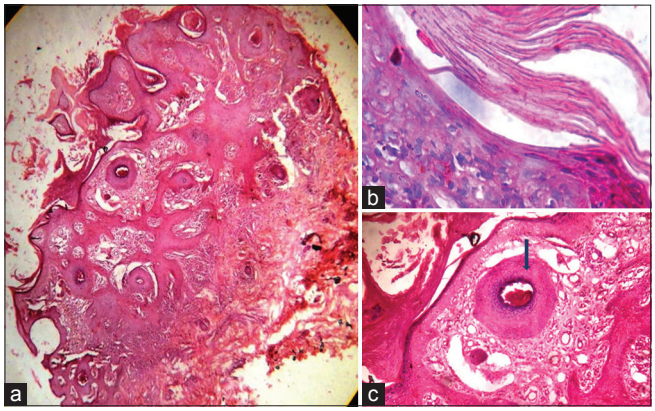

Figure 3: Whole mount of the skin bit showing alternate areas of acanthosis and atrophy, hyperkeratosis, inflammatory infiltrates, and follicular plugging (a) (H and E, ×40) (b). Close up view of parakeratosis and (c) follicular plugging (H and E, ×10)

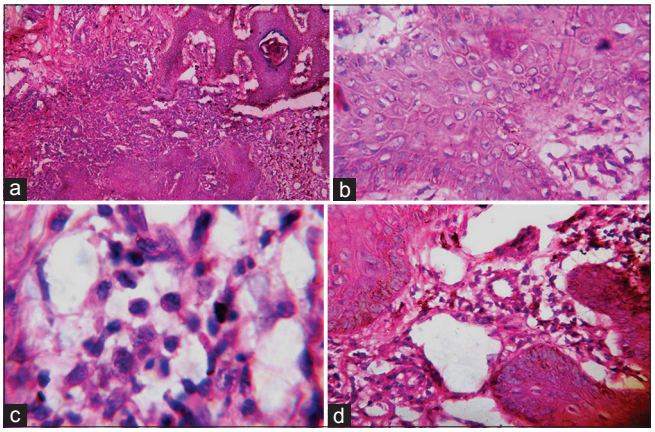

Figure 4: Inflammatory infiltrate obscuring dermoepidermal junction (a) (H and E, ×10). Areas of vacuolar alteration of cells (b) (H and E, ×40). Increased plasma cell infiltrate (c) (H and E, ×100). Dilated dermal capillaries (d) (H and E, ×40)

numerous necrotic keratinocytes at superficial and infundibular epidermis, chronic inflammatory infiltrate in the papillary dermis consisting of lymphocytes, histiocytes and plasma cells, and vascular dilatation.[8]

Some authors consider KLC as a variant of lichen planus that has evolved by antigen mimicry followed by epitope spreading, but now many consider the former as a distinct entity. Features that help to differentiate the two are highlighted in Table 1. KLC may sometimes be confused with Darier's disease, but presence of keratotic papules in linear/reticular arrangement and absence of acantholytic dyskeratosis, corp ronds, and acantholysis in the histopathology of the KLC delineates it from Darier's disease. Others such as lichen planus–psoriasis overlap, lichen planopilaris, and lupus erythematosus can mimic KLC and should be distinguished.

Topical agents are usually ineffective and systemic agents like steroids, dapsone, methotrexate, antimalarials, retinoids, and PUVA therapy can be tried. Although the disease is considered resistant to therapy, there are reports of KLC successfully treated with PUVA and efalizumab.[10,11]

Table 1: Clinical and histological differences between keratosis lichenoides chronica and lichen planus

Clinical features	Keratois lichenoides chronica	Lichen planus
Symmetry of lesions	+	–
Pruritus	–	+
Wickham striae	–	+
Response to therapy	Poor	Good
Histopathological features		
Parakeratosis	+	–
Follicular plugging	++	+
Hypergranulosis	–	++
Civatte bodies	–	+
Plasma cells	+	–
Dilated dermal blood vessels	+	–

+: Present, –: Absent, ++: Prominent, KLC: Keratosis lichenoides chronica

CONCLUSION

Nekam's disease is a rare disease. When a patient presents with constellation of features consistent with lichen planus, seborrheic dermatitis, and apthous ulcers, Nekam's disease should be considered and further evaluation of such cases can aid in detection of more number of cases.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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