



An Interesting and Rare Keratotic Disorder– Nekam Disease

Authors

Dr N.Devanand^{1*}, Dr M.S.Srinivasan²

¹Junior Resident, ²Head of the Department

Department of Dermatology, Venereology and Leprosy, Chettinad Hospital and Research Institute,
Kelambakkam

*Corresponding Author

Dr N. Devanand

Junior resident, Department of Dermatology, Venereology and Leprosy, Chettinad hospital And Research
Institute, Kelambakkam, India

Abstract

Keratosis lichenoides chronica describes rare lichenoid disorder involving limbs and trunks in a linear or reticulate pattern with relative symmetry. It is so rare that only 128 cases have been reported so far. The following report describes clinical features of this disease along with a short review of literature.

Keywords: Nekam disease, Keratosis lichenoides chronica.

Introduction

Keratosis lichenoides chronica was initially described in 1895 by Kaposi, and he named the disorder as lichen ruber acuminatus morbilliform disease^[1]. In 1938, Nekam called this disease as porokeratosis striate lichen, despite the absence of coronoid lamella^[2]. The most commonly used term keratosis lichenoides chronica was introduced by Margolis *et al.* in 1972^[3]. Keratosis lichenoides chronica describes rare lichenoid disorder involving limbs and trunks in a linear or reticulate pattern. It can be seen in all age groups although age group between 20 to 40 years were commonly affected. It had a slight male preponderance with male to female ratio of 1.7: 1^[4]. The cause of this disorder remains unknown. It was previously thought to be a variant of lichen planus. The mutation in NLRP1 gene which leads to release of IL1 leading to secretion of Keratinocyte growth factor and

Tumour necrosis factor alpha, resulting in epidermal hyperplasia and keratosis^[5]. It clinically manifests as violaceous hyperpigmented scaly papules or small nodules involving trunk and extremities. 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^[6].

Case Report

- A 46 years old female presented with complaints of itchy hyperpigmented oozy lesions over both legs and hands for past 4 years, itching and oozing increased in intensity for past 2 months.
- H/O previous treatment with steroids before 1 month but showed no improvement.

- No history suggestive of contact with any irritants.
- On examination: Multiple symmetrical hyperpigmented lichenified oozy papules and plaques of varying sizes over extensor aspect of bilateral forearms, hands and dorsal aspect of bilateral feet.
- Mucosa, Nails and Genitals are normal



Fig-1: Lesions over extensor aspect of hands



Fig-2: Lesions involving extensor aspect of feet

- Differential diagnoses considered were Lichen Planus, Contact dermatitis, Photo lichenoid dermatitis, chronic eczema and Pellagra.
- Investigation such as Complete blood count, Random blood sugar, Liver, renal and thyroid function tests, HbSAg, HCV, HIV and RPR were done and were normal
- Histopathological examination revealed
 - Focal Parakeratosis with Spongiosis
 - Focal lymphocytic exocytosis
 - Dermis shows Mild edema Perivascular and Periadnexal lymphocytic infiltrate

- Features confirmed **Keratosi s Lichenoides Chronica**

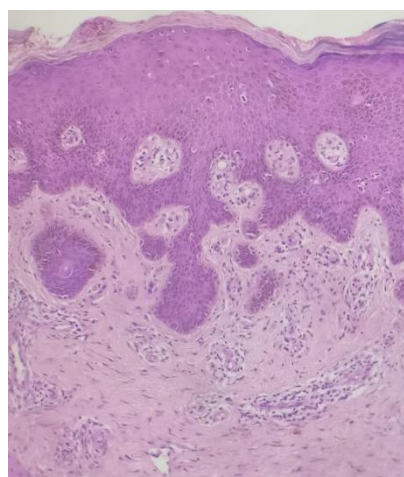


Fig 3: Histopathology showing parakeratosis, lymphocyte exocytosis, perivascular lymphocytic infiltrate.

- Patient was treated with oral dapsone 100mg during night time and oral prednisolone 30mg during morning time. Topically bland emollients like Liquid paraffin and topical mometasone furoate cream was given.

Conclusion

Keratosi s lichenoides chronica is a rare mucocutaneous disorder of unknown etiology. It is usually asymptomatic but itching can be seen in 20 % of cases. It can mimic hypertrophic lichen planus, Hypertrophic chronic cutaneous lupus erythematosus and chronic eczemas. Histologically it shows hyperkeratosis, Acanthosis, lichenoid band like infiltrate in basal layer, mixed infiltrate of lymphocytes, plasma cells and histiocytes [7]. It is usually very resistant to treatment. 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 [8][9].

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