

Keratosis lichenoides chronica: case report

Sunil Kumar Gupta, Monika, Deepika

Department of Dermatology, Venereology, Leprology, Dayanand Medical College & Hospital, Ludhiana

Abstract Keratosis lichenoides chronica (KLC) or Nekam's disease is a controversial rare dermatosis of unknown etiology. It is characterized by symmetrically arranged lichenoid linear and reticulate scaly plaques and hyperkeratotic papules most marked on extremities and buttocks and accompanied by facial lesions resembling seborrheic dermatitis. It usually affects adults between 20 to 40 years but children are affected occasionally. We report a case of middle-aged female showing chronic lichenoid plaques in a characteristic linear and reticulate fashion over buttocks and thighs with minimal pruritus, resistant to conventional treatment, and associated seborrheic dermatitis like eruption on face. KLC is a chronic and progressive disorder extending over many years and is very resistant to therapeutic approaches. Despite being a rare disorder, it is important to be familiar with KLC, which can be easily confused with Koebner's phenomena of lichen planus.

Key words

Keratosis lichenoides chronica.

Introduction

Keratosis lichenoides chronica (KLC), also known as Nekam's disease, is a rare dermatological disorder of keratinization characterized by development of violaceous, hyperkeratotic papules and nodules typically arranged in a linear or reticulate pattern, mostly localized on the extremities and buttocks and accompanied by facial lesions resembling seborrheic dermatitis.¹ It usually affects adults between 20 to 40 years but children are affected occasionally.² It has been a subject of controversy whether it is a distinct entity or represents an unusual variant of lichen planus. It was first described by Kaposi in 1985 and was given the name *Lichen ruber acuminatus morbilliform disease*.³ The current name *Keratosis lichenoides chronica* was given by Margolis *et al.* in 1972.⁴ We report a case of

keratosis lichenoides chronica in a middle aged female.

Case Report

A 40-year-old female, housewife, married, resident of Punjab, consulted for two year history of mildly pruritic, lichenoid eruption over lower back, buttocks and thighs. Patient also had associated erythematous scaly plaques over face. Palms and soles were spared, as were mucous membranes, hair and nails. The lesions had first appeared on lower back and had progressively spread over buttocks and thighs. There was only partial response to unspecified topical treatment taken over two years. She reported no co-morbidities or similar complaints in her family. Physical examination revealed lichenoid papules and nodules coalescing to form linear and reticular plaques located over lumbosacral area, buttocks, and posterior thighs (**Figure 1**). Few lesions were erythematous papules with a central hyperkeratotic plug as can be seen in **Figure 2**. Erythema and scaling affected the centropalpebral area, with a seborrheic

Address for correspondence

Prof. Dr. Sunil Kumar Gupta
Department of Dermatology, Venereology,
Leprology
Dayanand Medical College & Hospital,
Ludhiana, Punjab, India
Email: vsunilgupta@rediffmail.com



Figure 1 Lichenoid papules and nodules coalescing to form linear and reticular plaques.



Figure 2 erythematous papules with a central hyperkeratotic plug.

dermatitis like appearance. Hematological and biochemical investigations did not reveal any abnormality. Skin biopsy was planned but was not done due to financial constraints. Patient was started on PUVA-sol and topical retinoids but was unfortunately lost to follow up.

Discussion

Keratosi s lichenoides chronica (KLC) or Nekam's disease is controversial rare dermatoses of unknown etiology. The varied nomenclature of this disease includes *Lichen ruber acuminatus verrucosus et reticularis*, *porokeratosis striata lichenoides*, *lichen ruber moniliformis*.⁵ Kaposi first described this at the end of 19th century and the current terminology *Keratosi s lichenoides chronica* was proposed by Margolis in 1972.¹ There is ongoing discussion

about its independent nature. It commonly affects 20 to 40 years of age with slight male predominance (1.35:1).⁶ It is characterized by symmetrically arranged lichenoid linear and reticulate lesions, scaly plaques and hyperkeratotic papules most marked on extremities and buttocks. Individual lesions may show rough hyperkeratotic papules with a central plug that can only be removed with difficulty.⁷ In addition to the above characteristic features there can also be a seborrheic dermatoses like eruption over face in 75% of the cases.¹ Oral manifestations like recurrent aphthous ulcers, large chronic ulcers or erythrokeratotic papules can occur in 50% of patients.² There can be associated palmoplantar hyperkeratosis, alopecia, eye involvement, and even epiglottic infiltration.⁶ The nail involvement is seen in 30% of cases in the form of thickening and hyperkeratosis of nail bed and yellowish pigmentation of nails.³ A possible association of Nekam's disease with glomerulonephritis and lymphoproliferative disorders has also been reported.⁵ However, in our case we did not find any associated pathology.

Two distinct variants of keratosi s lichenoides chronica have been described. The pediatric variant shows characteristically erythematous - purpuric macules over face with alopecia and lesser involvement of nails, mucosae and eyes.⁸ The literature also describes a vascular variant of KLC in which reticulate telangiectasias are present in addition to the usual findings.⁹

The differential diagnosis usually includes lichen planus, pityriasis lichenoides et varioliformis acuta (PLEVA), nodular prurigo, reactive perforating collagenosis, and connective tissue diseases such as lupus erythematosus. The main differential diagnosis in our case was hypertrophic lichen planus with koebnerisation.

Histologically, it is characterized by lichenoid dermatitis with numerous necrotic keratinocytes and parakeratosis housing neutrophils in a staggered fashion, irregular acanthosis and corneal plug. The lichenoid infiltrate is often peri-infundibular or around the acrosyringial areas.³

KLC is chronic and progressive disorder extending over many years and is very resistant to therapeutic approaches. Topical treatments are usually ineffective, as well as, systemic steroids, methotrexate, antimalarials, and cyclosporine. Favourable responses to photochemotherapy with or without acitretin, etretinate, photodynamic therapy and efalizumab have been reported but further studies are called for regarding these therapies. Spontaneous remission of lesions is very uncommon.^{10,11}

We made the diagnosis of KLC on the basis of presentation in a middle-aged adult showing chronic lichenoid plaques in a characteristic linear and reticulate fashion over buttocks and thighs with minimal pruritus, resistant to conventional treatment, and associated seborrheic dermatitis like eruption on face. Despite being a rare disorder, it is important to be familiar with KLC, which can be easily confused with Koebner's phenomena of lichen planus. Rarity of this disorder propelled this discussion.

References

1. Ghislain PD, De Bier A, Creusy C, Modiano P. Keratosis lichenoides chronica: report of a new case, with success of PUVA therapy. *Dermatol Online J.* 2001;7:4-8.
2. Breathnach SM. Lichen planus and lichenoid disorders. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology. 8th edn. Vol 2.* Oxford: Wiley-Blackwell; 2010. p. 41.1-28.
3. Martins LCG, Horne M, Moreira DN, Follador I, Almeida VRP. Keratosis lichenoides chronica- case report. *An Bras Dermatol.* 2011;86:S148-51.
4. Adiflen E, Erdem O, Celipci S, Gurer MA. Easy to diagnose, difficult to treat: keratosis lichenoides chronica. *Clin Exp Dermatol.* 2008;35:47-50.
5. Masouye I, Saurat JH. Keratosis lichenoides chronica: the centenary of another Kaposi's disease. *Dermatology.* 1995;191:188-92.
6. Wozniacka A, Schwartz RA, Omulecki A, Lesiak A, Sysa-Jedrzejowska A. Keratosis lichenoides chronica: a diagnostic and therapeutic challenge. *Clin Exp Dermatol.* 2006;31:48-50.
7. Margolis MH, Cooper GA, Johnson SA. Keratosis lichenoides chronica. *Arch Dermatol.* 1984;120:1471-4.
8. Ruiz-Maldonado R, Duran-McKinster C, Orozco-Covarrubias L, Saez-De-Oscariz M, Palacios-Lopez C. Keratosis lichenoides chronica in pediatric patients: a different disease? *J Am Acad Dermatol.* 2007;56:S1-5.
9. Nijsten T, Mentens G, Lambert J. Vascular variant of Keratosis lichenoides chronica associated with hypothyroidism and response to tacalcitol and acitretin. *Acta Derm Venereol.* 2002;82:128-30.
10. Demirci E, Boyvat A, Arica IE, Kocyigit P, Ozdemir E, Heper AO. Keratosis lichenoides chronica: marked response to PUVA in combination with acitretin. *Acta Derm Venereol.* 2005;85:552-3.
11. Muñoz-Santos C, Yébenes M, Román J, Luelmo J. Response of keratosis lichenoides chronica to efalizumab therapy. *Arch Dermatol.* 2009;145:867-9.