Dermatology Practical & Conceptual

Keratosis Lichenoides Chronica (Nekam's Disease): Dermoscopic and in Vivo Reflectance Confocal Microscopy Findings

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Introduction

Keratosis lichenoides chronica (KLC), also known as Nekam disease, is a rare chronic inflammatory skin disorder characterized by asymptomatic hyperkeratotic papules in a linear or reticular pattern on the trunk and extremities, along with a facial rash resembling seborrheic dermatitis and oral ulcers [1]. Histopathology reveals lichenoid dermatitis features, and the disease follows a chronic, progressive course with challenging treatment [2]. This report presents a KLC case, highlighting dermoscopy and reflectance confocal microscopy (RCM) findings.

Case Presentation

A 39-year-old man presented with 3 years of reticular lichenoid lesions on the trunk and extremities, along with erythroderma (Figure 1). For the management of idiopathic pulmonary fibrosis, he was prescribed 100 mg of azathioprine and 4 mg of prednisolone. There was no family history of similar eruption, nor any skin disorders diagnosed.

A physical examination revealed erythroderma and generalized pink-brown infiltrated papules with reticular coalescence on the face, trunk, and extremities. Oral mucosa and nails were unaffected. Dermoscopy displayed globular Wickham striae, dotted and linear vessels, diffuse brown-gray granules, and perifollicular granules and dots (Figure 2B) [3].

RCM demonstrated a regular honeycomb pattern on the epidermal level, dense inflammatory infiltration featuring dendritic cells, and single-cell necrosis on basal layers. The irregular dermoepidermal junction (DEJ) and upper dermis displayed a sheet-like cell distribution, including scattered small bright round spots correlating lymphocytes, dendritic

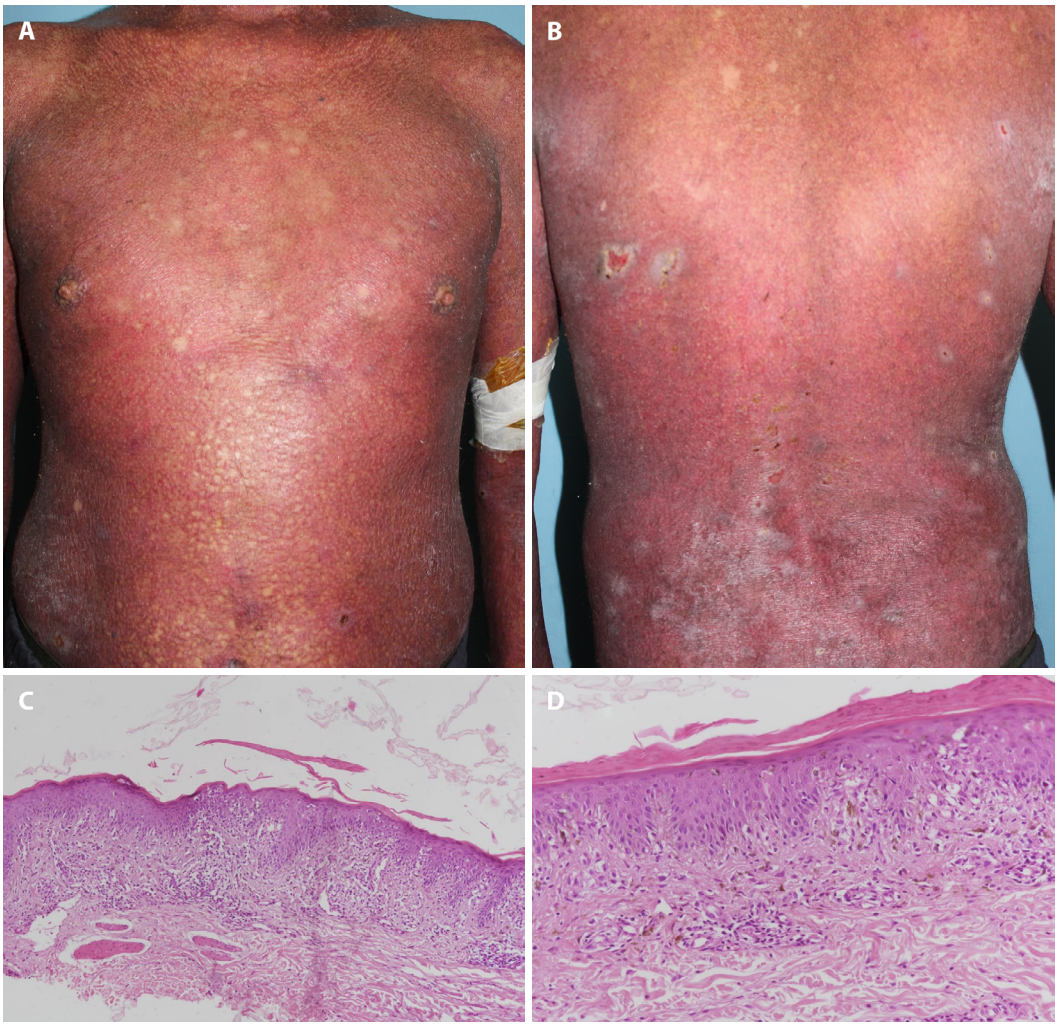

Figure 1. (A, B) Generalized lichenoid papules exhibiting a reticular coalescence resulting in erythroderma. (C, D) A lichenoid infiltration with parakeratosis, basal vacuolar degeneration, lymphoid exocytosis, and dermal inflammatory cell infiltrations with melanophages on histopathology (H&E x100, x200).

cells, single or aggregated plump bright triangular inflammatory cells corresponding to melanophages, satellite necrosis, vacuolization, and dilated vessels in the upper dermis (Figure 2, C and D).

Histopathology revealed hypergranulosis, epidermal basal necrotic keratinocytes, and basal lymphoid infiltration (Figure 1, C and D). Superficial dermis exhibited a dense band of mononuclear cell infiltration with some melanophages and dilated vessels. Immunofluorescence results were negative, while immunohistochemistry showed positivity for CD3, CD8, CD7, and CD4. Laboratory evaluation was unremarkable.

The patient was diagnosed with Nekam disease with clinicopathologic correlation. Treatment involved systemic prednisolone at 0.5 mg/kg combined with acitretin at 10 mg. Prednisolone dosage was gradually reduced while increasing acitretin therapy to 35 mg daily. The patient displayed a remarkable response to acitretin therapy, with no recurrence in two years, continuing acitretin as part of ongoing treatment.

Conclusion

Dermoscopy and RCM are established in vivo diagnostic techniques with notable significance, obviating the requirement for histopathologic examination in lesion diagnosis. The epidermis, DEJ, and upper dermal morphological features can be analyzed using RCM [4]. RCM findings of KLC have not been described in the literature.

The RCM findings of the patient correlated with the histopathologic examination. Bright round spots at the DEJ

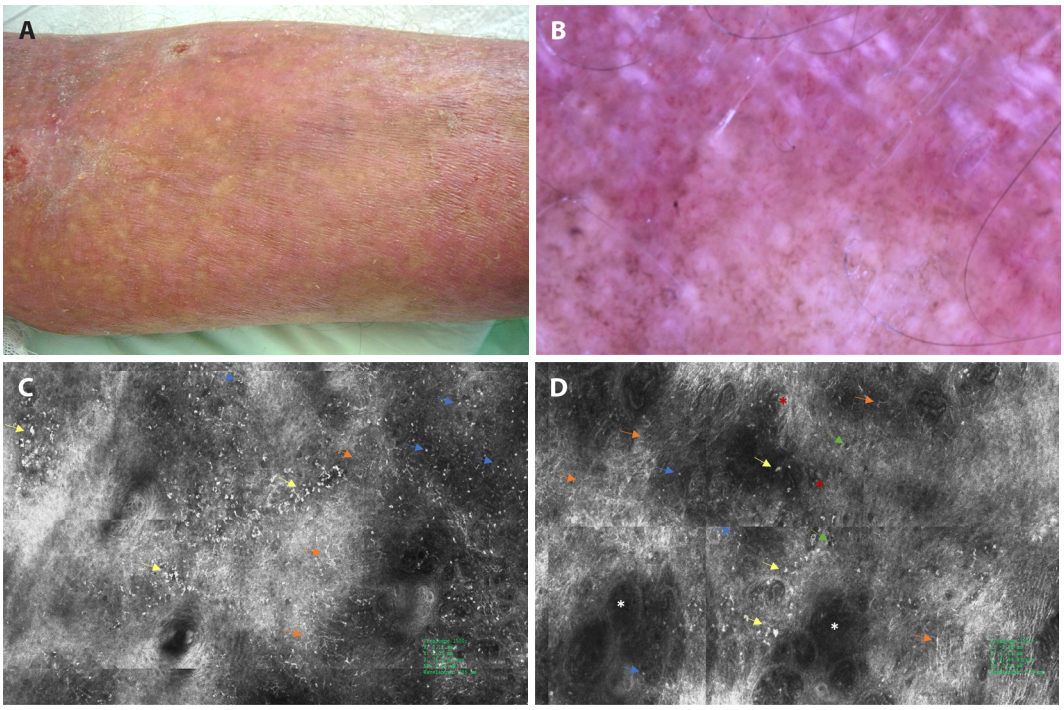

Figure 2. (A) Reticular erythematous lesions on the left forearm (dermoscopy and RCM imaging area). (B) Thick whitish lines, dotted and linear vessels, and brown-gray irregular and perifollicular granules and dots on dermoscopy. (C) RCM image revealing a sheet-like distribution of cells at the irregular DEJ, featuring scattered small, bright, round spots corresponding to lymphocytes (blue arrows), dendritic cells (orange arrows), and single or aggregated, plump, bright, triangular inflammatory cells correlating with melanophages (yellow arrows). (D) An RCM image showing sheets of inflammatory cells in the upper dermis and obscuring the DEJ, containing small, bright, round spots corresponding to lymphocytes (blue arrows), satellite necrosis (necrotic keratinocyte adjacent to a lymphocyte, red asterisk), dendritic cells (orange arrows), plump, bright inflammatory cells correlating with melanophages (yellow arrows), vacuolization (green arrows), and dilated vessels (white asterisk).

were associated with dense inflammatory infiltration involving dendritic cells. In certain areas, single-cell necrosis characteristic of Civatte bodies in lichen was observed. Irregularities at the DEJ and dilated vessels were associated with interface dermatitis. The plump-bright cells present at the DEJ and in the superficial dermis were indicative of melanophages.

This case report illustrates the dermoscopy and RCM features of KLC with histopathological correlation, emphasizing their diagnostic value. Combined with clinical findings, RCM and dermoscopy features can enhance KLC diagnosis.

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