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CASE REPORT

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Childhood-onset keratosis lichenoides chronica: A case report

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Abstract

Keratosis lichenoides chronica (KLC) is a rare dermatosis which represents different clinical characteristics between adult- and pediatric-onset cases. We described a childhood case of KLC with features typical for adult-onset disease. Acitretin led to partial improvement of her skin, but not mucosal, lesions.

KEYWORDS

alopecia, keratosis lichenoides chronica, nail dystrophy, rosacea, seborrheic dermatitis

1 | INTRODUCTION

Keratosis lichenoides chronica (KLC) is a rare dermatosis with a chronic progressive course. Characteristically, KLC presents as violaceous, keratotic, lichenoid papules with a linear or reticulated arrangement and symmetrical distribution on the extremities and the trunk.[1] Another prominent feature is a seborrheic dermatitis- or a rosacea-like eruption on the face.[2]

Although KLC manifests more commonly in adults (20-40 years), there are some reports of the disease appearing in childhood.[1,2] It has been suggested that the disease represents different clinical characteristics between adults and children.[3] We describe a patient with the manifestations of adult-onset KLC beginning from infancy.

2 | CASE REPORT

A 20-year-old female patient was referred to our clinic with a generalized pruritic erythematous-violaceous eruption. The lesions had appeared during the first year of life on her chest and gradually progressed to the other parts. Repeated courses of topical corticosteroids had been administered without any significant improvement. Her past medical and family history was unremarkable. Examination revealed keratotic violaceous papules arranged in a reticular pattern with symmetrical distribution over the extremities and the trunk (Figure 1). The lesions were more confluent on the lateral trunk, breasts, buttocks, and extremities.

There was an erythematous rosacea-like eruption on her face. The neck was involved circumferentially, and the

Soheila Nasiri and Azadeh Rakhshan have equal contribution.

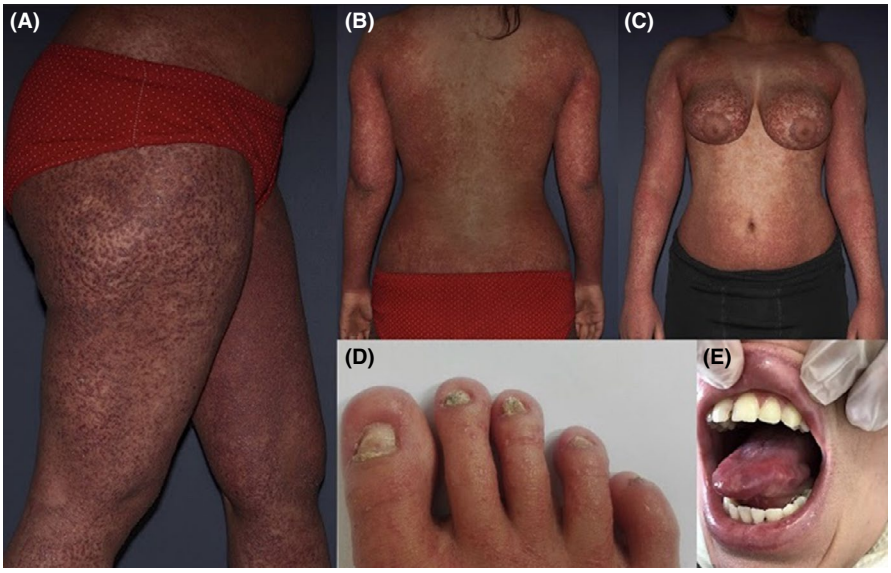

FIGURE 1 Erythematous, scaly, keratotic papules in a reticulated pattern on the (A) buttocks and lower extremity, (B) posterior trunk, (C) and anterior trunk pronounced on the breasts; (D) Dystrophic toe nails; (E) erosive lesions on the tongue

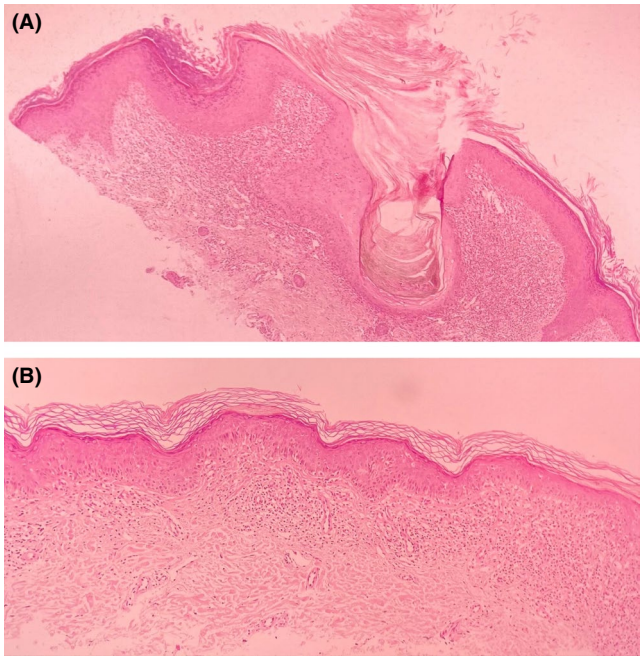

FIGURE 2 (A,B) Variable epidermal thickening with areas of acanthosis and atrophy, hyperkeratosis, focal parakeratosis with remnants of neutrophils, lichenoid interface reaction with band-like subepidermal infiltration of lymphocytes and a few plasma cells. (×4 objective)

scalp was scaly, but scalp hair, eyebrows, and eyelashes had a normal thickness and density. The nails were dystrophic and greatly thickened, and there were many keratotic papules on the palmoplantar surfaces. She had tender erosions along with soreness on her tongue. Examination of the other mucosae including genitalia and conjunctiva was insignificant. There were no signs of internal organ involvement or lymphadenopathy.

Routine laboratory data including complete blood count, ESR, CRP, liver, and renal function tests were normal.

We performed skin biopsy with differential diagnoses of KLC, psoriasis, hypertrophic lichen planus, and pityriasis rubra pilaris. Histologic examination revealed variable epidermal thickening with areas of acanthosis and atrophy, hyperkeratosis, focal parakeratosis with remnants of neutrophils, lichenoid interface reaction with band-like subepidermal infiltration of lymphocytes and a few plasma cells which were compatible with keratosis lichenoides chronica (Figure 2).

The diagnosis of KLC was considered based on typical clinical and histopathologic findings, and the patient was started in acitretin 25 mg/daily. This led to the gradual improvement of the skin lesions with prominent papular flattening and reduction of erythema in 3 months, but her oral lesions did not demonstrate any significant improvement.

3 | DISCUSSION

Keratosis lichenoides chronica is considered by some as a variant of lichen planus, while others believe that it has a distinct clinical and histological picture.[4] The combination of lichenoid keratotic papules with a characteristic linear or reticular arrangement and an erythematous facial eruption refers to the clinical diagnosis of KLC. Oral lesions occur in 50% of patients. Additional features that may occur less frequently include eye and genital involvement, and palmoplantar kertoderma. The other less common features of KLC are summarized in Table 1. The histologic features of KLC are variable and nonspecific; however, the presence of parakeratosis and heavier infiltration than what is usually seen in lichen planus may help in differentiation.[4]

Although children are occasionally affected, the majority of reported KLC cases are adults. In 2007, Ruiz-Maldonado et al studied 14 cases of pediatric-onset KLC and compared

TABLE 1 Additional less frequent features of KLC

Yellow discoloration of the nails[5]
Nail dystrophy[6]
Nodular infiltration of the epiglottis, which may cause hoarseness[7]
Vascular lesions[8]
Sclerodactyly[9]

them with adult-onset KLC patients.[3] They proposed that some features of KLC might characterize pediatric-onset KLC in comparison with adult-onset KLC including an early or congenital onset, a positive family history with a probable autosomal recessive inheritance, a greater proportion of male to female, the primary location of the lesions on the face as erythematous-purpuric macules, forehead, eyebrow, and eyelash alopecia, the higher frequency of pruritus, and a much lower frequency of mucosal involvement and systemic abnormalities.[3]

Our patient began to develop skin lesions during the first year of her life making her a case of pediatric-onset KLC but she presented with features more consistent with those of adult-onset KLC. First, the lesions had appeared on the chest instead of the face. Indeed, her facial rosacea-like lesions were a recent phenomenon. Second, extensive oral erosions and prominent nail involvement in our case were other dermatological alterations occurring infrequently in children but seen in 50% and 30% of adult-onset patients, respectively.[3] Negative family history and lack of alopecia were also in favor of adult-onset disease. In general, the presence of adult-like KLC manifestations in this patient, which started in infancy, along with the accumulation of rare symptoms of the disease such as palmoplantar keratotic papules, dystrophic nails, and oral erosions made this case memorable.

Keratosis lichenoides chronica has a chronic course with a gradual progression, and most cases have failed to show a favorable response to any treatment with a mean follow-up time of 14 years in adults.[3] Several anecdotal reports have shown the efficacy of ultraviolet A- and ultraviolet B-light phototherapies, natural light,

photochemotherapy, and oral retinoids (acitretin or isotretinoin) plus phototherapy.[3] In our case, the patient showed a partial response to the treatment with acitretin but the phototherapy was not feasible due to the COVID-19 outbreak. Based on the current data systemic corticosteroids, antimalarial agents, sulfones, gold, and cyclosporine are proven to be ineffective in the treatment of KLC.[4]

4 | CONCLUSION

This case was a pediatric-onset KLC patient that was remained undiagnosed for several years. It highlights the importance of dermatologists' vigilance in the early detection of pediatric-onset KLC cases.

5 | ETHICAL CONSIDERATIONS

The patient in this manuscript has given written informed consent to the publication of her case details.

ACKNOWLEDGMENT

None.

CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

Soheila Nasiri: involved in clinical evaluation and management of the patient, writing the manuscript, and supervision of the project. *Azadeh Rakhshan:* involved in histopathology evaluation of the patient and writing the related part in the manuscript. *Khatere Zahedi:* made contribution to data gathering and writing the manuscript in consultation with Mehdi Gheisari and Sahar Dadkhahfar. *Sahar Dadkhahfar* and *Mehdi Gheisari*: involved in taking photographs and writing and editing of the final draft of the manuscript.

DATA AVAILABILITY STATEMENT

The data are available from the corresponding author upon reasonable request.

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