

A case of pediatric keratosis lichenoides chronica with unusual presentation of severe oral erosions



Maryam Nasimi, MD,^a Arghavan Azizpour, MD,^a Vahideh Lajevardi, MD,^a
Azita Nikoo, MD,^b and Ifa Etesami, MD, MPH^a
Tebzan, Iran

Key words: keratosis lichenoides chronica; oral lesions; pediatric onset.

BACKGROUND

Keratosis lichenoides chronica (KLC) is a rare, chronic disorder of keratinization that typically affects adults.¹ The adult-onset disease commonly presents with linear or reticular keratotic papules and plaques over the trunk and extremities, rosacea, or seborrheic dermatitis–like rash on the face and oral ulcers. Pediatric-onset KLC is rare with different clinical characteristics.² Pediatric cases initially present with purpuric macules on the face, more commonly with alopecia of the forehead and eyebrows and less commonly affecting oral mucosa.³ We report a case of KLC starting in early childhood with extensive oral erosions.

CASE PRESENTATION

A 10-year-old girl presented to our clinic with erythematous, scaly, keratotic plaques over her extremities arranged in a reticulated pattern. The linear and reticulated plaques did not follow Blaschko lines (Fig 1). The plaques were slightly pruritic. She also complained of painful oral lesions that caused her discomfort during eating. On physical examination she had extensive oral erosions, large ulcers on her tongue and labial mucosa, and scales on her lips (Fig 2). Her disease started when she was 3 years old with a progressive course. Her nails were normal. Her facial skin was spared. The physical examination findings were otherwise normal. The routine laboratory data were normal. Based on the clinical findings, KLC was considered. Her medical history was otherwise normal, and her family history was negative for KLC. Two biopsies were done: one from her oral

Abbreviation used:

KLC: keratosis lichenoides chronica

lesions and the other from keratotic plaques on her extremities. Histopathology of cutaneous lesions found acanthosis, hyperkeratosis, and lichenoid reaction with many dyskeratotic cells in the epidermis compatible with KLC (Fig 3). Histopathology of oral lesions showed acanthosis and lichenoid reaction associated with telangiectasia and edema.

We started acitretin at a dose of 10 mg/d. After 2 months of retinoid therapy, the keratotic plaques on her extremities flattened, and the erythema was reduced, but her oral lesion did not show improvement.

DISCUSSION

KLC is a rare disease with a chronic course that mainly affects adults.¹ In adults, KLC is characterized by erythematous hyperkeratotic papules and plaques arranged in linear or reticulated patterns with preferred distribution on limbs. Most of the patients have seborrheic dermatitis or psoriasiform facial lesions. KLC affects oral mucosa as ulcers or infiltrating papules, and ocular involvement such as blepharitis and keratoconjunctivitis have been reported.² Pediatric KLC is rare and has different characteristics than those of adult-onset KLC. Pediatric KLC starts during early childhood or even congenitally with erythematous and purpuric macules of the face early during the course of the

From the Departments of Dermatology^a and Pathology,^b Razi Hospital, Tehran University of Medical Sciences.

Funding sources: None.

Conflicts of interest: None declared.

Correspondence to: Ifa Etesami, MD, MPH, Razi Hospital, Vahdat-eslami square, Tehran, Iran 1199663911. E-mail: ifa.etesami@gmail.com.

JAAD Case Reports 2018;4:267-9.
2352-5126

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<https://doi.org/10.1016/j.jcdr.2017.11.015>



Fig 1. Erythematous, scaly, keratotic plaques on the extremities arranged in a reticulated pattern. **A**, Lower extremity. **B**, Upper extremity.



Fig 2. Oral lesions.

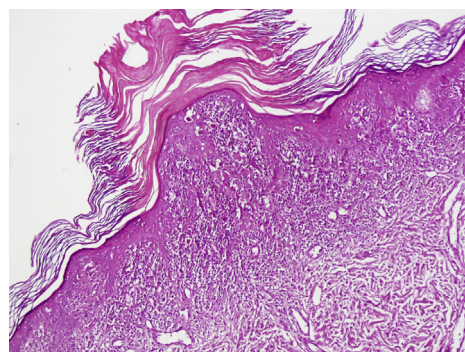


Fig 3. Acanthosis, hyperkeratosis, and lichenoid reaction with many dyskeratotic cells in the epidermis. (Hematoxylin-eosin stain; original magnification: $\times 100$.)

disease.⁴ In 2007 Ruiz-Maldonado et al³ studied 14 cases of pediatric-onset KLC and compared those patients with 40 reported adult-onset KLC patients.³ They provided differentiating features between adult-onset and pediatric-onset KLC (Table 1). The investigators found that pediatric cases are mostly familial and present with macular purpuric lesions on their faces and alopecia of eyelashes and eyebrows, which are not common in adults.³ Oral lesions are less frequently reported in pediatric cases than adult cases (7% vs 50%, respectively). These authors concluded that pediatric KLC may present as a different disease with special genetic and clinical features.

KLC can be associated with oral manifestations in 50% of cases, mostly presenting with recurrent aphthous oral lesions.⁵ Oral lesions are more resistant to therapy, and previous studies found partial response to retinoids (acitretin) and dapsone.^{5,6} One patient with KLC and oral aphthous ulcers was reported with partial response to dapsone and acitretin. The mucosal lesions relapsed after

discontinuation of dapsone. Our patient did not show significant improvement in her oral erosions after 2 months of receiving acitretin, and because our patient lived a long distance with poor access to medical care, dapsone was not started.

KLC has a chronic and progressive course that is difficult to treat. Phototherapy and oral retinoids are reported as successful therapies.^{5,7}

CONCLUSION

Our case is unique because as a case of pediatric-onset KLC, our patient presented with features that are more similar to those of adult-onset KLC. She had severe and extensive oral erosions that are unusual for pediatric KLC cases. Our patient had no family history for KLC despite the autosomal-recessive inheritance suggested for pediatric KLC.⁴ Our patient did not show alopecia of eyebrows and eyelashes and lacked macular or purpuric macules that are frequently seen in children with KLC. Her keratotic linear and reticulated lesions on the limbs resolved in

Table I. Comparing adult-onset with pediatric-onset keratosis lichenoides chronica

	Adult-onset KLC	Pediatric-onset KLC
Gender ratio M:F	1.35:1	2.16:1
Age at onset	28.5 y	7.4 mo
Congenital cases	0	21%
Familial cases	0	57%
Inheritance	None	Autosomal recessive
Initial location of lesions	Limbs	Face
Type of facial lesions	Seborrheic dermatitis–like, psoriasiform	Erythematous purpuric
Alopecia of forehead, eyebrows, eyelashes	0	29%
Pruritus	23%	43%
Oral lesions	50%	7%
Genital lesions	35%	0%
Nail lesions	30%	0%
Lichenoid	87%	90%

Data from Ruiz-Maldonado et al.³

response to acitretin administration, but her oral lesions did not improve by acitretin, highlighting the beneficial effects of retinoids on cutaneous lesions without successful influence on mucosal lesions.

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