Keratosis Lichenoides Chronica: A Case Report

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Keratosis lichenoides chronica (KLC) is a rare chronic hyperkeratotic disorder that typically affects patients aged 20 to 50 years. Its distinct clinical presentation in the pediatric population has raised speculation that the adult and pediatric variants of this disorder may be entirely separate disease entities. We present a case of adult-type KLC manifesting during childhood in a 14-year-old adolescent girl. We also review the literature on this rare disorder.

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Keratosis lichenoides chronica (KLC) is a rare papulosquamous hyperkeratotic disorder that usually presents in patients aged 20 to 50 years and is uncommon in the pediatric population. In adults, it manifests as violaceous, keratotic, lichenoid papules or patches arranged in a reticulate or linear pattern, usually on the extremities, trunk, abdomen, and lower back. By contrast, pediatric cases of KLC have involved a congenital or early childhood eruption of lichenoid papules or plaques on the face, especially on the cheeks, accompanied by alopecia. When present, facial lesions in adults have been reported to be seborrheic dermatitis–like. Thus, it has been proposed that childhood-onset KLC might be a different disease altogether.[1] We present a patient with adult-pattern KLC that began in childhood.

Case Report

A 14-year-old adolescent girl was referred to the Department of Dermatology at the University of Virginia, Charlottesville, for evaluation of an asymptomatic eruption that had been present for several months. She was otherwise in excellent health and there was no personal or family history of skin disease.

On physical examination, the patient appeared healthy. She had keratotic, erythematous, 2- to 4-mm lichenoid papules on the distal arms (Figure 1A), linear lesions on the antecubital region as well as popliteal fossa, and punctate keratotic papules on the soles and dorsal feet (Figure 1B). Her palms were clear. There was a seborrheic dermatitis–like eruption on the scalp.

A clinical diagnosis of KLC was made and biopsies were performed. The biopsies were obtained from lesions on the left posterior arm and left forearm. The findings were similar in the 2 specimens and included focal parakeratosis, vacuolar alteration of the basilar layer with Civatte body formation, telangiectasia, and a patchy perivascular and periadnexal infiltrate comprised of lymphocytes and plasma cells (Figure 2). The specimen from the left posterior arm also showed variable atrophy and acanthosis. In addition, inflammation was noted around the acrosyringium, and squamous metaplasia of these structures was focally evident (Figure 2B). These changes supported the diagnosis of KLC.

The patient failed a 5-month course of isotretinoin 1 mg/kg daily and was then lost to follow-up. Five years later she returned for reevaluation and to discuss treatment options. In the intervening 5 years, she had developed more diffuse, erythematous, keratotic papules on the arms, legs, and trunk (Figure 3). The lesions had spread with substantial involvement of the dorsal feet and toes as well as Achilles tendons. The breasts, buttocks, and neck were clear. Her face was clear, except for some inflammation of her eyelid margins. She had noted dramatic improvement of her eruption after sun exposure.

She was treated with numerous modalities including topical imiquimod, calcipotriene, topical tacrolimus, tretinoin cream, tazarotene cream, ammonium lactate lotion 12%, salicylic acid cream, urea

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The authors report no conflict of interest.

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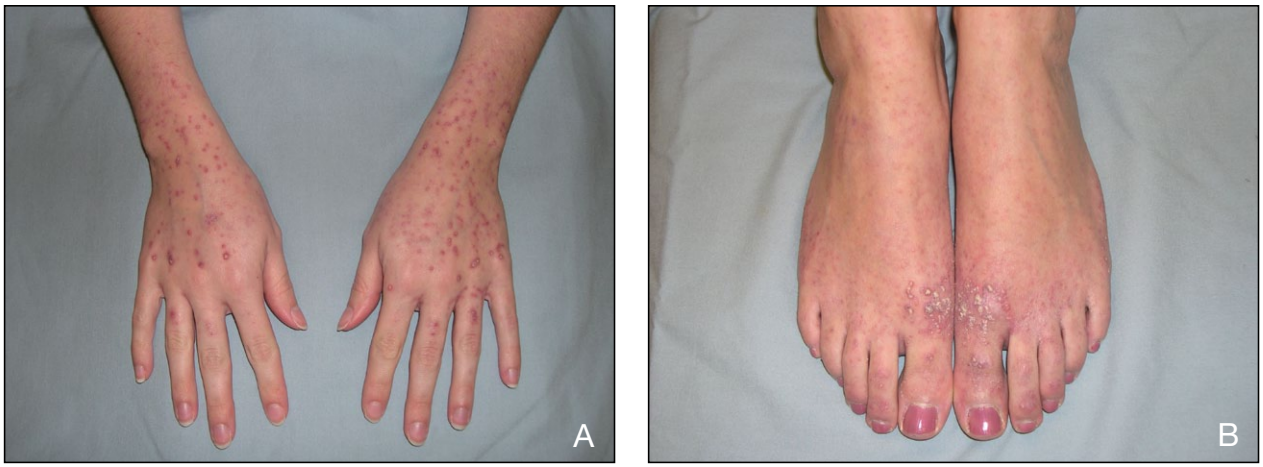

Figure 1. Lichenoid keratotic papules on the dorsum of the hands at the patient's initial visit (age, 14 years)(A). She also had punctate keratotic papules on the dorsum of the feet (B).

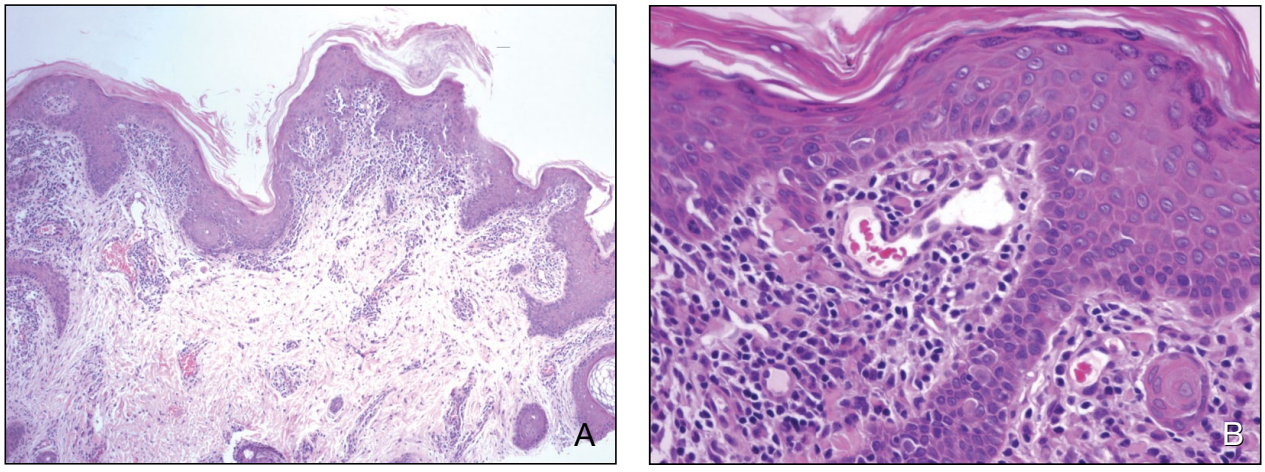

Figure 2. Histologic examination of the lesion from the left posterior arm showed focal parakeratosis, variable atrophy and acanthosis, vacuolar alteration of the basilar layer, telangiectasia, and a superficial and deep inflammatory infiltrate (A)(H&E, original magnification ×100). On higher magnification, Civatte body formation was evident (B) (H&E, original magnification ×200). The inflammatory infiltrate included lymphocytes and plasma cells. On the lower right (B), note squamous metaplasia involving the superficial portion of an eccrine sweat duct.

cream 40%, and topical steroids without improvement. Methotrexate was started but discontinued after 1 month because of side effects. On her last visit, phototherapy was discussed as a possible treatment option.

Comment

Keratosis lichenoides chronica is a rare acquired dermatosis of young to middle-aged adults. The disease was first described by Kaposi[2] in 1895 and later by Nekam[3] in 1938.[4] The term *keratosis lichenoides chronica* was introduced in 1972 by Margolis et al.[5] It is a chronic keratinization disorder that typically is characterized by violaceous, keratotic, lichenoid papules that are arranged in a reticulate or linear pattern. The lesions usually are distributed symmetrically on the limbs and trunk with a seborrheic dermatitis–like mediofacial eruption or psoriasislike scaling plaques.[6] The lesions usually are asymptomatic, are rarely accompanied by pruritus, and are typically thinner on the trunk compared to the extremities.[7] In 50% of adult cases, the disorder involves the oral or genital mucous membranes and presents as inflammation, ulceration, or infiltration. There is nail involvement in 30% of cases.[8] Nail changes include longitudinal ridges with red discoloration and slight distal onycholysis as well as thickening, ridging of the nail plate, brownish discoloration, and hyperkeratosis of the nail bed.[9] In other cases, the nail changes have been described as superficially resembling psoriasis but without pitting or pustulosis.[10]

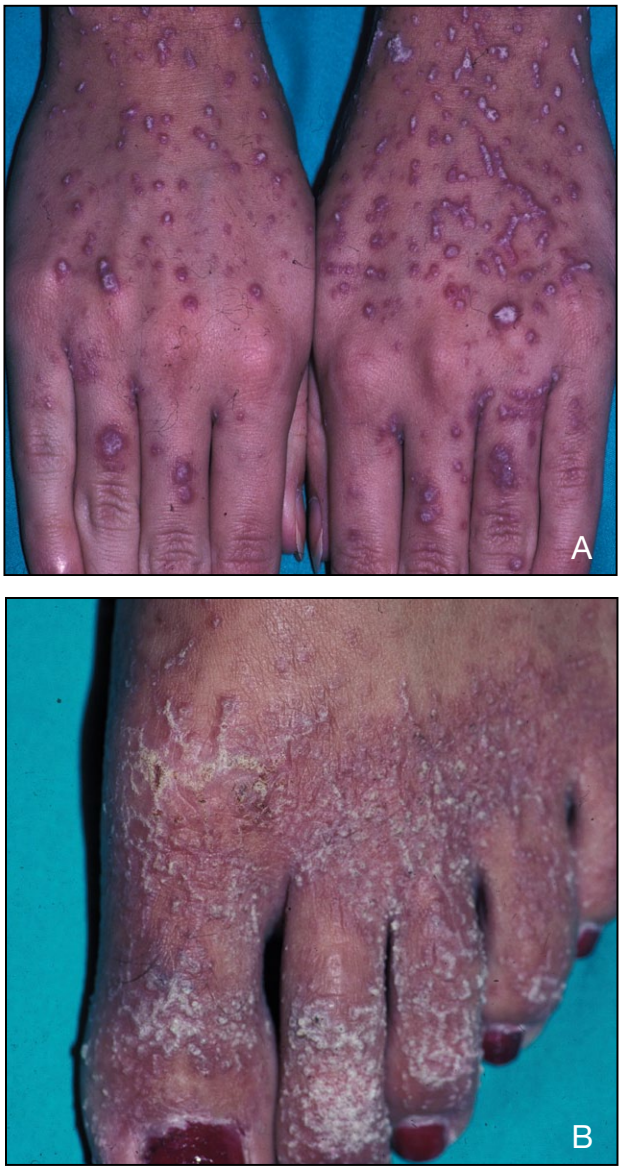

Figure 3. After 5 years' progression, the papules on the hands were more keratotic and lichenoid, and the linear pattern was more obvious (A). The papules on the feet also were more extensive and keratotic with substantial involvement of the dorsal feet and toes (B).

Pediatric occurrence of KLC is much less common than adult presentation; however, it may be more common than previously thought. This finding was described in a study conducted in 2007 in which 6 new pediatric cases of KLC were presented and compared to the total number of cases reported thus far.[1] Of the 54 cases studied, 14 were pediatric cases. The authors suggest that pediatric KLC might represent a different disease or a subset of adult-onset KLC because of the notable difference in clinical presentation. Unlike adult-onset KLC, pediatric KLC is characterized by probable autosomal-recessive inheritance; early or congenital onset with facial erythematopurpuric macules on the cheeks and chin; forehead, eyebrows, and eyelash alopecia; pruritus; and a low frequency of other cutaneous or systemic abnormalities.[1] Our patient's condition began during childhood, but the clinical findings were typical of adult-onset KLC.

The pathophysiology of KLC is not known. Some of the associations with KLC mentioned in the literature thus far include appearance after drug-induced erythroderma,[11] prolonged exposure to a source of heat (infrared radiation),[12] or trauma or carbamazepine treatment[13]; association with multiple eruptive keratoacanthomalike lesions in a patient with multiple myeloma[14]; association with mantle cell lymphoma and leg panniculitis, which seemed to improve after chemotherapy for the lymphoma[15]; association with atypical sarcoidal granulomatous inflammation (suggested that both KLC and the inflammation might have been a response to the same unknown antigenic exposure)[16]; association with hypothyroidism (2 cases reported)[17]; and association with a number of other systemic disorders such as toxoplasmosis, chronic lymphocytic leukemia, cutaneous amyloidosis, multiple sclerosis, chronic hepatitis, and glomerulonephritis.[7] Our patient had no relevant medical history and no systemic or other associated pathology.

The microscopic features of KLC include focal parakeratosis, variable atrophy and acanthosis, vacuolar alteration of the basilar layer with Civatte body formation, telangiectasia, and an inflammatory infiltrate that may include plasma cells and involve both deeper vessels and appendages.[18,19] Involvement of the acrosyringium also has been reported in a few cases, including squamous metaplasia, overlying hyperkeratosis, and hypergranulosis, as well as a lichenoid tissue reaction.[20,21] Virtually all of these features were encountered in our case, including perieccrine inflammation and squamous metaplasia, though the changes did not appear to be particularly focused on eccrine ducts. This combination of findings is quite characteristic, if not pathognomonic, for KLC, though the differential diagnosis would include lichen striatus or linear porokeratosis with a lichenoid tissue reaction.

The course of KLC is chronic and progressive; it rarely, if ever, shows signs of remission. In our patient, the course of the disease fluctuated yet became slowly but steadily more severe and extensive. Rarely, spontaneous resolution occurred.[22,23]

Treatment of KLC has been disappointing thus far. Unsuccessful therapies include topical and systemic steroids, topical coal tar preparations, radiation

therapy, ammoniated mercury, salicylic acid, anthralin, tretinoin, dapsone, erythromycin, tetracycline, gold, methotrexate, cyclosporine, griseofulvin, liquid nitrogen, and chloroquine sulfate.[5,11,24-26] There have been reported responses to psoralen plus UVA (PUVA), etretinate (or a combination of PUVA and etretinate), and calcipotriene.[25-29] In one patient treated with PUVA, all lesions disappeared and the results lasted for at least 2 years.[28] Similarly, a combination of photochemotherapy (PUVA) and retinoids with or without the inclusion of tacalcitol also has been suggested to be potentially beneficial.[30] In our patient, sun exposure resulted in dramatic improvement in the appearance of her lesions and she was nearly in remission during the summer. Review of the literature on KLC suggests a similar beneficial effect of sunlight in many patients with KLC.[1,17,31] These findings as well as the reported response to PUVA therapy suggest that phototherapy may be the most promising treatment of this recalcitrant condition.

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