CASE REPORT

Keratosis lichenoides chronica : A case report

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ABSTRACT

Keratosis lichenoides chronica(KLC) is a rare disease, with only about 70 cases reported in literature. KLC is characterized by violaceous, keratotic, lichenoid papules that are arranged in a characteristic linear and reticulated pattern. Lesions are usually symmetrically distributed on the limbs and trunk. KLC can sometimes be associated with oral, genital, ophthalmologic and systemic diseases. The disease is chronic, progressive and resistant to various treatment options. We report a 53-year-old male, who presented with asymptomatic symmetrically distributed keratotic papules, that were arranged in a characteristic linear and reticular pattern over both lower limbs

KEY WORDS: Keratosis lichenoides chronica, PUVA, acitretin

INTRODUCTION

KLC was first described in 1895 by Kaposi. He used the term “lichen ruber acuminatus verrucosus et reticularis” to describe a patient with a linear, warty lichenoid eruption.[1] Since his original description, patients with similar lesions have been reported, but with a confusing nomenclature which include lichen verrucosus et reticularis, porokeratosis striata lichenoides, keratosis lichenoides chronica, lichenoid trikeratosis, and Nekam's disease.

Over time, keratosis lichenoides chronica has become the most consistently used name for this disorder.[2]

KLC can occur in people of any race, age, or sex, The majority of cases have been described in adults aged 20-40 years; 24% of cases reported are children. The male to female ratio is 1.73 and most reported cases have been Caucasians. KLC is characterized by brownish-violaceous, thick scaly papules or small nodules on the trunk and extremities.[3] The papules are generally arranged in a linear or reticular pattern and are symmetrically distributed. They are usually asymptomatic but can be itchy in around 20% of cases. Keratosis lichenoides chronica has a chronic and often progressive course. In 70% of cases, patients also present with facial lesions resembling seborrhoeic dermatitis or rosacea.

Histopathologic features of KLC are often nonspecific, and include mild papillomatosis, focal parakeratosis, variable atrophy and epidermal acanthosis, vacuolar basal cell degeneration and keratinocytes necrosis.

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In this case report, we describe a 53-year-old male patient with characteristic clinical and histopathological features consistent with the diagnosis of keratosis lichenoides chronica.

CASE REPORT

We report a 53-year old male patient who developed skin lesions on the both lower limbs more than 38 years back. The lesions were largely asymptomatic, symmetrically distributed linear and reticular brownish to violaceous papules with white scaly surface.

Previously, he had been diagnosed at the age of 12 as a case of psoriasis, lupus and lichen planus. His lesions at that time were highly pruritic, involving mainly both upper limbs, and to a lesser extent the lower limbs. With the passage of time, the upper limb lesions disappeared and the lesions concentrated more over both lower limbs and became asymptomatic.

Topical corticosteroids, calcineurin inhibitors, phototherapy, acitretin and methotrexate had been tried with little to no benefit.

Skin examination at the time of presentation to our dermatology clinic at King Hussein Medical Center, revealed symmetrical brownish-violaceous papulonodular lesions with a keratotic surface, arranged in a linear and reticular pattern on the both lower limbs. (Fig.1)

Mild erythematous scaly lesions were present on the face, mainly over cheeks. There was no palmo-plantar, nail or mucosal involvement.

No regional lymphadenopathy was detected. Also, there was no genital involvement and his eyes appear normal with no obvious changes.

No scalp lesions were observed and there was no family history of similar lesions.

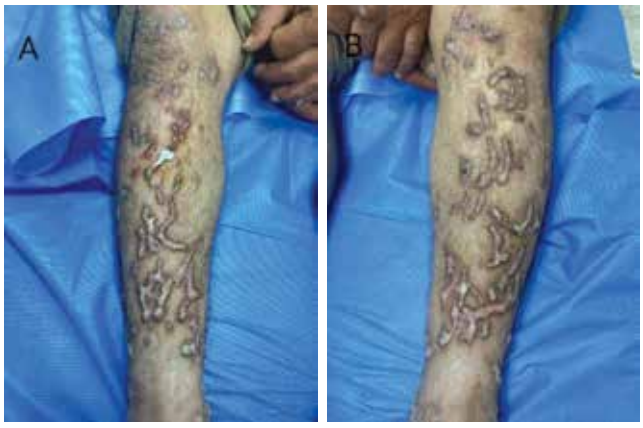

Fig. 1 A,B Multiple scaly keratotic papulo-nodular lesions in linear and reticular pattern over both lower limb

Laboratory investigations showed normal hematologic indices. But, biochemical and endocrinological tests showed hyperlipidemia and high thyroid stimulating hormone. Thus, he was diagnosed as a case of hypothyroidism with hyperlipidemia (high cholesterol and triglycerides). He was started on simvastatin and thyroxin by the attending physician. The hepatitis and HIV profile was negative.

ECG and echocardiogram showed a ventricular tachycardia and low ejection fraction, respectively.

Pacemaker was implanted and the patient was started on digoxin, warfarin and furosemide.

Histologic examination of lesional skin biopsy specimen showed papillomatosis, acanthosis, focal parakeratosis, and vacuolar degeneration of basal cell layer with necrotic keratinocytes (Fig. 2), PAS stain was negative. According to the clinicopathologic findings, he was diagnosed as a case of keratosis lichenoides chronica.

The patient was started on systemic retinoid (acitretin capsules: 25 mg BD) and PUVA (Re-PUVA modality of treatment).

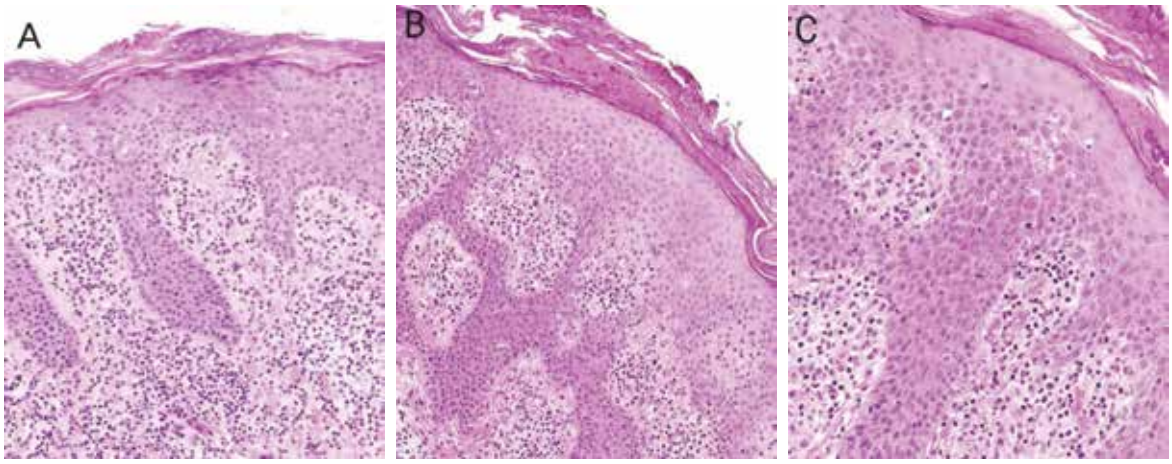

Fig. 2 (A) Acanthosis and dense lymphocytic infiltrates (H&E) (B) Hyperkeratosis, acanthosis, focal parakeratosis (H&E), (C) Variable vacuolar degeneration of basal cell layer, necrotic keratinocytes and mixed inflammatory cells, lymphocytes and plasma cells (H&E)

Follow up after 3 months revealed a satisfactory response to the treatment with flattening of most of the lesions. (Fig.3)

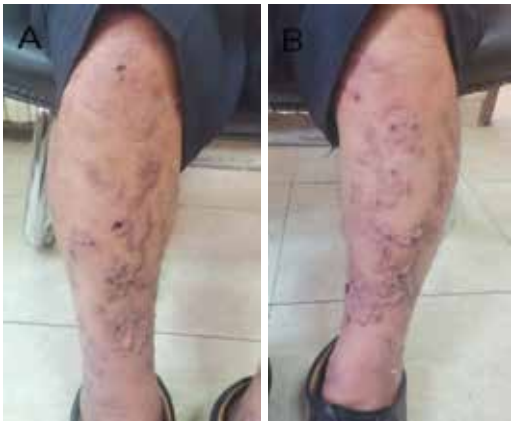

Fig. 3 (A,B) Flattening of the lesions on both lower limb after 3 months of Re-PUVA modality of treatment

DISCUSSION

The cause of keratosis lichenoides chronica is not well understood. It was initially thought to be a rare variant of lichen planus,[4,5] but many now consider it to be a distinct condition.

Some familial cases of keratosis lichenoides chronica are due to a germline mutation in NLRP, an inflammasome sensor gene that activates inflammatory cytokines. The aberrant activation of NLRP1 leads to the localized release of interleukin 1, secondary secretion of tumour necrosis factor-alpha and keratinocyte growth factor, resulting in epidermal hyperplasia and keratosis The criteria for diagnosing keratosis lichenoides chronica have not been well established. However, characteristic clinical features have been described in the literature. The lesions in KLC are typically scaly pink-red to violaceous papules and nodules. which are arranged in linear or reticular patterns.

A concurrent seborrheic dermatitis-like facial eruption or rosacea is also often present in almost 70% of patients.

KLC is usually asymptomatic, but has been shown to be pruritic in 20% of the total reported cases. Although uncommon, mucosal involvement has been reported and includes oral ulceration, genital ulceration,and ocular involvement (including blepharitis, conjunctivitis, uveítis, and iridocyclitis), nail changes including onychodystrophy and nail plate discoloration may be seen. Ruiz-maldonado et al studied 14 cases of pediatric onset KLC and compared those patients with 40 reported adult-onset KLC pateints.[6] They found that pediatric cases are mostly familial and

present with macular purpuric lesions on their faces with upper limbs lesional predilection and alopecia of eyelashes and eyebrows, which are not common in adults. Oral lesions are less frequently reported in pediatric cases. Also, itching was a more prominent symptom.

These finding support the initial signs and symptoms observed in our case, when it started at the age of 12 years. But, it changed dramatically as the disease progressed.

Associated systemic findings that are occasionally observed in the reported cases include hypothyroidism, cardiomyopathy, ischemic heart disease, hepatosplenomegaly and lymphadenopathy.

Histopathologic findings in KLC show a lichenoid, lymphocytic reaction pattern with focal basal vacuolar alteration. Perivascular and periappendageal lymphocytic infiltrate, sometimes with a few plasma cells, can be seen. Epidermal changes including alternating areas of atrophy and acanthosis with focal parakeratosis, follicular plugging, and sometimes neutrophilic debris can also be seen. Immunofluorescence is typically negative.[7] Together these histologic features help differentiate KLC from lichen planus, which lacks parakeratosis and shows wedge-shaped hypergranulosis. Some investigators, however, consider KLC a variant of lichen planus.

Management of KLC can be difficult and has not been studied in detail.[8] A recent review of KLC suggested that oral retinoids[9] and phototherapy, either alone or in combination,[10] were the most effective forms of treatment.

Our patient's disease was refractory to treatment with systemic corticosteroids, methotrexate, acitretin and NBUVB, consistent with prior observations.

Current treatment with acitretin with PUVA (Re-PUVA) resulted in partial improvement in his cutaneous lesions.

Long term follow up while on treatment is needed to assess the response and the sustainability of clinical improvement achieved.

CONCLUSION

KLC is a rare chronic progressive disease and represents a distinct entity with a wide range of differential diagnosis.

The diagnosis of KLC is highly dependent on clinical picture, pathological findings and the poor response to treatments.

KLC represents a mucocutaneous disease with a variety of associated skin and systemic diseases according to the data collected from the reported cases in literature.

REFRENCES

1. Margolis MH, Cooper GA, Johnson SA. Keratosis lichenoides chronica. Arch Dermatol. 1972; 105: 739-43.
2. Böer A. Keratosis lichenoides chronica: proposal of a concept. Am J Dermatopathol. 2006; 28:260-75.
3. Konstantinov KN, Sondergaard J, Izuno G, Obreshkova E. Keratosis lichenoides chronica. J Am Acad Dermatol. 1998; 38:306-309.
4. Petrozzi JW. Keratosis lichenoides chronica. Possible variant of lichen planus. Arch Dermatol. 1976; 112:709-11.
5. Kersey P, Ive FA. Keratosis lichenoides chronica is synonymous with lichen planus. Clin Exp Dermatol. 1982; 7:49-54.
6. Taberner R, Puig L, Fernandez-Figueras T, Alomar A. Keratosis lichenoides chronica. J Eur Acad Dermatol Venereol. 2001; 15:84-85.
7. Alpsoy E, Uzun S, Akman A, Acar MA, Memisoglu HR, Basaran E. Histological and immunofluorescence

findings of non-follicular papulopustular lesions in patients with Behcet's disease. J Eur Acad Dermatol Venereol. 2003; 17:521-24.

8. Pistoni F, Peroni A, Colato C, Schena D, Girolomoni G. Keratosis lichenoides chronica: Case-based review of treatment options. J Dermatolog Treat. 2016; 27:383-88.
9. Avermaete A, Kreuter JA, Stucker M, Von Kobyletzki G, Altmeyer P, Jansen T. Keratosis lichenoides chronica: characteristics and response to acitretin. Br J Dermatol. 2001; 144:422-24.
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; 85:552-53.