



Familial Actinic Lichen Planus: Three Cases from the Same Family

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Actinic lichen planus (ALP) that affects sun-exposed areas of the skin is an uncommon variant of lichen planus. While ALP is commonly triggered by ultraviolet radiation exposure, genetic predisposition may also be important in the pathogenesis of the disease. Herein, we report three patients with ALP from the same family, which supports the genetic etiopathogenetic factors of ALP.

Keywords: Actinic, Familial, Inheritance, Lichen planus

INTRODUCTION

Lichen planus (LP) is a papulosquamous inflammatory skin disorder of unknown origin that is relatively uncommon in children. Actinic lichen planus (ALP) is a rare clinical variant of LP. Children and young adults with darker skin types in tropical and subtropical regions are commonly affected, and lesions are almost always asymptomatic. ALP often occurs in the spring or summer seasons and develops on sun-exposed areas of the skin^{1,2}. The pathogenesis of ALP has not been well established, but several studies have found that lesions can be reproduced by exposure to ultraviolet radiation^{1,2}. As ALP has a racial predilection affecting predominantly individuals of Middle Eastern origin, it is likely that a combination of environmental factors and genetic predispositions may be important in the pathogenesis of the disease³. Familial ALP cases have been reported very rarely in the literature.

Herein, we present three patients with ALP from the same family in order to contribute to familial ALP knowledge in the literature.

CASE REPORT

Case 1

A 5-year-old boy with a Fitzpatrick type-IV skin type presented to our outpatient clinic with a 4-month history of facial hyperpigmentation. All the lesions were located on sun-exposed areas of the skin. The lesions first began in the spring season but were asymptomatic. A physical examination revealed dark brown macules on his malar area and upper lip. We also observed papular lesions on the dorsum of his hands (Fig. 1A). Skin biopsies were taken and histopathological analysis indicated the lesions were identical to LP with a demarcated band-like lymphocytic infiltrate (Fig. 1B).

Case 2

A 11-year-old boy with a Fitzpatrick type-IV skin type, who was the brother of the first case, complained of recurrent lesions located in sun-exposed skin areas during every summer season for 3 years. A physical examination revealed several dark brown patches on his right forehead and upper lip. Li-

chenoid papular lesions were present on the back of his hands (Fig. 2A). Histopathological analysis of a skin biopsy revealed the lesions were consistent with interface dermatitis (Fig. 2B).

Case 3

A 43-year-old male with a Fitzpatrick type-IV skin type, who was the father of the above-mentioned patients, presented with a complaint of a photodistributed dermatitis that erupted each summer season for the last 5 years. A physical examination found several papular lesions with scaling on the back of his hands (Fig. 3A). There were no other lesions. This patient

didn't have no skin lesions on his face in the past. Histopathological analysis of a skin biopsy revealed the lesions were consistent with psoriasiform dermatitis (Fig. 3B).

Common findings and treatments of all cases

All the family members admitted to outpatient clinic in month of august.

All three patients were not using any medications, and none had systemic diseases. Physical examinations revealed no signs of LP in other parts of their bodies including the oral mucosa, hair, or nails. Other physical examination findings were normal.

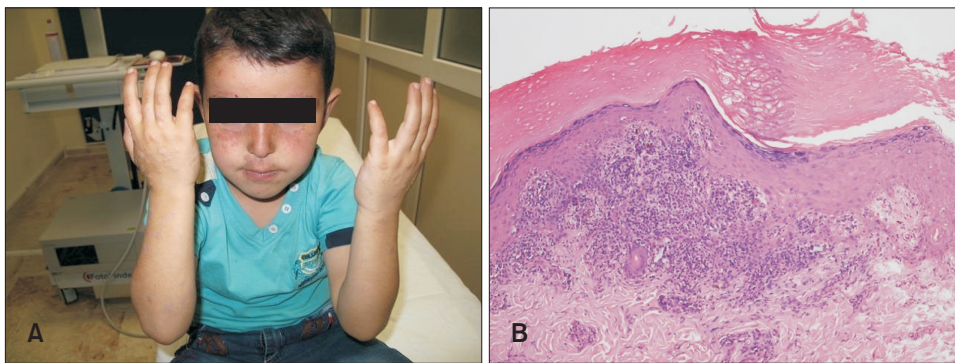


Fig. 1. A case of a 5-year-old boy with actinic lichen planus. (A) Melasma-like dark brown patches on the malar area, macules on the upper lip, and papular lesions on the back of hands. (B) Histopathological appearance of a skin biopsy showing hyperkeratosis, hypergranulosis, and lichenoid infiltration at the dermo-epidermal junction in the epidermis (H&E, $\times 100$).

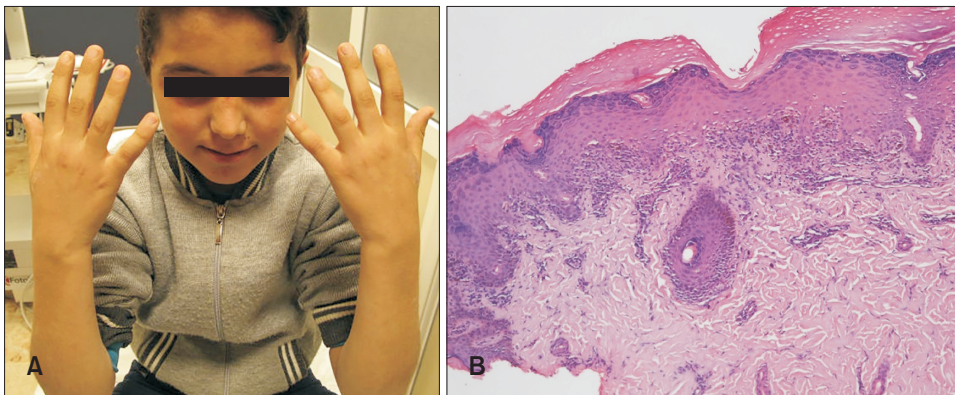


Fig. 2. A case of a 11-year-old boy with actinic lichen planus. (A) Dark brown patches on the right forehead and upper lip, papular lesions on the back of hands. (B) Histopathological appearance of a skin biopsy showing parakeratotic epidermis under hyperkeratosis, and interface dermatitis at the dermoepidermal junction (H&E, $\times 100$).

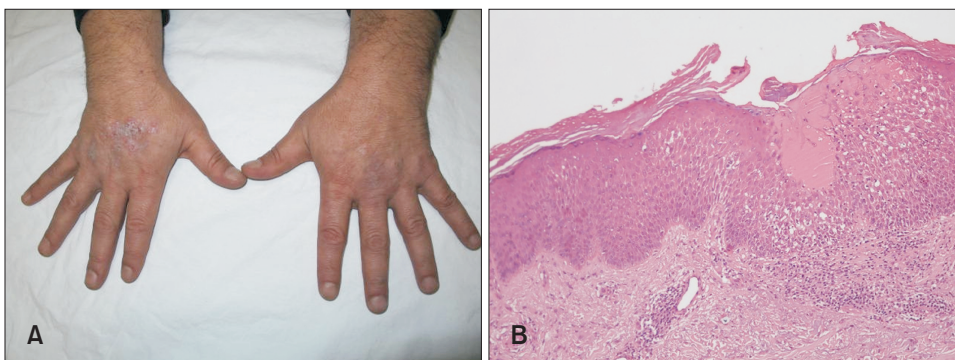


Fig. 3. A case of a 43-year-old male with actinic lichen planus. (A) Papular lesions with scaling on the back of hands. (B) Histopathological appearance of a skin biopsy showing exocytosis in the acanthotic and spongiotic epidermis, and serosity on the surface (H&E, $\times 100$).

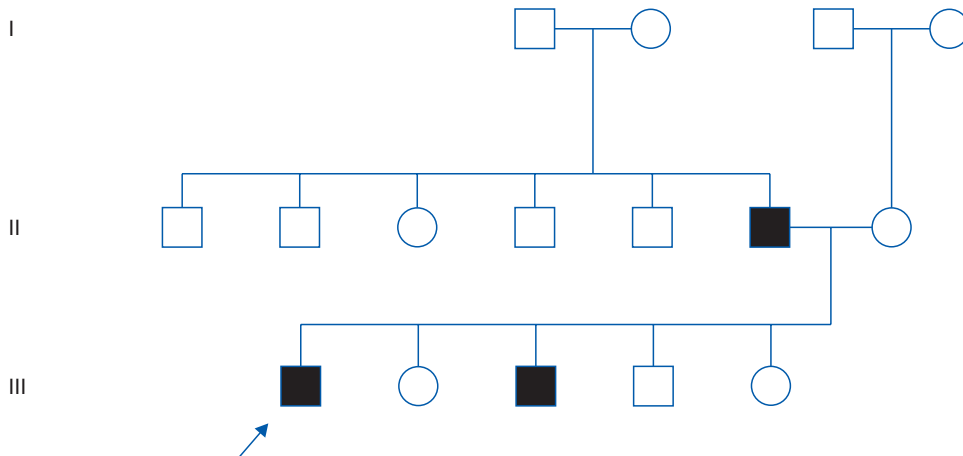


Fig. 4. The pedigree of the family history.

All the skin biopsies were taken from the back of their hands and forearms.

A blood test analysis indicated the three patients were hepatitis B surface antigen and anti-hepatitis C virus Ab negative, but all were anti-hepatitis B surface positive. HLA typing revealed that there was no increase of HLA-B7, HLA-DR1, or DR10.

The pedigree of the family history can be seen in Fig. 4.

The patients were advised to use sun protection and were given a topical corticosteroid cream for treatment.

All patients provided written and oral consent. The procedures in this report are in accordance with the 1975 Declaration of Helsinki and were approved by our institutional review board.

We received the patient's consent form about publishing all photographic materials.

DISCUSSION

LP affects between 0.5% and 1% of the population, with many of the LP variants occurring much more infrequently than classic LP¹. The ALP variant of LP is more common in females than males and its occurrence in children is rare⁴. In the international literature, the frequency of ALP varied from 2% to 11.5%⁵. In the present report, two of three patients were boys and all patients were male.

The clinical features of ALP are variable. In addition to lichenoid or annular papules, macular hyperpigmentation and infiltrated erythematous plaques with variable borders and scaling can be observed¹. Various clinical types have been identified, such as annular, plaque-like, dyschromic, and pigmented⁶. The most common form, annular ALP, is character-

ized by erythematous brownish plaques in an annular configuration with or without atrophy. Pigmented forms are present as hypermelanotic patches with a melasma-like appearance. Dyschromic-type ALP lesions are the most rare and are characterized by whitish pinhead and coalescent papules⁶. We report two cases with the pigmented form of ALP and one case with the plaque-like form. All cases were individuals from the same family. Frequently affected sites include the face, especially the forehead, cheeks, lips, and the dorsal surface of the hands². In all of the present cases, the lesions were localized to these areas.

Diagnosis is typically made based on the observed distribution of lesions in sun-exposed areas in combination with histological findings consistent with those of classic LP. Histologically, any papular element will usually show features of LP. Even macular areas may show subtle evidence of interface dermatitis with prominent dermal melanophages². In addition to the classical LP-like findings, there may be various histopathological findings ranging from nonspecific eczematous dermatitis to lichenoid tissue reactions⁶. Histopathological analysis of our cases revealed classical LP, interface dermatitis, and psoriasiform dermatitis, respectively.

Based on clinical and histological features of case 3, several skin diseases, including chronic actinic dermatitis, may be considered in differential diagnosis. But, clinically actinic dermatitis predominantly affects middle-aged or elderly male. Case 3 is younger patient. As in this patient, it may be difficult to distinguish ALP from chronic actinic dermatitis clinically and histopathologically. This is in the limitation in our study.

The aetiology and pathogenesis of LP remains unclear. An autoimmune reaction in which CD8+T lymphocytes attack basal keratinocytes and lead to apoptosis of the cell has been

favoured². Various potential triggers, such as viral or bacterial antigens, metal ions, drugs, or physical factors, could initiate the autoimmune process. In general, LP has not been shown to have a racial predilection; however, certain populations seem to have a higher incidence of the disease^{1,2}. ALP has been reported commonly from Middle East areas, similar to LP. The high proportion of cases reported among these populations suggests a genetic predisposition for LP susceptibility. This idea is further supported by cases of familial LP, reported in 2% of childhood LP cases⁷. Many reports describe the tendency of familial lichen planus to develop at an early age, to becoming severe or chronic, and can have widespread atypical manifestations⁸.

The fact that lesions are not widespread, even if ALP cases are familial, and that disease manifestation is localized to sun-exposed areas supports that triggering factors can determine the location of the lesions.

Genetic linkage studies have established HLA is associated with LP. Copeman et al.⁹ found an association between familial LP and HLA-B7. Although the relationship of the disease with HLA types has been reported in familial cases, it is suspected that the test for HLA typing is not useful for demonstrating the inheritance of LP¹⁰. HLA typing in our cases was not compatible with the HLA types mentioned in the literature.

Another precipitating factor of LP is vaccination⁷. LP may appear after any dose of vaccination and the latent period from the latest vaccination varies from days to three months. Kanwar and De⁷ reported that 15% of patients in their study in India developed LP after vaccination. The mean interval between vaccination and LP onset was three years, and ranged between three months and 11 years. In patients with shorter intervals, hepatitis B vaccination was suspected to be causally related. The patients in our cases had been routinely vaccinated, but the immunization dates were years before the onset of the lesions. However, the relationship between vaccination and LP may be weak because the rate of administration of hepatitis B vaccination is much higher in children in Turkey than India.

A majority of previous studies could not effectively demonstrate the association of LP with genetics, viral infections, or vaccinations. The fact that our three cases are from the same family supports the theory that genetic background can influence ALP susceptibility after ultraviolet exposure. However, genetic inheritance of ALP is a matter that must be confirmed by further studies.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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