Original Article

Childhood Lichen Planus: A Series of 42 Patients

Abhinav Kumar, Vibhu Mendiratta, Shilpi Agarwal[1], Ram Chander, Sarita Sanke

Departments of Dermatology and STD and [1]Pathology, Lady Hardinge Medical College and Associated Hospitals, New Delhi, India

Abstract

Background: Lichen planus (LP) is a papulosquamous disease of unknown etiology that is relatively uncommon in children. There is a paucity of data on the clinical profile of LP in children. Available case series are few and majority being retrospective. A cross-sectional observational study to evaluate the clinical profile of childhood LP was performed. **Materials and Methods:** All childhood cases (<18 years) with histopathologically confirmed diagnosis of LP were evaluated. Detailed clinical history, examination, and investigations were performed according to a proforma. **Observation and Results:** There were 42 children and childhood LP constituted 1.4% of the pediatric dermatoses. There were 26 females (61.9%) and 16 (38.1%) males with male to female ratio of 1:1.6. The age ranged from 2 to 18 years with a mean age of 11.6 ± 5.1 years. The duration of the disease ranged from 15 days to 5 years with a mean of 8.6 ± 9.4 months. History of recent hepatitis B vaccination was found in 6 (14.3%) patients, and exposure to X-ray radiation was seen in 3 (7.1%). The most common morphological presentation of LP was papules and plaques observed in 34 (81%) patients. Classical LP was the most common variant, found in 29 (69%) patients, followed by hypertrophic variant in 7 (16.7%) patients. Koebnerization was found in 31 (73.8%) patients. Oral mucosa involvement was seen in 28.6% of patients, nail in 42.85%, and scalp in 7.1% of patients. **Conclusion:** Childhood LP resembles adult LP in most of the aspects. However, etiological factors in childhood LP may be different. It is an under-reported disease. Large multi-centric prospective studies should be undertaken to acquire a better understanding of the clinical profile of childhood LP.

Keywords: *Childhood lichen planus, nail lichen planus, oral lichen planus, papulosquamous disorder*

Introduction

Lichen planus (LP) is a papulosquamous disease of unknown etiology. It is considered to be an autoimmune T-cell mediated disease that affects skin, mucous membrane, hair follicles, and nail. Childhood LP is considered traditionally rare, as reflected by the relative paucity of data on the same. The supposed "rarity" of childhood LP could be due to several factors such as underreporting, lesser exposure to environmental triggers and rare coexistence of other associated autoimmune diseases which manifest later in life. The clinical profile, etiopathogenesis, course, and treatment options in childhood LP are different from those in the adult LP.

Aims

This aim was to study the clinical profile of LP in children.

Materials and Methods

This was a cross-sectional observational study, conducted in the Department of Dermatology of a Tertiary Care Hospital. The study period was from January 2014 to March 2015. All patients of age group 6 months to 18 years with clinically and histopathologically confirmed the diagnosis of LP was taken in our study after informed consent. Detailed clinical history, examination, and investigations (complete blood count, erythrocyte sedimentation rate, liver function tests, lipid profile, thyroid function tests, antinuclear anybody, hepatitis B and C serology) were done according to a predesigned proforma. Body surface area involved was calculated. The intensity of itching was graded as mild, moderate, and severe according to scoring system as shown in Table 1. The intensity of pruritus for children <10 years was measured by parents/guardian by noting the frequency of disturbance in sleep and intensity of scratching throughout the day.

Statistical analysis

This was analyzed using SPSS (Statistical software for social sciences, Version 18.0, Chicago, USA.) developed by IBM

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Address for correspondence:
Dr. Sarita Sanke, Department of Dermatology and STD, Lady Hardinge Medical College and Associated Hospitals, Shaheed Bhagat Singh Marg, New Delhi - 110 001, India.
E-mail: sankesarita@gmail.com

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corporation, using appropriate tests. *P* value was considered statistically significant if <0.05.

Observations and Results

Childhood cases of LP constituted 1.4% of the pediatric dermatoses in our hospital. A total of 42 patients with clinically and histopathologically confirmed the diagnosis of LP were included in the study. There were 26 females (61.9%) and 16 (38.1%) males with male to female ratio of 1:1.6. The age ranged from 2 to 18 years with a mean age of 11.6 ± 5.1 years. The duration of the disease ranged from 15 days to 5 years with a mean of 8.6 ± 9.4 months. The distribution of age, duration of illness, and body surface area involved is shown in Table 2.

Progressive disease (appearance of new lesions in the last 6–12 weeks) was seen in 33 (78.6%) patients, whereas 9 (21.4%) patients had stable disease (no new lesions in the last 6 months). The itching was severe in 18 (42.85%) patients, moderate in 12 (28.57%), and mild in rest of the patients.

On evaluating the associated risk factors, history of recent hepatitis B vaccination (HBV) was found in 6 (14.3%) patients, and exposure to X-ray radiation was seen in 3 (7.1%). None of the patients gave a history of blood transfusion. History of autoimmune disease in the family was present in several patients as shown in Table 3. Treatment history was present in 11 patients (26.2%), of which 3 (7.1%) gave a history of homeopathic medication, and 8 (19%) gave a history of intralesional injections.

Classical LP was the most common variant, found in 29 (69%) patients, followed by hypertrophic variant in 7 (16.7%) patients. Koebnerization was found in 31 (73.8%) of the patients. The most common morphological presentation of LP was papules and plaques observed in 34 (81%) patients. The detailed clinical characteristics of the patients are shown in Table 4, and extracutaneous involvement is shown in Table 5.

On investigating the patients, 10 (23.8%) patients were found to have anemia, 6 (14.3%) had abnormal liver function test, 3 (7.1%) had altered lipid profile, and 3 patients (7.1%) had hypothyroidism. None showed seropositivity for hepatitis B, hepatitis C, or antinuclear antibody positivity. Erythrocyte sedimentation rate was not increased in any patient.

Discussion

Childhood cases of LP constituted 1.4% of the pediatric dermatoses. Handa and Sahoo have reported the prevalence of 2.5% of childhood LP among the pediatric dermatology patients.[1] In India, different studies have quoted prevalence of LP among dermatology patients to be 0.38%–1.4%.[2,3] The prevalence in our study coincides with the prevalence of LP in general population and rarity of childhood LP is due to underreporting of cases and lack of concerned data.

Table 1: Scoring system for severity of itching

Severity	Score of the patient
Mild	0-4
Moderate	5-7
Severe	8-10

Table 2: Distribution of age, duration of illness and body surface area involved in the patients

	Number of patients (%)
Age group	
6 months to 6 years	8 (19.0)
>6-12 years	11 (26.2)
>12-18 years	23 (54.8)
Duration of illness (months)	
1-3	13 (31.0)
3-6	9 (21.4)
6-12	5 (11.9)
>12	15 (35.7)
Body surface area involvement (%)	
<5	9 (21.4)
5-10	12 (28.6)
10-20	19 (45.2)
>20	2 (4.8)

Table 3: Family history of autoimmune disease

Family history	Number of patients (%)
Diabetes	6 (14.3)
Psoriasis	4 (9.5)
Vitiligo	2 (4.8)
Thyroid disease	2 (4.8)
Others (LP)	2 (4.8)
Rheumatoid arthritis	1 (2.4)
Atopic dermatitis	0
Alopecia areata	0

LP - Lichen planus

The age group of our study population ranged from 6 months to 18 years. The youngest patient in our study was 2 years. The earliest reported age in previous studies is 2 weeks.[4] The mean age of our study population was 11.61 ± 5.09 years, whereas the mean age in larger studies has been 7.1–8.4 years.[1,5] As the majority of children in our study were in the age group of 12–18 years (54.8%), we had a higher mean age. The majority of children had disease between 5 and 9 years in a study by Handa and Sahoo.[1] The variability of the affected age in childhood LP depends on the age composition in different study groups and underlying etiopathogenetic mechanisms. Several factors are responsible for early onset such as genetic predisposition, vaccines, and drugs.

The male to female ratio was 1:1.6 which shows a higher female preponderance, whereas earlier studies have shown either equal sex ratio or male preponderance.[1] Higher male to female ratio in Indian studies may be due to reporting bias in the Indian society. The duration of disease ranged from

Table 4: Clinical characteristics of the patients

Clinical variants of cutaneous LP	Frequency (%)
Classical	29 (69)
Hypertrophic	7 (16.7)
Zosteriform	2 (4.8)
Classical	1 (2.4)
Follicular	1 (2.4)
Eruptive	1 (2.4)
Site of cutaneous lesions	**Number of patients (%)**
Leg	34 (81.0)
Foot	26 (61.9)
Arm	16 (38.1)
Hand	10 (23.8)
Chest	9 (21.4)
Back	7 (16.7)
Face	1 (2.4)
Number of body sites involved (single/multiple)	**Frequency (%)**
Single	11 (26.2)
Multiple	31 (73.8)
Extracutaneous involvement	**Frequency (%)**
Finger nail	14 (33.3)
Oral mucosa	12 (28.6)
Toe nail	3 (7.1)
Scalp	3 (7.1)

LP - Lichen planus

Table 5: Extracutaneous involvement of lichen planus in children

	Frequency (%)
Extracutaneous involvement	
Oral mucosa	12 (28.6)
Reticulate	9 (29.0)
Atrophic	2 (4.8)
Papular	1 (2.4)
Erosive	1 (2.4)
Nail involvement	17 (40.47)
Finger nail	14 (33.3)
Toe nail	3 (7.1)
Melanonychia	7 (16.66)
Longitudinal striation	5 (11.90)
Horizontal ridging	4 (9.52)
Periungual hyperpigmentation	2 (4.76)
Pterygium	1 (2.38)
Nail plate thinning	2 (4.76)
Onychorrhexis	2 (4.76)
leukonychia	1 (2.38)
Scalp involvement	3 (7.1)
Genital mucosa	0
Palms and soles	0

15 days to 5 years with a mean of 8.63 ± 9.41. The duration was more than 1 year in most of the patients (35.7%), which indicates a delay in reporting to the clinician. LP is thus a subacute to chronic disease of childhood.

Progressive disease was seen in 78.6% of patients, whereas 21.4% had stable disease suggesting that disease was active at the time of presentation in the majority of patients. Nearly 7.1% of the patients had taken homeopathic and 19% had a history of intralesional injections indicating that various modalities of treatment were explored before presenting to us. Homeopathy is popular in the treatment of pediatric skin disorder. Some of these drugs contain heavy metals which may contribute to the aggravation of LP. History of HBV was found in 35.7% of patients. Cross-reactivity between antigen in HBV and epitopes on keratinocyte has been suggested.[6] However, causal relationship between LP and HBV cannot be established. Hepatitis C virus (HCV) has been suspected to be a risk factor in LP. Positive associations have been suspected in studies from Japan, Italy, and India;[7] however, none could prove such an association. Nearly 7.1% of patients gave a history of exposure to X-ray radiation, but none had a history of LP lesions appearing on the site of exposure. LP at the site of radiation exposure has been reported and is called "isoradiotopic response."[8]

The family history of LP was present in 4.8%. The incidence of familial childhood LP is 1%–2% according to various studies.[9] Familial LP differs from the classical form clinically, with earlier age at onset, more generalized involvement, and more common mucosal involvement with a tendency for erosive, ulcerative and linear forms, with prolonged course and frequent relapses.

History of autoimmune disease in the family was present in 22.6% of the patients. This suggests that LP is an autoimmune disease with a strong association and predilection for other autoimmune diseases. Coexistence of both psoriasis and vitiligo, together with LP has been reported.[10] A study from Pakistan reported the prevalence of thyroid disease in cutaneous LP to be 1.5%.[11]

Classical LP was the most common variant in 69% of our patients. The findings were consistent with the observation in various studies where classical LP ranged between 42% and 70%.[1] Hypertrophic variant occurred in 16.7% of patients and was mainly on the extensor aspect of legs and foot. Other studies have reported a lower incidence of LP hypertrophicus in children (8%–10%).[4,5] Higher prevalence of hypertrophic variant in our study could be on account of older children (13–18 years) in our study who undergo hair removal procedures (shaving and waxing), inducing koebnerization and thus, a hypertrophic variant of LP. Zosteriform LP constituted a total of 4.8% of the patients, which is lower than various studies who report a prevalence of 8%–12%.[1] However, eruptive variant was rare limited to 1 patient in contrast to other studies which reported a higher prevalence.[1,4] This may be explained by the fact that most of patients in our study group has disease duration of more than 1 year, whereas eruptive variant appears in a very short time and spreads rapidly to involve a large part of the body. Leg was the most

common site with 81% involvement followed by foot with 61.9% involvement which is consistent with various studies carried out in the past.[5] Koebner phenomenon was seen in 73.8% of patients, which is higher compared to other studies that report an incidence between 24% and 28%.[1,5]

Mucosal involvement with cutaneous lesion was found in 28.6% of patients which was in agreement with other studies quoting a frequency of 13.7%–30%.[1,6] Nail findings were found in 42.9% of patients. Melanonychia was most common finding in 22.2% of the patients, followed by periungual hyperpigmentation and longitudinal striation in 16.7%. Patients with multiple nail changes were seen in 33.3% of patients. This is important as nail involvement is believed to be rare in children and is often not reported by the clinician. Tosti *et al*. reported nail involvement in 11% of childhood LP cases.[12] The most common changes are an exaggeration of the longitudinal lines and linear depressions, elevated ridges, and pterygium unguis.[13] Rarely, the nail is completely shed with clinical evidence of LP at the base of the nail before shedding. LP of the nail bed may give increase to hyperpigmentation, longitudinal melanonychia, subungual hyperkeratosis, or onycholysis or changes mimicking the yellow nail syndrome. Scalp involvement with cicatricial alopecia was seen in 7.1% of our patients, which is in agreement with various studies.[5,14]

High incidence of anemia (23.8%) in our patients could be incidental, considering the high prevalence of anemia in Indian population. Dyslipidemia was seen in 7.1% of our cases, which can be explained by the fact that lymphocytes attack keratinocytes resulting in the generation of reactive oxygen species. During this process, keratinocytes release more cytokines that in turn attack more lymphocytes.[15] These cytokines involved in LP pathogenesis (such as tumor necrosis factor-alpha, interleukin [IL]-6, IL-10, and IL-4) could explain the association with dyslipidemia.[15] Very few studies have looked into the lipid profile in childhood LP. Dyslipidemia is a surrogate marker for cardiovascular morbidity. Hence, LP (generalized) might increase the risk of cardiovascular morbidity.

Conclusion

Childhood LP resembles adult LP in most of the aspects. However, it is an under-reported disease. Large multi-centric prospective studies should be undertaken to acquire a better understanding of the clinical profile of childhood LP.

We had few limitations in our study like small sample size. We could not establish causal relationship with viral markers nor with vaccination for hepatitis B. Human leukocyte antigen typing could not be done in familial cases.

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Conflicts of interest

There are no conflicts of interest.

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