

# Non – classical clinical variants of lichen planus . Clinicopathological study of 43 cases

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## Abstract

**Background:** Lichen planus is a well known inflammatory skin disease with characteristic clinical features in its classical form. Yet, sometimes it presents clinically in a non-classical forms. These non-classical variants of lichen planus must be highlighted for their early diagnosis.

**Design:** It is a retrospective study

**Objective:** To focus on the clinical and histopathological feature of the non-classical variants of lichen planus, to show the distribution of these variants of lichen planus in a group of our patients in Jordan.

**Materials and Methods:** All cases of lichen planus with non-classical presentations that have been seen by us in our practice at Jordan University Hospital, Al-Karak Teaching Governmental Hospital and in private practice from Jan. 2014 to Dec. 2017 were included in this study. Their clinical data were retrieved and analyzed. Skin biopsy was performed in all cases. Their histopathological features were analyzed.

**Results:** Out of a total number of 135 cases of lichen planus cases that have been seen during the period of this study from Jan. 2012 to Dec. 2017 there were 43 cases of lichen planus with non-classical clinical presentations representing 32% of the total number of all lichen planus cases with age range from 3 to 80 year old (mean age 36 year).

There were 25 males and 18 females with male to female ratio 1.4: 1. Different non-classical variants of lichen planus were seen in our study. The main diagnostic histopathological features of lichen planus were present in all cases but with some variation.

**Conclusion:** It is not uncommon for lichen planus to present in non-classical variants with variable clinical presentations but the histopathological features are almost always diagnostic and by histopathological examination and clinicopathological correlation the diagnosis can be made precisely.

**Keywords:** clinical variants, lichen planus.

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## Introduction

Lichen planus is an idiopathic inflammatory disease of the skin, mucous membranes, nails and scalp. The overall prevalence of lichen

planus is estimated to be less than 1% of the general population <sup>(1)</sup>. The etiology of lichen planus is not known, but a cell Mediated immune response to an epidermal antigen in genetically predisposed person is considered in

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its pathogenesis<sup>(2)</sup>. Various possible triggers like; viral or bacterial antigen, drugs, metal ions or physical factors could induce the autoimmune process<sup>(3)</sup>. Different clinical variants of lichen planus have been described<sup>(4)</sup>. In its classical variant lichen planus has typical diagnostic clinical presentation and the diagnosis can be made based on clinical basis alone but lichen planus may be present in a non-classical variant that may pose a diagnostic challenge. In such cases the histopathological examination is always needed to make the proper diagnosis and should not be delayed.

In this study we will present and discuss the clinicopathologic features of some non-classic presentations and variants of lichen planus in a group of our patients in Jordan.

### Materials and methods:

From Jan, 2014 to Dec. 2017 all the cases that have been diagnosed as non-classical variants of lichen planus at Jordan University Hospital, Al-Karak Teaching Government Hospital and in private practice in Amman were included in this study. Their clinical and demographic features were retrieved and analyzed. Skin biopsy was performed for all 43 cases and the diagnosis was confirmed by histopathological interpretation by a dermatopathologist. All the skin biopsy slides were reviewed and seen by the researches and the histopathological features were analyzed.

### Results:

From the total number of 135 cases of lichen planus that have been seen and diagnosed by us during the study period from Jan. 2014 to Dec. 2017, there were 43 cases of non-classical variants of lichen planus cases representing 32% of all lichen planus cases that have been seen. Different non-classical variants of lichen

planus have been seen in our study including : lichen planus actinicus , lichen planopilaris , eruptive or exanthematous lichen planus , lichen planus pigmentosus inversa Figure 1, lichen planus pigmentosus , lichen planus pemphigoides , linear lichen planus, hypertrophic lichen planus Figure 2, plantar lichen planus Figure 3, erythrodermic lichen planus Figure 4 and isolated lip lichen planus .

Lichen planus actinicus represent the majority of cases (9/43) (20.9%) followed by lichen planus pigmentosus 7/43 (16.2%) and lichen planopilaris (6/43) (13.9%).

Table 1 shows the distribution and demographic features of these variants of lichen planus.

Table 2 shows the age distribution of these cases.

The characteristic symptom of lichen planus, itching was present in (38/43) (88.3%) patients, which is more intense in erythrodermic and hypertrophic variants of lichen planus and less prominent in lichen planus pigmentosus and lichen planus pigmentosus inversa. Associated oral mucosal involvement was present in (5/43) patients (11.6%) , both cases of lichen planus pemphigoides showed mucous membrane lesions, nail involvement was present in (3/43) patients (7%) . Male genitalia was involved in (1/43) patient (2.3%).

The majority of patients with non-classic variants of lichen planus in this study were in the age group (21-50), there were (24/43) patients (55.8%).

The main diagnostic histopathological features that are needed for the diagnosis of lichen planus in general were present in all cases and include: hyperkeratosis, hypergranulosis or accentuation of the granular

cell layer, lichenoid or focally lichenoid inflammatory lymphoid cell infiltrate, hydropic degeneration of the basal cell layer and pigmentary incontinence. Some variation in histopathological features were seen in some variants like; epidermal atrophy, less prominent lichenoid inflammatory cell infiltrate, less prominent hypergranulosis and more prominent pigmentary incontinence was seen in cases of lichen planus pigmentosus Figure 5 , lichen planus pigmentosus inversa and in few cases of lichen planus actinicus . Bullous lesions of lichen planus pemphigoides showed subepidermal blister with inflammatory cell infiltrate that is focally lichenoid and containing polymorphs ( eosinophils and neutrophils) but the lichenoid papular lesions of lichen planus pemphigoides cases showed typical histopathological changes of lichen planus.

Marked epidermal hyperplasia, prominent hyperkeratosis and prominent hypergranulosis were seen in hypertrophic lichen planus. Lichen planopilaris cases showed lichenoid inflammatory reaction involving the follicular epithelium and sparing the interfollicular epidermis Figure 6 .The other non-classical variants of lichen planus in our study showed histopathological changes similar to that of classical variant Figure 7. Colloid bodies were seen in (30/43) patients (70%). Sawtoothing was present in (35/43) patients (81%). Squamous cell carcinoma proved by skin biopsy was seen in one patient with hypertrophic lichen planus Figure 3. Skin biopsy was diagnostic for all cases that have been biopsied in this study (43/43) (100%).

### Discussion:

Classical lichen planus has its characteristic diagnostic clinical and histopathological features, clinically characterized by itchy

violaceous polygonal papules and plaques on extremities and histologically characterized by hyperkeratosis, wedge-shape hypergranulosis, hydropic degeneration of the basal cell layer, sawtoothing , lichenoid inflammatory cell infiltrate hugging the epidermis, colloid body formation and pigmentary incontinence <sup>(1,2)</sup> Figure 7.

Our study focused on the non-classical variants of lichen planus . There are many variants of lichen planus other than the classical type that have been reported including; annular lichen planus <sup>(5)</sup> hypertrophic lichen planus, atrophic lichen planus , ulcerative lichen planus, Bullous lichen planus , lichen planus pemphigoides, lichen planus pigmentosus, lichen planus pigmentosus inversa, erythrodermic lichen planus, inverse lichen planus, linear lichen planus, lichen planopilaris, lichen planus actinicus , lip lichen planus, oral lichen planus, eyelid lichen planus , nail lichen planus, eruptive lichen planus and palmoplantar lichen planus .

Different variants of non- classical lichen planus were present in our study as shown in tab.1 with their distribution, frequency and demographic features. They represent the majority of the above mentioned reported variants of lichen planus. The majority of patient in our study fall in the (21-50) years age group which is younger than that in other studies for lichen planus in general. The most prevalent variant in our study is lichen planus actinicus. Lichen planus actinicus is known to occur in persons with middle eastern descent. The second most frequent variant in our study is lichen planus pigmentosus with female predominance. Lichen planus pigmentosus is also a variant of lichen planus that may be more common in middle eastern population and more frequently in female. Lichen planopilaris cases

in our study include one case with a rare linear presentation of lichen planopilaris over the forehead ( Linear lichen planopilaris).

This rare variant has been reported<sup>(16, 17)</sup>. It is important to note that cases of eruptive or exanthematous lichen planus in our study were all young patients (<18 yr). Although this is rare in the literature but eruptive lichen planus have been reported in children. One of our patients in the study with lichen planus pigmentosus inversa had eyelid lesion. Eyelid lichen planus have been reported.

Lichen planus pemphigoides cases were two so it is a rare variant with more involvement of the extremities by bullous lesions in addition to the presence of typical lichenoid papules with the presence of mucous membrane lesions and these presentation are similar to that reported in the literature but in our patients the age is younger, palmoplantar lichen planus is also a rare variant that has been reported to occur in older age group with involvement of the palms and soles, mucous membrane lesions and pruritus. Our patients with palmoplantar lichen planus in the study showed similar features to the reported cases in the literature. Malignant changes of lichen planus are a very rare event but it has been reported in hypertrophic lichen planus. One of our patients developed squamous cell carcinoma in association with hypertrophic lichen planus after having it for 15 years Figure 3. We had one case of each erythrodermic lichen planus Figure 4 and isolated lip lichen planus with clinicopathological features of these two cases were similar to the reported cases in the

literature<sup>(9)</sup>. The diagnosis was made by histopathological examination in both cases.

The histopathological examination was diagnostic in all cases in our study and this concurs with other researchers. These variants of lichen planus are clinically different but their histopathological findings are almost similar and always diagnostic and the diagnosis can be certainly made. One exception is that lichen planopilaris cannot be differentiated from frontal fibrosing alopecia by histopathological examination and in such cases clinicopathological correlation is needed to make the diagnosis. It is important to note that 10/34 (23%) of our patients in the study were children under 18 years. Incidence of lichen planus in children is thought to be as low as (1.2%), but in an Indian studies it was (11%-19%). Our figures are close to that from India. We suggest that the incidence of non-classical lichen planus in children is not low. Other studies with larger groups of patients are needed to draw conclusions.

### **Conclusion:**

There are different non-classical variants of lichen planus affecting all age groups and they can present with variable clinical presentation but the histopathological features of the skin biopsy is almost always diagnostic for lichen planus. So skin biopsy should not be delayed for proper-early management. Very rare variants of lichen planus including; erythrodermic, linear lichen planopilaris and isolated lip lichen planus were present in our study.

**Table 1** Non –classical variants of lichen planus in our study and their demographic features

Variant of lichen planus	M	F	Age range : ( mean)	Number of patient (%)
Lichen planus actinicus	8	1	3-45 (26.6) years	9 (20.9%)
Lichen planopilaris	4	2	28-58 (41) years	6 (13.9%)
Eruptive or exanthematous lichen planus	4	1	6-17 (10.6) years	5 (11.6%)
Lichen planus pigmentosus inversa	0	3	20-38 (33) years	3 (6.9%)
Lichen planus pigmentosus	3	4	10-65 (33.7) years	7 (16.2%)
Lichen planus pemphigoides	0	2	10-13 (11.5) years	2 (4.6%)
Linear lichen planus	1	2	27-48 (36.6) years	3 (6.8%)
Hypertrophic lichen planus	1	2	30-80 (52.2) years	3 (6.9%)
Plamoplantor lichen planus	2	1	29-70 (55.6) years	3 (6.9%)
Erythrodermic lichen planus	1	0	65 years	1 (2.3%)
Isolated lip lichen planus	1	0	70 years	1 (2.3%)

**Table 2** Age distribution of non-classical variants of lichen planus in our study

Age group in years	Number of patients (%)
0-10	6
11-20	5
21-30	9
31-40	7
41-50	8
51-60	2
61-70	5
71-80	1
Total ( 0-80 )	43



**Figure-1:** Lichen planus pigmentosus inversa, eyelids lesions.  
 Note: The patient had also similar lesions on retro-auricular area and neck



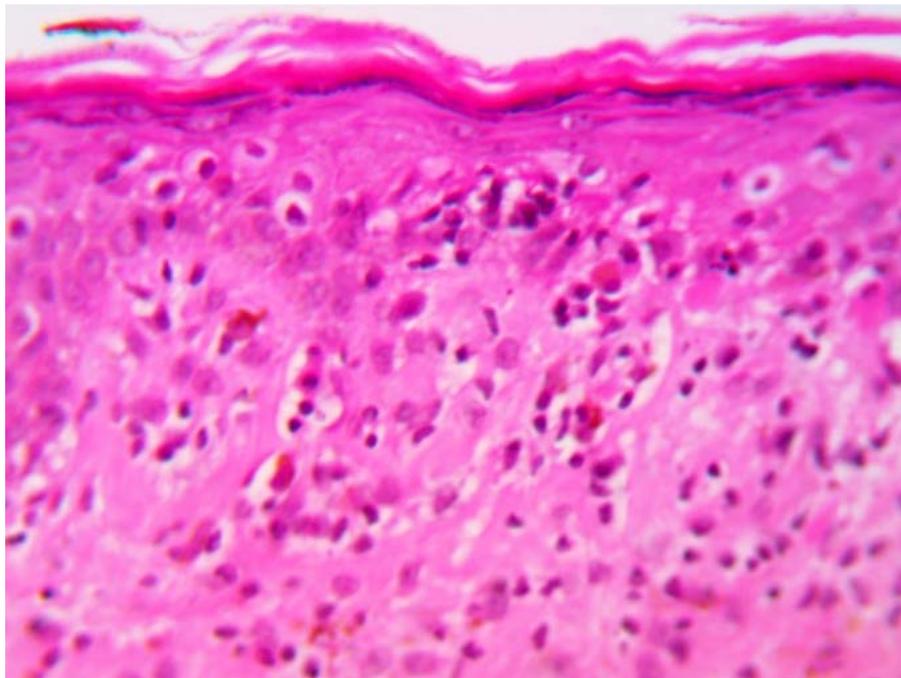
**Figure-2:** Hypertrophic lichen planus. Lesions on both legs and on dorsum of hands with squamous cell carcinoma on the Left leg



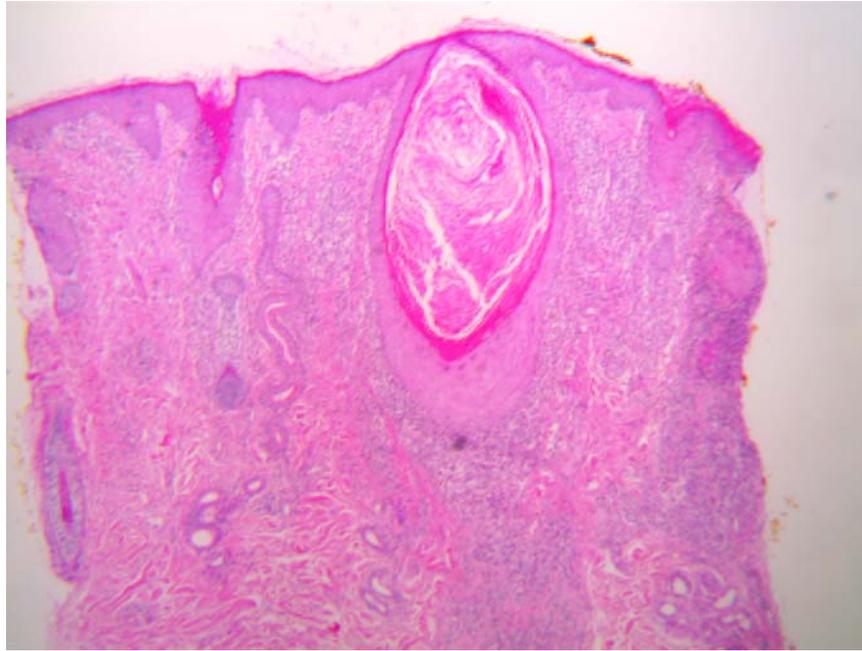
**Figure-3:** Palmoplantar lichen planus, plantar lesions  
Note: the patient had palmar lesions too



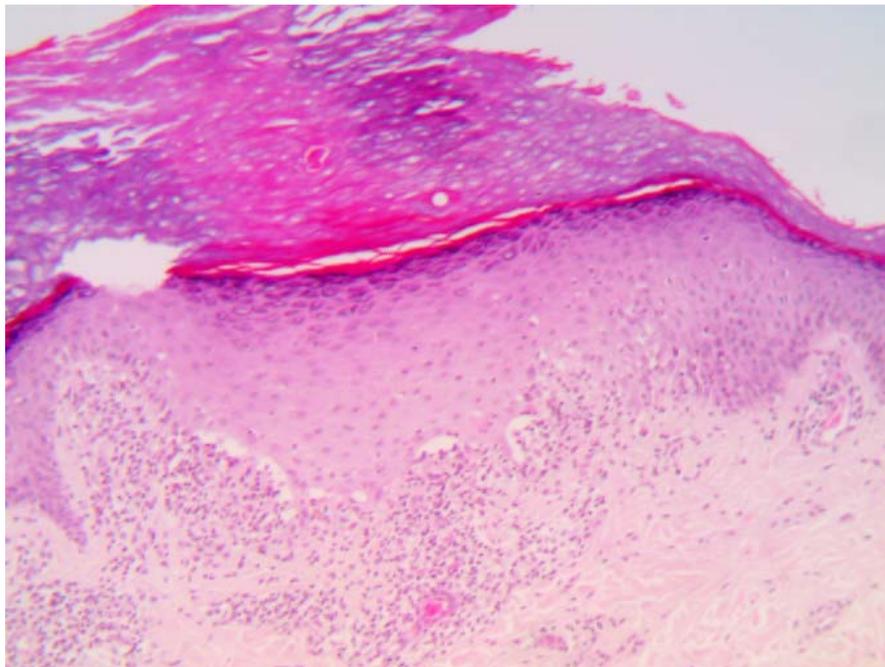
**Figure-4:**Erythrodermic lichen planus. Wide spread erythematous papulosquamous skin lesions



**Figure-5:**Lichen planus pigmentosus. Intermediate power view x 20 Shows: hyperkeratosis, accentuation of the granular cell layer, sawtoothing , hydropic degeneration of the basal cell layer, colloid body formation, pigmentary incontinence and mild lichenoid lymphoid cell infiltrate



**Figure-6:** Lichen planopilaris .Low power view x10 shows: hyperkeratosis , follicular hyperkeratosis,lichenoid inflammatory cell infiltrate of the hair follicle hugging the follicular epithelium and sparing the interfollicular epidermis



**Figure-7:** Lichen planus with classical diagnostic histopathological features .Intermediate power view x 20 shows: Hyperkeratosis, hypergranulosis, epidermal hyperplasia , lichenoid inflammatory cell infiltrate hugging the epidermis , hydropic degeneration ,Max-Joseph space formation, colloid bodies and pigmentary incontinence

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## الأنواع السريرية غير التقليدية للحزاز المسطح دراسة للعلامات السريرية والمجهرية المرضية لثلاثة وأربعين (43) حالة

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### الملخص

الخلفية: الحزاز المسطح هو مرض جلدي التهابي معروف بخصائصه السريرية المميزة في شكله التقليدي ، ولكنه أحياناً يظهر سريريا في أشكال غير تقليدية. يجب تسليط الضوء على هذه الأشكال غير التقليدية من الحزاز المسطح من اجل التشخيص المبكر لها. التصميم : دراسة استعادية.

الهدف: التركيز على الخصائص السريرية والتشريحية المرضية للأنواع غير التقليدية من الحزاز المسطح ، لإظهار توزيع هذه الأنواع من الحزاز المسطح في مجموعة من مرضانا في الأردن.

الأساليب والأدوات: تم شمول جميع حالات الحزاز المسطح غير التقليدية التي تم تشخيصها أثناء ممارستنا في مستشفى الجامعة الأردنية، ومستشفى الكرك التعليمي الحكومي وفي القطاع الخاص من يناير 2014 إلى ديسمبر 2017 في هذه الدراسة. تم استرجاع تاريخهم السريري وتحليله. تم إجراء فحص خزعة الجلد في جميع الحالات وتم تحليل العلامات النسيجية المرضية.

النتائج: من بين إجمالي 135 حالة من حالات الحزاز المسطح التي شوهدت خلال فترة الدراسة من يناير 2012 إلى ديسمبر 2017 كان هناك 43 حالة من الحزاز المسطح لها علامات سريرية غير تقليدية تمثل 32% من العدد الإجمالي من جميع حالات الحزاز المسطح وفي الفئة العمرية تتراوح بين 3 إلى 80 سنة (متوسط العمر 36 سنة).

كان هناك 25 من الذكور و 18 من الإناث وكانت نسبة الذكور إلى الإناث هي 1.4 : 1. وشوهدت أنواع مختلفة من الأنواع غير التقليدية للحزاز المسطح في دراستنا. وكانت العلامات التشخيصية التشريحية المرضية المميزة للحزاز المسطح موجودة في جميع الحالات ولكن مع بعض الاختلاف.

الخلاصة: ليس من غير المألوف أن يظهر الحزاز المسطح سريريا بأنواع غير تقليدية مع اعراض وعلامات سريرية متعددة لكن العلامات النسيجية هي دائما مختلفة وتؤدي الى التشخيص السليم وبواسطة الفحص النسيجي المجهرى وربطه بالعلامات السريرية يمكن عمل التشخيص بدقة.

الكلمات الدالة: سريري مرضي، الحزاز المسطح، أنواع سريرية.