Histopathologic Evaluation of Linear Lichen Planus and Lichen Striatus

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Background **: Linear lichen planus (LLP) and Lichen striatus (LS) are rare disorder that can be confused because they can share similar clinical and histopathologic features.**

Objective **: The purpose of this study was to evaluate the characteristic differences and common features between the two disorders histopathologically.**

Methods **: We reviewed the clinical records of patients who had been diagnosed as LLP or LS in our dermatology clinic during the 15-year period between 1985 and 1999. We classified twenty seven cases, which were differentiated from other possible linear dermatoses, into LLP and LS on the basis of clinical features, and then compared them histopathologically, and appreciated the characteristic differences or common features of the two disorders.**

Results **: In cases diagnosed as LLP clinically, epidermal changes were mainly composed of hyperkeratosis (78%), acanthosis (78%), basal degeneration (78%), granular layer thickening (67%) and saw-toothed appearance of rete ridges (44%). In dermis, colloid bodies (78%), band-like inflammatory cell infiltration with pigmentary incontinence (78%) were striking findings. In cases with clinical features of LS, parakeratosis (50%), dyskeratotic cells scattered in the epidermis (61%) and intercellular edema (39%) were noted in the epidermis. Dermal cellular deposits were focally band-like infiltration(89%), more frequently perivascular infiltration (83%) and often involved deep dermis (50%), hair follicles (44%) and eccrine glands (22%) in contrast to LLP.**

Conclusion **: This study presents a comparative histopathologic features of LLP and LS. Three cases of LLP with overlapping histopathologic features suggest the possibility that there may be an intermediate form between either end of a spectrum, LLP and LS.**

(Ann Dermatol 13(4) 211~217, 2001).

Key Words : Linear lichen planus, Lichen striatus

Linear lichen planus (LLP) is a rare variant of lichen planus (LP) accounting for 0.24-0.62% of all patients with LP showing pruritic, polyangular, purple papules arranged in linear patterns. Like classic LP, it usually develops insidiously, lasts for more than 1 year and sometimes leaves hyperpigmentations, but it is mostly localized in a portion of the body and known to mainly affects children in contrast to classic LP[1].

Lichen striatus (LS) is an uncommon disorder that manifests as continuous or interrupted erythematous papules in linear, unilateral patterns mainly occurring in children from 5 to 15 years of age[2]. The eruption is usually asymptomatic showing spontaneous clearance within 3-6 months in most cases, but it often leaves hypopigmentation especially in dark-skinned patients[3]. The distribution of the lesions is thought to follow Blaschko's lines on the extremities, trunk, or neck in many cases[4].

Received August 5, 2000.
Accepted for publication June 27, 2001.
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Annals of Dermatology
Vol. 13, No. 4, October 2001

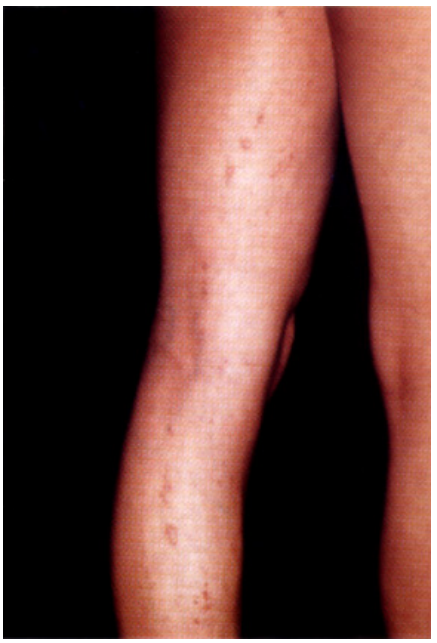

Fig. 1. A 36-year-old female patient with features of LLP showing linearly arranged lichenoid papules along a length of left leg with postinflammatory hyperpigmentations.

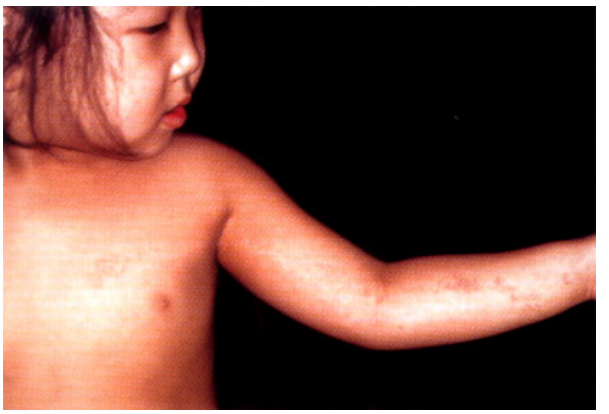

Fig. 2. A 4-year-old female child with features of LS showing oblique, linear lesions on the left side of her chest that extend along nearly whole length of the upper extremity following Blaschko's lines. The lesions are accompanied by hypopigmentations.

Though there are some other diseases that present with localized, linear eruption besides LLP and LS such as linear form of psoriasis, linear epidermal nevus and so on, LLP and LS occasionally misdiagnosed as each other especially because they share similar clinical appearance partly and there can be many overlapping features in histopathologic findings[5]. In order to evaluate the helpful informations for differentiating both conditions, we classified twenty seven cases, which were differentiated from other possible linear dermatoses, into LLP and LS on the basis of clinical features, and compared them histopathologically, and appreciated the characteristic differences or common features of the two disorders.

MATERIALS AND METHODS

We reviewed the clinical records of patients who had previously been diagnosed confirmatively or presumptively as LLP or LS between 1985 and 1999. All patients presented with grouped erythematous or violaceous papules which were arranged in a linear or segmental patterns and confined to a localized area of the body.

We excluded the conditions that mimicked LLP or LS by thorough clinical and histopathologic review, most of which proved to linear psoriasis, linear epidermal nevus, inflammatory linear verrucous epidermal nevus (ILVEN), lichen simplex chronicus linearis, and so on. After ruling out the above conditions, we got twenty seven cases compatible with LLP or LS.

We first classified the cases into LLP and LS by considering solely clinical criteria (onset, duration, nature of papules, itching, and pigmentary changes) described in dermatologic texts and literatures as follows ; The cases presenting with violaceous papules of characteristic Wickham's striae, pruritic papules of insidious onset and chronic course, or sequelae of hyperpigmentation were regarded as LLP (9 cases), while the cases of relatively early age, acute onset, papules rather erythematous than violaceous, or nonpruritic lesions with hypopigmentation were regarded as LS (18 cases). Then we compared the pathologic specimens (hematoxylin-eosin stain) obtained from the patients of two clinically classified groups.

RESULTS

Besides the criteria we relied on to classify the cases to LLP and LS clinically (onset, duration, nature of papules, itching, and pigmentary changes), there were no striking clinical differences between two groups (Table 1). The sites of prevalence were commonly extremities sometimes

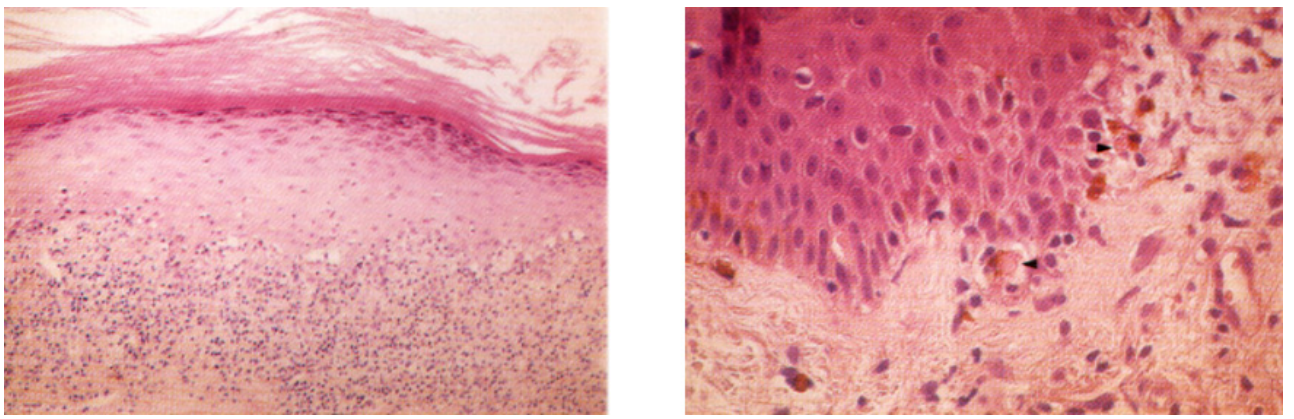

Fig. 3. Histopathologic findings of LLP showing hyperkeratosis without parakeratosis, acanthosis, focal thickening of granular layer, saw-toothed rete ridges, and band-like cellular infiltration in the upper dermis with basal layer degeneration (A: H&E, ×100). The presence of colloid bodies (arrows) and pigmentary incontinence are observed in the upper papillary dermis (B: H&E, ×400).

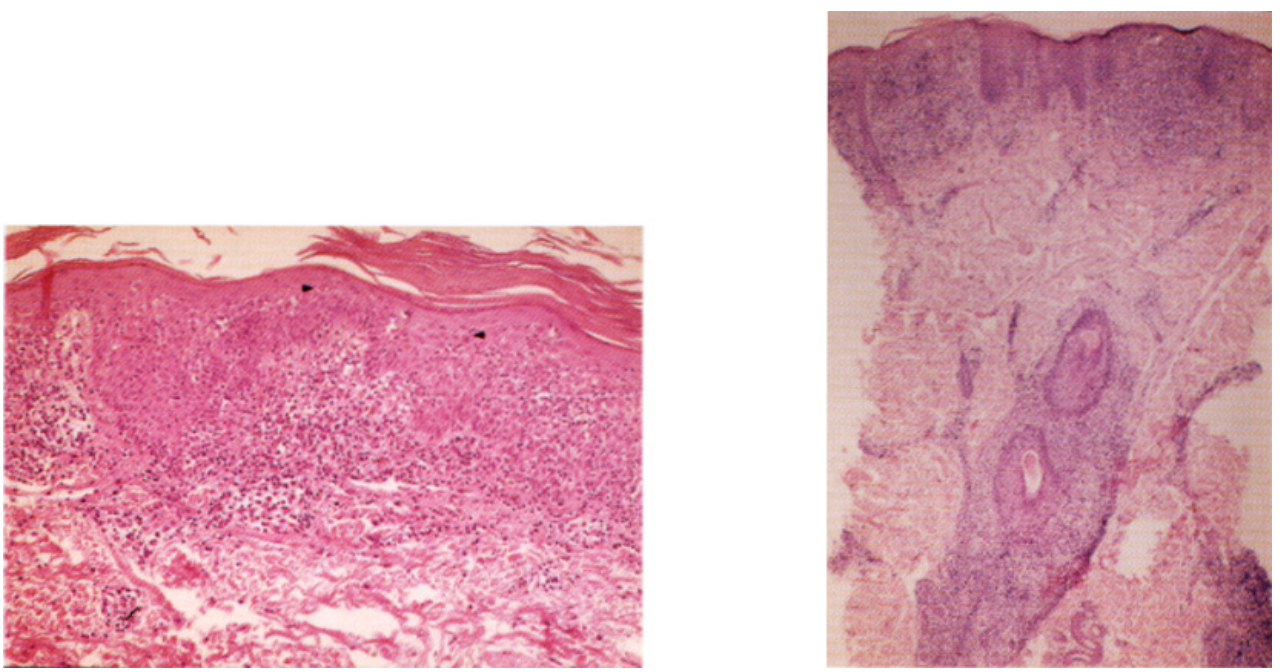

Fig. 4. Histopathologic findings of LS showing focal band-like lymphocytic infiltration in the papillary dermis and focal parakeratosis, relatively irregular acanthosis, mild intercellular edema with exocytosis, and scattered dyskeratotic cells (arrows) in the epidermis (A: H&E, ×100). Focal band-like cellular infiltration is observed in papillary dermis and dense cellular infiltration is also observed around the deep follicles and sweat glands. (B: H&E, ×40).

involving nearly whole length of a limb (Fig. 1), and two cases of LSs presented lesions on the trunk and arm that showed oblique distribution along Blaschko's lines (Fig. 2).

Histopathologic comparison between the two groups is summarized in Table 2. They shared similar epidermal changes including hyperkeratosis, mild acanthosis and basal degeneration, but the findings were somewhat more prominent in clinical LLPs than clinical LSs, and the latter often showed mild atrophy. Parakeratosis and intercellular edema were observed in 9 (50%) and 7 (39%) cases of 18 LSs respectively while only 2 (22%) and 1 (11%) cases of 9 LLPs revealed such findings. 11 cases (61%) of 18 LSs showed scattered dyskeratotic cells in the epidermis, and the finding was less fre-

Table 1. Profiles of patients diagnosed as linear lichen planus (LLP) or lichen striatus (LS) on the basis of clinical examinations (*: cases showing intermediate histopathological findings such as deep dermal infiltration and follicular involvement)

LLP

No.	Age(y)/sex	Duration	Site	Itching	Pigmentary alteration
1	30/F	3 months	Lt. knee	-	hyper
2	27/F	3 years	Rt. thigh-knee	-	hyper
3	9/F	6 months	Rt. thigh	+	no
4	31/M	1 year	both arms	+	no
5*	24/F	6 months	Lt. arms	-	hyper
6*	23/F	10 months	Lt. arms and chest	+	hyper
7*	12/M	3 years	Lt. thigh	+	no
8	44/F	2 years	Rt. leg	-	hyper
9	36/F	1 year	Lt. leg	+	hyper

LS

No.	Age(y)/sex	Duration	Site	Itching	Pigmentary alteration
1	3/F	5 months	Lt. arm	-	no
2	5/F	2 months	Rt. arm	-	no
3	30/F	1 month	Rt. thigh-leg	+	hypo
4	3/M	6 months	Rt. leg	-	hypo
5	2/F	1 month	Lt. leg	-	no
6	8/M	1 week	Rt. arm	-	no
7	4/M	1 year	Rt. leg	-	hyper and hypo
8	9/F	1 month	Rt. cheek and arm	-	no
9	8/F	2 months	Rt. chin	-	no
10	4/F	1 year	Lt. chest-arm	-	hypo
11	5/F	3 months	Rt. arm	-	no
12	5/M	5 months	Rt. arm-hand	-	no
13	2/M	7 months	Lt. back	-	hypo
14	15/F	2 months	Lt. leg	+	hypo
15	6/F	1 year	Rt. leg	-	no
16	3/F	3 months	Rt. arm	-	hypo
17	28/F	2 weeks	Rt. thigh-leg	-	no
18	3/F	1 month	Rt. thigh	-	no

quent in LLPs (33%). On the other hand, focal granular layer thickening was observed in 6 cases (67%) of 9 LLPs, but not common in LSs (4%). Saw-toothed appearance of rete ridges was also prevalent in LLPs (44%), but not observed in LSs.

Colloid bodies were observed in the papillary dermis in both groups, but it was much easily detected in LLPs (78%) than LSs (44%). Papillary dermis appeared to be edematous with varying degrees of accompanying exocytosis in 4 cases (22%) of 18 LSs while the finding was not observed in LLPs. Each group showed different patterns of inflammatory cell infiltration in the dermis. Band-like infiltration was observed in LLPs, focal band-like infiltration in LSs (78% and 89% respectively) Perivascular infiltration was more dominant in

Table 2. Histopathologic comparison between LLP and LS

Characteristics	LLP	LS
Epidermis		
hyperkeratosis	7 (78)	10 (56)
parakeratosis	2 (22)	9 (50)
acanthosis	7 (78)	10 (56)
granular layer thickening	6 (67)	4 (22)
saw-toothed rete ridges	4 (44)	0 (0)
dyskeratotic cells	3 (33)	11 (61)
basal degeneration	7 (78)	10 (56)
intercellular edema	1 (11)	7 (39)
Dermis		
colloid bodies	7 (78)	8 (44)
papillary dermal edema	0 (0)	4 (22)
band-like infiltration	7 (78)	16(89)
RBC extravasation	0 (0)	5 (28)
pigmentary incontinence	7 (78)	8 (44)
perivascular infiltration	4 (44)	15 (83)
inflammatory cells		
lymphocytes	9 (100)	18 (100)
histiocytes	9 (100)	18 (100)
eosinophil	0 (0)	1 (6)
plasma cell	0 (0)	1 (6)
deep dermal infiltration	1 (11)	9 (50)
follicular involvement	2 (22)	8 (44)
sweat gland involvement	0 (0)	4 (22)
Tested cases (%)	9 (100)	18 (100)

LSs than in LLPs (83% and 44% respectively). In addition, LSs often revealed cell infiltrations in the mid to deep reticular dermis (50%), around hair follicles (44%) or sweat glands (22%), but these findings were rare in LLPs. Three cases of LLP showed intermediate histopathological findings such as deep dermal infiltration(11%) and follicular involvement(22%). The infiltrated cells were mostly composed of lymphocytes and histiocytes, but there were one case showing eosinophils and one case showing plasma cells in 18 LSs. RBC extravasation was observed in 5 cases (28%) in 18 LSs, but not in LLPs. The characteristic histopathologic features of each condition are illustrated on Fig. 3 and Fig. 4.

DISCUSSION

It is sometimes difficult to differentiate a variety of dermatoses when a patient presents with linear or segmental lesions localized to some portion of the body. Rothmans and Niederman[6] suggested that in cases of linear streaks of lichenoid papules, four conditions should be considered: LLP, LS, linear psoriasis, and linear epidermal nevus. Other forms of linear lesions to be considered are lichen simplex chronicus linearis, verrucae vulgaris linearis, incontinentia pigmenti, linear morphea, linear porokeratosis, linear lichen nitidus, and so on[7]. Some of the above dermatoses can be easily confirmed due to their congenital nature, characteristic histopathologic features or laboratory findings, but differentiating LLP or LS from each other can be the puzzling problem for several reasons. Both conditions are fairly uncommon disease of unknown origin, and can share similar clinical and histopathologic features partly[3]. In addition, in contrast that typical LP generally develops in adult between the ages of 30 and 60 years[8], the linear variant usually affects children as LS does[9]. So we intended to classify twenty seven cases into clinical LLP and LS on the basis of clinical features, and then compare them histopathologically, and appreciate the characteristic differences or common features of the two dermatoses.

As a result, the most relevant histopathologic features of LSs were focal band-like lymphocytic infiltrates and dense perivascular lymphohistiocytic infiltrates in the dermis that often involved deep or periadnexal regions, and epidermal changes including hyperkeratosis with spotty parakeratosis, spongiosis along with a varying degree of exocytosis and scattered dyskeratotic cells. Acanthosis, though common findings, was relatively focal and some specimens showed mild atrophic changes. LLPs, on the other hand, were characterized by band-like dermal cell infiltrates, focal hypergranulosis, acanthosis, saw-toothed rete ridges and the presence of colloid bodies in the papillary dermis and lowermost epidermis, which were generally compatible with classic LP. In general, epidermal findings such as parakeratosis, scattered dyskeratotic cells and intercellular edema, and dermal perivascular infiltration of cells extending to deep dermis or adnexa were mainly observed in LSs, while granular layer thickening, saw-toothed rete ridges, colloid

bodies and band-like cell infiltration in the dermis without deep dermal infiltration or adnexal involvement were striking in LLPs though some mixed features of above findings existed. These results were generally consistent with those suggested by Rubio et al[3] and Herd et al[5] previously, and in most cases the differentiation between LLP and LS on the basis of clinical features was in accordance with histopathologic differentiation, though 5 cases presented inconsistent findings between clinical and histopathologic impressions.

Linear lesions frequently occur in patients with LP or other dermatoses and are attributed to scratching as a consequence of pruritus (Koebner phenomenon)[10]. However, the peculiar patterns that reveal isolated, relatively wider and longer linear lesions involving as long as whole length of a limb do not seem to be induced by the Koebner phenomenon[11,12]. In our cases, the most patients denied trauma or scratching prior to the eruption and another lesions were hardly seen in remote sites of the body.

It is generally accepted that there is usual correlation between the distribution of childhood LS and the line of Blaschko[4]. Yoshihito et al[13] suggested that though LS in adults are rare and has some different features when compared with that in childhood (pruritus, more inflammatory nature and tendency to recurrence), its distributions are also consistent on Blaschko's lines. There has been relatively few suggestion about the pattern of distribution in LLP, but recently some evidences were reported that LLP might follow Blaschko's lines[14]. Taieb et al[4] suggested the acronym BLAISE (Blaschko linear acquired inflammatory skin eruption) or blaschkitis to describe acquired inflammatory skin lesions occurring diffusely or locally along Blaschko's lines. The current embryologic interpretation of Blaschko's lines is that they correspond to the direction of growth of clones of cutaneous cells (epidermal and dermal) derived from a limited number of precursor as a result of postzygotic genomic event[15,16]. On the trunk, the lines follow a bizarre S-shape on the abdomen, V shape near the posterior midline, and inverted U shape from the breast to the upper arm, which is due to longitudinal growth and the flexion of the embryo[17]. In our cases, two cases of LSs involving trunk showed lesions following Blaschko's lines, but in other cases involving extremities, it was not obvious whether the distributions were consistent with Blaschko's lines, for the lines are not so characteristic in extremities.

Our study confers some diagnostic references differentiating LLP and LS, but both diseases are not always clearly differentiated on the clinical or histopathologic basis only, and we found considerable discrepancy between clinical impression and histopathologic findings in 3 cases of LLP. These questionable conditions were previously proposed by several authors[3,5,7]. Rubio et al[3] reported a case of unsolved diagnostic problem which revealed some mixed findings of LLP and LS, and they suggested an intermediate form between these two entities. Herd et al[5] prompted the question whether the distinction of the two diseases was justified or whether they merely represented either end of a spectrum. The more fundamental basis of the disease process as well as clinical and histopathologic characteristics should be elucidated to resolve the question.

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