

Case Report / Olgu Sunumu

Linear lichen planus: a case report

Lineer lichen planus: olgu sunumu

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Abstract

Lichen planus (LP) is an inflammatory skin disease which is often seen in middle ages and affects the skin, mucous membranes, nails and scalp. Etiology is not clear and there are different clinical types of the disease which are named according to localization, distribution and morphological characteristics of the lesion. Linear lichen planus (LLP) is a rare variant of LP that is characterized by lichenoid, pruritic, violaceous papules, arranging in a linear pattern. Unilateral linear strip-like or segmental lesions follow the lines of Blaschko but does not demonstrate dermatomal distribution. Lichen striatus, linear lichen nitidus, inflammatory linear verrucous epidermal nevus, linear psoriasis, lichen simplex chronicus, incontinensia pigments, linear contact dermatitis can be considered in differential diagnosis. In this case, a case of 24 year-old woman with lesions on left arm is presented and differential diagnosis of this rare clinical form is discussed.

Keywords: Linear lichen planus

Özet

Liken planus (LP), orta yaşlarda sık olan ve deri, mukoza, tırnak ve saçlı deriyi tutan inflamatuvar bir deri hastalığıdır. Etiyolojisi belli olmayan bu hastalığın, lezyonlarının lokalizasyon, yayılım ve morfolojik özelliklerine göre farklı klinik tipleri tanımlanır. Lineer liken planus (LLP) bu hastalığın nadir bir formudur. LLP lineer olarak dizilen likenoid, pruritik, violasöz papüller ile dikkati çeker. Unilateral lineer şerit şeklinde veya segmental lezyonlar Blaschko çizgilerini izler ama dermatomal yayılım göstermez. Liken striatus, linear liken nitidus, inflamatuvar lineer verrüköz epidermal nevus, lineer psoriasis, liken simpleks kronikus, inkontinensiya pigmenti, lineer kontakt dermatit ayırıcı tanıda düşünülmelidir. Bu olgu sunumunda 24 yaşında bir kadının sol kolundaki lezyonların ayırıcı tanısı ve nadir görülen LLP hastalığının klinik özellikleri irdelenmiştir.

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Anahtar sözcükler: Lineer liken planus

Introduction

Lichen planus is an inflammatory disease that has an acute initiation, but mostly expresses a chronic continuation; and, can be present at skin, mucosa, scalp and nails. Although the etiology and pathogenesis are not clearly defined, the chief autoimmune result is basal keratinocyte apoptosis induced by CD8 T-lymphocytes. Viral and bacterial antigens, metal ions, medications and physical factors are accepted as the potential triggering factors of the autoimmune pathway [1]. Even though the prevalence is higher in the middle age group, the cases presented at childhood and adolescence is not at a negligible frequency [2]. There is no difference in prevalence amongst different genders; and, although the condition is known to be self-limiting between average durations of 1 month to 7 years, the presentation could also be continuous [3, 4]. Typical lesion in the classical form is in the shape of violet papules or plaques, in polygonal shape, with smooth surface and sharp margins of few millimeters. Reticular, white squamas named as Wickham lines, is observed on cutaneous and oral lichen planus lesions. Primary predicted presentation regions are lower and higher extremities; the most frequently affected regions are especially extensor surface of the lower extremities, volar surface of the upper arm, flexor of the wrist and ankles. Trunk and lumbar regions are the other frequently affected locations, face is generally not affected [1, 2]. Although an accompanying rash is present at approximately 80% of the patients, secondary excoriation is very rarely observed [4]. Isomorphic response secondary to trauma (Koebner phenomenon) is observed in approximately 50% of the patients at the acute phase [3]. As lichen planus is the autoimmune disease that most frequently affects the oral mucosa, oral symptoms are observed in 30-70% of the LP cases [2, 5]. Lesions in reticular, plaque-like, atrophic, papillary, erosive or bullous morphology can emerge [6]. As genital mucosa involvement is located in vulva and vagina in females, gans penis is the most frequent location of involvement in males [1]. The form of lichen planus with involvement of hair follicles is named as lichen planopillaris; and, could result with cicatricial alopecia [7]. Various nail signs could take place, such as longitudinal lines, pitting, pterygium, trachonychia, onycholysis, onychirrhaxis, koilonychia, subungual hyperkeratosis, onychomadesis, thinning of the nail plaque, depending on the nail involvement [8]. Lichen planus can be observed in different clinical manifestations, according to distribution and morphology of the lesions (Tables 1-3) [2].

Table 1. Distribution of lesions

Linear lichen planus	Linearly distributed lichenoid lesions
Blaschkoid LP	Lesions follow Blaschko lines
Zosteriform LP	Lesions follow dermatomal lines
Inverse LP	Lesions settle in intertriginous areas, might not be squamous
Mucosal LP	White reticular plaques in the mucosal region
Lichen planopilar	Follicular invasion on the scalp, secondary cicatricial alopecia

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Table 2. Morphology of lesions

Hypertrophic LP	Hypertrophic squamous pruritic nodules in the pretibial area
Bullous LP	Vesicles and bullae on the LP lesions
Actinic LP	Raised hyper pigmented atrophic plaques in sun-prone regions
Anular LP	Violet annular atrophic plaques
Erosive LP	Painful erosive or ulcerative lesions mostly in oral mucosa, can create scar
LP pigmentosus	Hyperpigmented lichenoid plaques on sun-exposed or intertriginous areas
Invisible LP	Presence of a rash without any clinical symptoms, outcome of Wood's lamp examination of skin biopsy taken from lesions is concordant with LP

Table 3. Overlap Syndromes

LP pemphigoides	Co-occurrence of LP and bullous pemphigoid
LP erythematosus	Co-occurrence of LP and lupus erythematosus

Even though, generally, clinical signs are sufficient for the diagnosis, typical histopathological features are helpful in the differential diagnosis. Orthohyperkeratosis, satellite necrotic keratinocytes named as Civatte bodies, wedge shaped hypergranulosis, saw tooth shaped irregular acanthosis, band shaped lymphohistiocytic infiltration in the dermoepidermal junction are the characteristic histopathological signs [4].

Case

24 year-old female patient presented with erythema and rash of the left arm that has been ongoing for two months. This patient neither had past medical history of any systemic or dermatological conditions, nor had a history of contact with any specific materials. In the dermatological examination, slightly squamous, erythematous papules were found at the flexor surface of the left arm, localized at distal of the axillary region (Figures 1 and 2). No pathological symptoms were found at the scalp, oral mucosa and nails. The skin biopsy was taken from the lesion for differential diagnosis of lichen planus, lichen striatus, linear lichen nitidus, inflammatory linear verrucous epidermal naevus, linear psoriasis, linear lichen simplex chronicus, incontinentia pigment and linear contact dermatitis; the results were coherent with lichen planus (Figure 3). No pathological signs were detected in the investigations including full blood count, fasting blood glucose, hepatitis B and C. After topical steroid treatment with medium potency was initiated for this patient with LLP diagnosis, complaints ceased within the first month and new lesions did not generate in the one year follow-up period.



Figure 1. Lateral view of the lightly squamous, erythematous, linearly localized papules on the left upper arm.



Figure 2. Lateral view of the lightly squamous, erythematous, linearly localized papules on the left upper arm.

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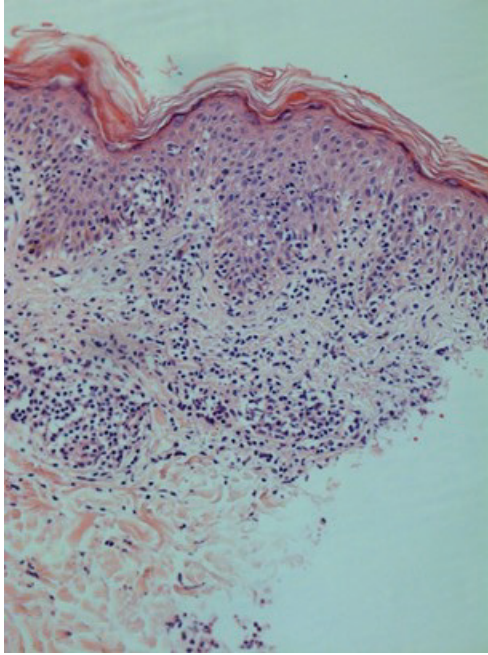


Figure 3. Histopathological examination demonstrated hyperkeratation, focal hypergranulation and Civatte bodies in epidermis as well as medium-severity lichenoid interface dermatitis in the dermis.

Discussion

Linear LP is a rare LP variant commonly observed in childhood or the adolescent period. Lesions that are clinically line-shaped, could demonstrate distribution that is segmental, zosteriform or following the Blaschko lines. Lesions that are commonly observed as typical papules with a smooth surface, could also be purpuric, vesicular, hyperkeratotic or resembling annular morphology [1]. It is seen that there is no dermatomal arrangement in linear LP, but Blaschko lines are trailed. In the literature, it is reported that 0.2-0.6% of the LP cases is linear LP [9]. As generally zosteriform LP has a dermatomal localization and linear LP is used for defining lesions localizing on the Blaschko lines, discrimination of linear and zosteriform variants is not always possible; and differentiation based solely on clinical signs may not be accurate [10]. Linear LP defines spontaneously appearing linearly arranged lichen lesions in the shape of narrow lines that follow the Blaschko lines, which does not follow vascular or neuronal structures in skin, with no previous herpes infection or trauma history [9].

Mizukawa et al., reported the presence of the VZV antigen with eccrine epithelial localization, from the immunohistochemical examinations performed on the biopsies taken from the lesions, in two cases with positive VZV IgG titers and no herpes zoster history; and emphasized that zosteriform LP could be pathogenically different to linear LP [10]. Lutz et al. reported two zosteriform cases that presented on the location of healing herpes zoster, lesions following peripheral cutaneous nerves and nerve branches, where no herpes simplex virus or varicella zoster virus have been detected by the polymerase chain reaction

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(PCR) [11]. Zosteriform lichen planus developing over healed herpes infection is reported in the isotopic response of Wolf [12, 13]. Kalkan et al. presented a 40 year-old male patient with an S-shaped linear elongation on the Blaschko line localized in left arm and right trunk [9]. Horowitz et al. reported a 30 month-old child case diagnosed with linear LP elongated over Blaschko lines, localized in abdominal and femoral region [14]. Batra et al. reported a 50 year-old male case diagnosed with linear LP with a distribution following the Blaschko lines, located unilaterally at the lower extremity [15]. Krasowska et al. reported a case of unilateral linear LP following Blaschko lines [16]. In the literature, adolescent cases with linear LP diagnosis that is morphologically similar to epidermal naevus but histopathologically diagnosed with LP are reported [17]. A 32 year-old female case was reported with diagnosis of Blaschkoid LP, localized at the left trunk [18]. A case of 30 year-old male with unilateral Blaschkoid diagnosis, localized in half-body has been reported [19].

This case was found as worthy of presentation as it is a very rare clinical form of lichen planus and attention was paid to the thought of this case in differential diagnosis of erythematous, squamous diseases with a linear distribution.

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