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Atypical Forms of Lichen Planus

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Abstract: Lichen planus (LP), the most typical and best characterized lichenoid dematosis, is an idiopathic inflammatory skin disease affecting the skin and mucosal membranes. Classic LP typically presents as pruritic, polygonal, violaceous flat-topped papules and plaques; many variants in morphology and location also exist, including oral, nail, linear, annular, atrophic, hypertrophic, inverse, eruptive, bullous, ulcerative, lichen planus pigmentosus, lichen planopilaris, vulvovaginal, actinic, lichen planus-lupus erythematosus overlap syndrome, and lichen planus pemphigoides. Clinical presentation of the rarer variant lesions may be largely dissimilar to classic LP and therefore difficult to diagnose based solely on clinical examination. This article contains an array of reports on the forms in presentation and successful management of LP and its variants. A familiarity with LP and its variants is important in achieving timely recognition and management of the dermatoses.

Keywords: lichen planus, hypertrophic, atrophic, inverse ,mucosal, linear, annular, bullous, lichen planus pemphigoides, ulcerative, lichen planus pigmentosus, lichen planopilaris, vulvovaginal, actinic.

Introduction: Lichen planus (LP) is a unique inflammatory cutaneous and mucous membrane reaction pattern of unknown etiology.The disorder occurs most commonly in adulthood(mean age of onset is 40.3 years in males and 46.4 years in females). The main eruption clears within 1 year in68% of patients, but 49% of eruptions recur. Althoughthe disease may occur at any age, it is rare in children younger than 5 years. Approximately 10% of patients havea positive family history. This supports the hypothesisthat genetic factors are of etiologic importance. Liverdisease is a risk factor for LP although not a specific marker of it. Cutaneous and oral LP may be associated with hepatitis C virus (HCV)–related, chronic, active hepatitis.Although it is not clear that hepatitis C causes lichen planus, some authors believe that hepatitis C may induce alterations in cytokine and chemokine expression, leading to the development of LP. Basal keratinocytes in LP show increased expression of intercellular adhesion molecule-1 (ICAM-1), which interacts with CD4+ and especially CD8+ T lymphocytes. This shift to the T-helper-1 arm of cellular immunity leads to basal keratinocyte apoptosis. Patients with cutaneous lichen planus are not at increased risk for cutaneous cancer, however patients with oral lichen planus (OLP) and vulvar lichen planus are at increased risk for cancer, especially squamous cell carcinoma (SCC). [1].

Purpose of the study Study atypicaly forms of lichen planus based on the results of clinical diagnoses and treatment.

Results and analysis

Hypertrophic Lichen Planus: Hypertrophic LP usually occurs on the shins but may occur anywhere. The anterior lower leg below the knee is the sole area of involvement in most patients. The typical lesions are verrucous plaques with variable amounts of scale. At the edges of the plaques, small, flat-topped, polygonal papules may at times be discovered. Superficial inspection of the lesion often suggests psoriasis or a keratinocytic neoplasm rather than LP, but the typical appearance resembling rapidly cooled igneous rock (igneous rock sign) may be useful in suggesting LP over keratinocytic neoplasms. The lesions are of variable size but are frequently several centimeters in diameter and larger than the lesions of classic LP. Dermoscopy of hypertrophic LP may demonstrate pearly white areas and peripheral striations, which can help distinguish it from mimickers in some cases; however, clinical diagnosis may be difficult, and biopsy is often required. Histologically, the pseudoepitheliomatous keratinocyte hyperplasia may be marked, leading to the erroneous diagnosis of squamous cell carcinoma (SCC). Eosinophils are much more often present in the dermal infiltrate of hypertrophic LP than classic LP. True SCC may also evolve from long-standing hypertrophic LP, over long as 11–12 years. In addition, keratoacanthoma-like proliferations may occur in lesions of hypertrophic LP. This has also been called "hypertrophic lichen planus–like reactions combined with infundibulocystic hyperplasia." Hypertrophic LP is chronic and often refractory to topical therapy. Hypertrophic lupus erythematosus (LE) resembles hypertrophic LP both clinically and histologically. Hypertrophic LE tends to affect the distal extremities, face, and scalp. The finding of continuous granular immunoglobulin on DIF strongly suggests a diagnosis of hypertrophic LE rather than LP.

Atrophic lichen planus.

Atrophic LP may represent a resolving phase of LP, given the history of the lesions: papules coalesce to form larger plaques that over time become atrophic centrally, with residual hyperpigmentation. The clinical appearance of atrophic LP is likely a result of thinning of the epidermis rather than degeneration of elastic fibers, and the epidermal atrophy may be accentuated by the use of potent topical corticosteroids Common sites of involvement include the intertriginous zones and the lower extremities (Fig. 11.9A). Simultaneous occurrence of LP and lichen sclerosus has been reported as has an annular atrophic variant with complete loss of elastic fibers in the center of the lesions**.**

Inverse lichen planus.

In this unusual variant, an inverse distribution pattern is observed. Pink to violaceous papules and plaques appear in intertriginous zones (axillae > inguinal and inframammary folds) and less often in the popliteal and antecubital fossae [11]. Occasionally, there are LP lesions elsewhere on the body. Hyperpigmentation is usually present as well and it may be the sole manifestation, leading to overlap with LP pigmentosus**.**

Mucosal lichen planus.

Lesions confined to the mouth, or with minimal accompanying skin involvement, are not uncommon [1–6], and account for about 15% of all cases. Mucosal lichen sclerosis/lichen planus overlap syndrome can also be observed [1]. The lesions do not differ from those found in connection with skin lesions, but, being confined to the mouth, may lead to great difficulty in diagnosis. They are often referred first to a dental surgeon. Distinct clinical subtypes such as reticular, atrophic, hypertrophic and erosive forms are well recognized, and more than one type may be present [2]. On the tongue and buccal mucosa, lesions are most likely to be mistaken for leukoplakia and on the gum margin for gingivitis or chronic candidiasis; the latter may coexist. Other conditions that must be excluded are 'smoker's patches', which characteristically involve the palate, and white-sponge naevi, in which the mucous membrane is thickened, irregularly

folded and feels soft to the touch. These occur mainly on the floor of the mouth and histologically many of the prickle cells in the epidermis are vacuolated. It is important to bear in mind the possibility of a lichenoid drug reaction in patients with oral lichenoid changes. Oral lichenoid reactions may be asymmetrical on the buccal mucosa and occur adjacent to dental amalgam fillings. If patch testing reveals mercury allergy, changing to another type of filling may prove beneficial [9–14]. Very occasionally, LP lesions extend to the larynx or oesophagus [15–19]. Oesophageal LP may result in dysphagia and the formation of benign strictures. In young men, the lesions of LP are sometimes restricted to the genitalia and/or mouth [20,21]. Genital lesions, which are usually characteristic, may be present on the penile shaft (see Figure 37.2), glans penis, prepuce or scrotum. Ulceration is very unlikely, and syphilis can usually be excluded without difficulty. The presence of buccal mucosal lesions will usually confirm the diagnosis. Circumcision may be helpful in clearing up LP [20]. Lesions on the female genitalia are fairly common [22–29]; they may occur alone, be combined with lesions in the mouth only, or be part of widespread involvement. The clinical presentation of LP of the vulva spans a spectrum from subtle, fine, reticulate papules to severe erosive disease accompanied by dyspareunia, scarring and loss of the normal vulvar architecture. The condition should be distinguished from lichen sclerosus or leukoplakia, but this may be difficult when there is coexisting atrophy or vaginal stenosis. Diagnostic criteria have been proposed recently [30] and include well-demarcated erosions/erythematous areas at the vaginal introitus, the presence of a hyperkeratotic border to lesions and/or Wickham's striae in the surrounding skin, symptoms of pain/burning, scarring/loss of normal architecture, the presence of vaginal inflammation and the involvement of other mucosal surfaces. Histologically, a well-defined inflammatory band involving the dermal–epidermal junction and consisting predominantly of lymphocytes and signs of basal layer degeneration are seen. The association of erosive LP of the vulva and vagina with desquamative gingivitis has been termed the vulvo-vaginal–gingival syndrome [21,22]. Coexisting vulval lichen sclerosus and lichenoid oral lesions have been described [31].

Linear lichen planus.

Although linear lesions frequently occur in sites of scratching or trauma in patients with LP as a result of the Koebner phenomenon, the term linear LP is usually reserved for lesions that appear spontaneously within the lines of Blaschko. This form has also been referred to as zosteriform, but with the exception of LP developing within the site of previous herpes zoster, the distribution pattern of LP is generally not dermatomal. The possibility exists that when LP has a strictly dermatomal pattern, it may have been preceded by "zoster sine herpete".

Bullous lichen planus and lichen planus pemphigoides.

Lichen ruber pemphigoides was first described by Kaposi in 1892.

Bullous LP and LP pemphigoides were in the past differentiated solely on clinical and histological criteria [1], but can now be differentiated using immunofluorescence (IMF) procedures and immunoelectron microscopy. In bullous LP, blisters arise only on or near the lesions of LP, as a result of severe liquefaction degeneration of the basal cell layer [3]. Histologically, there is subepidermal bulla formation with typical changes of LP, and direct and indirect IMF are negative [2,3]. The eruption is usually only of short duration [2]. In LP pemphigoides the LP tends to be acute and generalized and is followed by the sudden appearance of large bullae on both involved and uninvolved skin [4–7]. Occasionally, even in LP pemphigoides, blisters may arise only on the lesions of LP [8]. LP pemphigoides has been precipitated by psoralen and

UVA (PUVA) [9] and has evolved into pemphigoid nodularis [10]. An LP pemphigoides-like eruption has been reported to overlap with paraneoplastic pemphigus [11,12]. In LP pemphigoides, the histology shows a subepidermal bulla with no evidence of associated LP [2]. Direct IMF shows linear basement membrane zone deposition of IgG and C3 in perilesional skin

[2,7]. Immunoelectron microscopic studies reveal deposition of IgG and C3 in the base of the bulla and not in the roof as found in bullous pemphigoid[13].

Immunoblotting data have revealed that circulating autoantibodies in LP pemphigoides react with an epitope within the Cterminal NC16A domain of bullous pemphigoid 180 kDa antigen, and also with a 200 kDa antigen detected in bullous pemphigoid [14–18]. It seems that epidermal damage from liquefaction degeneration in LP exposes basement membrane antigens, and a consequent stimulation of autoantibody production. The mean age of patients with LP pemphigoides is lower than that of those with classic bullous pemphigoid, and the course of the disease also tends to be less severe.

Annular lichen planus

Although small annular lesions are common in LP, cases showing a few large annular lesions only are unusual. They may be widely scattered, and usually have a very narrow rim of activity and a depressed, slightly atrophic centre [23]. Much less often, the margin is wide and the central area is quite small. Annular lesions are characteristically found on the penis sometimes associated with lesions on the buccal mucosa. The differential diagnosis includes granuloma annulare distinct entity termed annular lichenoid dermatitis of youth [2,3,24,25] has been described, characterized by persistent, asymptomatic erythematous macules and round, annular patches with a red-brownish border and central hypopigmentation, mostly distributed on the groin and flanks, in children and adolescents. Histology reveals a lichenoid dermatitis with necrosis/apoptosis of the keratinocytes limited to the tips of rete ridges**.**

Folicullar lichen planus.

Follicular lesions usually appear during the course of typical LP, but occasionally they predominate and diagnosis may then be difficult. An absence of arrector pili muscles and sebaceous glands, a perivascular and perifollicular lymphocytic infiltrate in the reticular dermis and mucinous perifollicular fibroplasia within the upper dermis with an absence of interfollicular mucin, and superficial perifollicular wedge-shaped scarring are all characteristic features of the histology [1]. Presentation with alopecia of the trunk is recorded [2]. Follicular lesions occurring in the scalp are accompanied by some scaling and are likely to lead to a scarring alopecia. Very rarely, the scalp alone is involved. The so-called Graham Little–Piccardi–Lassueur syndrome comprises the triad of multifocal scalp cicatricial alopecia, non-scarring alopecia of the axillae and/or groin, and keratotic lichenoid follicular papules [3–8]. The clinical, histological and immunofluorescence overlap between this syndrome and LP with follicular involvement (lichen planopilaris) suggest that both are variants of LP. Follicular LP must be distinguished by biopsy from keratosis pilaris, Darier disease, follicular mucinosis, lichen scrofulosorum and, in the scalp, from lupus erythematosus.

Lichen planus pigmentosus.

LP pigmentosus typically presents as brown to gray–brown macules in sun-exposed areas of the face and neck, usually with no preceding erythema and often evolving into diffuse or reticulated pigmentation.This variant favors individuals with skin types III and IV, in particular those from South Asia, Latin America, and the Middle East. Involvement of intertriginous sites is occasionally observed and a linear distribution following the lines of Blaschko has also been described. In the photodistributed form, topical application of mustard oil, which contains a potential photosensitizer, has been implicated as a possible trigger. Given the similarity of histologic findings, distinction from erythema dyschromicum perstans (EDP) is based primarily upon clinical features[8].

In EDP, there is often truncal involvement, a younger mean age ofonset, and lack of either diffuse pigmentation or coexisting LP lesions. The latter is seen in ~20% of patients with LP pigmentosus. Occasionally, small nests of keratinocytes are seen within the basal layer and these may be confused with nests of melanocytes.

Lichen planopilaris

In lichen planopilaris, involvement of the hair follicle is observed, both clinically and histologically. This variant is also called follicular LP and LP acuminatus. Multiple, keratotic plugs surrounded by a narrow violaceous rim are observed primarily on the scalp, although other hairbearing areas can also be affected [11-13]. The inflammatory process may result in scarring and loss of follicular structure, i.e. a permanent alopecia. Over time, the areas of involvement often "burn out" centrally and are indistinguishable from other causes of "endstage" cicatricial alopecia . However, examination of the periphery may reveal the primary lesions. Women are more frequently affected than men, and this form may occur alone or with typical LP lesions elsewhere.

A variant of lichen planopilaris known as Graham-Little–Piccardi–Lassueur syndrome is characterized by the triad of: (1) non-cicatricial loss of pubic and axillary hairs and disseminated spinous or acuminated follicular papules [14-15] typical cutaneous or mucosal(2) LP; and (3) scarring alopecia of the scalp with or without atrophy. These features need not be present simultaneously.

Another more recently recognized variant of LP of the scalp is frontal fibrosing alopecia which occurs primarily in older women and can affect the eyebrows as well.

Ulcerative lichen planus.

Ulcerations can occur within palmoplantar lesions of LP, particularly those on the soles. Palmoplantar LP is not as rare as was once thought and usually appears between the third and fifth decade of life. Although palmoplantar LP is more common in men than in women, ulcerative LP prevails in female patients. Typical LP lesions may be present in additional sites of the body. The ulcers are intensely painful and often recalcitrant to conventional therapy. Chronic ulcerative lesions are at risk of developing SCC.

Vulvovaginal lichen planus.

LP of the vulva can present with several clinical variants, but the most common appears to be erosive disease. Vaginal involvement occurs in up to 70% of women with erosive vulvar LP, and because there is often oral mucosal involvement as well, the term "vulvovaginal–gingival syndrome" has been introduced. The differential diagnosis of vulvovaginal LP includes lichen sclerosus and blistering diseases . Since scarring may be a sequela of vulvovaginal LP, patients should be carefully monitored for the development of SCC, including after the resolution of active disease.

Actinic lichen planus.

This variant is reported under a variety of names, including LP actinicus,

LP subtropicus, LP tropicus and lichenoid melanodermatitis. Although the majority of reported patients have been from Middle Eastern countries, this variant has been observed worldwide. Most patients are young adults or children, but there is no gender predilection. Onset of this variant is typically during the spring and summer, and lesions primarily involve sun-exposed skin of the face, followed by the neck and dorsal surfaces of the hands and arms. The lesions usually consist of red–brown plaques with an annular configuration, but melasma-like hyperpigmented patches have been observed.[7-9]. In temperate climates, spontaneous improvement may occur during the winter months.

Conclusion.

Lichen planus is an inflammatory skin disease with characteristic clinical and histopathological findings. In addition to classic LP, a myriad of LP variants exist, including oral, linear, annular, atrophic, hypertrophic, inverse, eruptive, bullous, ulcerative, LP pigmentosus, lichen planopilaris, vulvovaginal, actinic, and LP pemphigoides. The pruritic, polygonal, violaceous, flat-topped papules and plaques of classic LP are the most common presentation of the disease,

but morphology and location vary greatly among the variants. However, histopathological findings among the variants are largely consistent. Therefore, while clinical examination may be sufficient for diagnosis in some cases, histological examination is often valuable in confirming diagnosis of LP variants. Management of classic LP and its variants is mostly consistent, with topical TCS as a first-line therapy among a diverse array of treatment options that have been reported in the literature with varying degrees of success. Ultimately, familiarity with the characteristics of LP and its atypical variants is essential in achieving timely recognition and effective management.

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