

Pityriasis Lichenoides

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Pityriasis lichenoides (PL) is a skin disorder of unknown etiology that mainly affects children and young adults. Pityriasis lichenoides is perhaps best considered a disease spectrum with acute and chronic types: pityriasis lichenoides et varioliformis acuta (PLEVA) and pityriasis lichenoides chronica (PLC). Pityriasis lichenoides et varioliformis acuta, the acute variant, is characterized by the onset of papulovesicular eruptions that evolve into necrotic lesions over the trunk and extremities, and hence the name “varioliformis,” which means “like variola” (like smallpox). Pityriasis lichenoides chronica, the chronic form, involves recurrent crops of papules that may last from several months to several years. Both types are further characterized by independent evolution of each lesion, so that lesions in all different states of progression are found next to each other in a random array. A third disease, lymphomatoid papulosis (LP), is also included in this grouping by some authors. Pityriasis lichenoides is regarded under the rubric of parapsoriasis, although it is distinct from large and small plaque parapsoriasis, the other principle entities that comprise the parapsoriasis grouping.^{1,3}

History

Pityriasis lichenoides was first described in 1894 by Jadassohn⁴ and Neisser⁵ in two separate reports. The chronic form was described again by Juliusberg⁶ in 1899 and given the name PLC. In 1902, Brocq⁷ classified PL under the group parapsoriasis, which he proposed has guttate, plaque, and lichenoid variants. Mucha⁸ in 1916, and later Habermann⁹ in 1925, described acute PL as a disease entity distinct from the other parapsoriasis, including PLC. In 1981, Lambert and Everett¹ reclassified the parapsoriasis group to clearly delineate

PL from small and large plaque parapsoriasis, including digitate dermatosis, xanthoerythrodermia perstans, and the rare entity, retiform parapsoriasis.

Incidence

Pityriasis lichenoides most commonly occurs in children and young adults; however, its precise incidence is unknown. Both PLC and PLEVA appear to be about as common in males as in females, with PLC occurring more frequently. Pityriasis lichenoides et varioliformis acuta most often occurs during the second or third decade of life. However, the disease may also arise in children and the elderly, and may even be evident at birth.¹⁰ There is no apparent racial or geographic predisposition.^{1,2}

Characteristics

Pityriasis lichenoides is perhaps best considered as a disease spectrum with three forms: PLEVA, PLC, and LP. There is controversy regarding the inclusion of LP into the PL spectrum. Pityriasis lichenoides et varioliformis acuta, also known as Mucha-Habermann disease, is characterized by the acute onset of firm, pinkish papules, 2- to 10-mm in diameter, associated with dense inflammatory infiltrates.^{1,11} The papules commonly involve glabrous skin over the entire body surface with relative uniformity (Figures 1 to 3).^{12,13} The lesions generally occur on the anterior trunk and flexor surfaces and tend to be more numerous on the proximal rather than distal parts of the extremities.¹⁴ An important diagnostic sign is that lesions at all different stages are present at the same time in close proximity to each other. The face, palmoplantar surfaces, and mucous membranes are usually spared. Pruritus is sometimes present, but the lesions are more often asymptomatic.¹²⁻¹⁴

As PL progresses, the papules become hemorrhagic, vesicular, necrotic, and crusted, leaving scars with postinflammatory hyperpigmentation or hypopigmentation.¹⁵ The patient's general health is usually unaffected; however, low-grade fever, generalized

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FIGURE 1. Recurrent crops of sessile pinkish papules.

lymphadenopathy, malaise, and headache are occasionally seen.² A rare acute febrile ulceronecrotic type of PLEVA, known as “febrile ulceronecrotic Mucha-Habermann disease,” has been noted to occur in children. This severe form of PLEVA is characterized by the acute onset of diffuse, coalescent, large, ulceronecrotic skin lesions often associated with a high fever. A mild eruption is usually observed before the acute fulminating course.^{2,3} Pityriasis lichenoides et varioliformis acuta usually follows an acute course that spontaneously resolves in several weeks. However, after an acute phase, PLEVA may change into PLC, which has a prolonged chronic course.¹⁴

Pityriasis lichenoides chronica is characterized by recurrent crops of lichenoid, reddish-brown papules that are covered by a light scale and generally involute within 3 to 6 weeks. This solitary micaceous scale on the papule can easily be scraped off to demonstrate a typical shiny brown surface. Papules tend to flatten into a hyperpigmented, gradually fading macule. The disease may last from several months to several years with frequent exacerbations and remissions.^{1,2,6,16,17} The differentiation between PLEVA and PLC is sometimes unclear. Patients may have lesions of both the acute and chronic form simultaneously, as well as transitional lesions. This suggests that PLEVA and PLC may be a continuous spectrum of the same disease process, rather than two distinct entities.^{2,12,13} Because of the frequent association between the two PL variants, Gelmetti *et al.*¹² suggested an alternative classification. They described three patterns based on anatomic distribution: a diffuse form involving

glabrous skin over the whole body surface, a central variety involving the neck, trunk, and proximal extremities, and a peripheral form with an acral distribution.^{12,13} The diffuse type usually has the shortest duration and the peripheral type has the longest.

Lymphomatoid papulosis, a PL variant, is characterized by the uninterrupted appearance of papulonecrotic, nodular, and occasionally large, plaque-like, self-healing eruptions that undergo exacerbations and remissions for an indefinite period of time. The lesions show some histologic features resembling malignant lymphoma, but are clinically benign.¹⁸⁻²³ It may be clinically difficult to distinguish LP from PLEVA. However, LP lesions tend to be nodules and plaques, whereas PLEVA lesions are papular, more necrotic than vesicular, less numerous, and evolve slowly. The most significant difference between the two is that about 10 to 20% of patients with LP eventually develop mycosis fungoides (MF), Hodgkin’s disease, or non-Hodgkin’s lymphoma.¹⁶ Conversely, patients with MF may develop lesions that are clinically and histologically identifiable as LP.²⁴

Histopathology

The histology of the three entities comprising PL are distinct.^{21,24-28} In PLEVA, the epidermis shows intercellular and intracellular edema, focal necrosis, and, occasionally, neutrophils in the stratum corneum. The dermis has pronounced perivascular and diffuse infiltrates, consisting mainly of mononuclear cells, and extravasation and diffusion of erythrocytes into the epidermis. The vascular changes consist of en-



FIGURE 2. Close-up of the papules in Figure 1.

endothelial swelling and hemorrhage, with small deposits of fibrin present within vessel walls.

Pityriasis lichenoides chronica characteristically features dermal–epidermal interface dermatitis in all stages of the lesions. Lymphocytes and histiocytes, melanophages, and extravasated erythrocytes are the cells most commonly observed. Focal mild parakeratosis may also be seen.²⁹

Both PLEVA and PLC show a distinctive pattern in their dermal inflammation, which is wedge-shaped. The apex of the wedge is deep and the base is at the dermal–epidermal interface, which is covered by the epidermal component of the disease. Within this wedge, the inflammation is often focused around blood vessels.

In LP, two types of atypical cells, histiocytes and lymphocytes, are seen, with either one or the other predominating. When histiocytes predominate, the condition is called “Willemze Type A,” and when lymphocytes are prevalent, the condition is known as “Willemze Type B.” In Type A LP, the atypical cells are characteristically large with pleomorphic, bizarrely-shaped nuclei, prominent nucleoli, and abundant cytoplasm. Multinucleated cells resembling Reed-Sternberg cells may also be observed. In addition, many neutrophils and monocytes are seen in the infiltrate. In Type B LP, the atypical cells are smaller, but are still relatively large compared with other neoplasms of lymphocytes, and neutrophils are less common.^{18,19}

Etiology

The underlying etiology and pathogenesis of PL remain unknown.^{25,30,31} Pityriasis lichenoides et vario-

liformis acuta, PLC, and LP have several immunohistologic similarities.³⁰ All three diseases are characterized by an infiltrate of activated T cells mixed with macrophages in the epidermis and dermis.²⁷⁻³² Both PLEVA and LP have been shown, in some cases, to undergo rearrangement of T cell receptor genes, indicating a clonal T cell lymphoproliferative process. This finding supports the hypothesis that these three entities represent a spectrum of a single disease.

The presence of IgM and C3 in the dermal vessel walls and along the dermal–epidermal junction has been demonstrated in patients with PLEVA. Serologic, epidemiologic, and therapeutic evidence suggests that PLEVA may be the result of a hypersensitivity reaction to an infectious agent,³² although a specific agent has not been isolated with reproducible results in patients with PLEVA. The above data suggest that PL may be due to persistent antigen stimulation and resulting immune complex deposition.

Differential Diagnosis

Pityriasis lichenoides chronica and PLEVA may be difficult to differentiate from other papular eruptions. The individual lesions of PLC may resemble small plaque parapsoriasis. However, small plaque parapsoriasis can usually be differentiated since its lesions are flat rather than nodular. The differential diagnosis of PLEVA may include lymphomatoid papulosis, Gianotti-Crosti syndrome, varicella, erythema multiforme, leukocytoclastic vasculitis, and secondary syphilis.² Although LP may clinically resemble PLEVA, the lesions of LP are usually larger, less numerous, and have a slower evolution.



FIGURE 3. Coalescent papules forming a plaque.

Histologically, LP shows a dense lymphoma-like infiltrate, which in Type A LP is Ki-1 antigen-positive.^{33,36} Secondary syphilis may also be considered. However, the results of serologic tests (FT-ABS) are conclusive, and the palms and soles are often involved.³³ Gianotti-Crosti disease can be clinically differentiated from PLEVA by its lack of necrotic lesions and the presence of lymphadenopathy and acute hepatitis.^{2,37} Histologically, Gianotti-Crosti disease does not show erythrocyte exocytosis or epidermal necrosis as seen in PLEVA. In addition, erythema multiforme and varicella typically involve the mucous membranes. On histologic examination, erythema multiforme and PLEVA both contain necrotic keratinocytes and mononuclear cell infiltrates; however, PLEVA characteristically has a wedge-shaped infiltrate extending deep into the reticular dermis.²

Course

Pityriasis lichenoides is usually a self-limited process that evolves after several weeks to months. Pityriasis lichenoides et varioliformis acuta may change to PLC after an acute phase.¹ Pityriasis lichenoides chronica usually lasts considerably longer than PLEVA, in some instances, for years, with frequent exacerbations and remissions. Lymphomatoid papulosis has a well-documented potential to progress to MF or another lymphoma in about 10 to 20% of cases.

Treatment

The treatment for PL includes topical corticosteroids, oral antibiotics, psoralen-ultraviolet A

(PUVA) and ultraviolet B phototherapy, and methotrexate.^{2,11,34-37} Topical corticosteroids and antihistamines are given for pruritus, but do not affect the disease course.² Tetracycline and erythromycin^{11,14,37} have both been successfully used to induce clearing of PL. The anti-inflammatory effect of antibiotics, related to their inhibition of monocyte chemotaxis, has been speculated to be the mechanism of action of these drugs.² Children with PL should be treated with erythromycin rather than tetracycline due to the adverse effects of tetracycline on dentition. Ultraviolet B phototherapy in children has also proved a valuable and safe therapeutic option.³⁵ In some cases, methotrexate may be needed to clear PL, particularly the febrile ulceronecrotic PLEVA variant, when other more conservative measures have failed.³² Combination therapy, such as erythromycin and PUVA, may be useful.

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