

Purplish, pruritic papules on the limbs

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A 46-year-old white man presented with an acute 3- to 4-week history of an intensely pruritic eruption that mainly involved the limbs. Individual lesions appeared to predominate around the wrist flexures. Examination revealed multiple, flat, purplish lesions, 5 to 15 mm in diameter, with coalescence in areas of scratching (*Figure 1*). Whitish plaques were

seen on the oral mucous membrane (*Figure 2*). The patient was otherwise well, although on further questioning he stated that he felt less energetic than usual. He reported no recent illness or medication ingestion.

What is the diagnosis, and what further laboratory tests are indicated?



Figure 1. Purplish, flat-topped papules on the wrist; a few are in a linear arrangement.



Figure 2. A white reticulated pattern can be seen inside the right cheek; lesions cannot be removed with a tongue blade.

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DIAGNOSIS: Lichen planus.

DISCUSSION

Lichen planus is a T-lymphocyte-mediated disease that affects both the skin and mucous membranes (1). Typically, skin lesions are pruritic, purple, polygonal, flat-topped papules that preferentially involve the flexor surfaces (especially the wrist) and trunk and only rarely involve the face (2). Close examination of a papule with a magnifying lens reveals a network of gray streaks (Wickham's striae). Rare variants of lichen planus include lesions that are hypertrophic (on the shins), bullous (on the oral mucosa), and ulcerative or erosive (on the oral mucosa, palms, and soles). The isomorphic response, or Koebner's phenomenon, is characterized by the production of lesions at areas of trauma (i.e., scratches, bites, surgical sites, and healed lesions of herpes zoster) and occurs commonly in lichen planus. Koebner's phenomenon is a helpful clinical clue because it is found in only a few dermatological diseases (Table 1). Resolution of lesions may result in hyper- or hypopigmentation, frequently persisting for months to years.

Mucous membranes are involved in two thirds of patients with skin lesions and may be the sole area of involvement. The inside cheeks (buccal mucosa) characteristically have silver-white, pin-sized papules in a lacelike distribution. Lesions may also occur on the lips, palate, and tongue and less commonly on the vaginal mucous membranes. Rarely, oral lichen planus may ulcerate, producing chronic, painful erosions of the gums and mucosa. Long-standing mucosal lichen planus, especially oral ulcerative lichen planus, has been associated with carcinoma (3, 4). On the glans penis, minute, flat, polygonal papules may be arranged in rings.

Nail changes are seen in 5% to 10% of patients (2). Pterygium formation is distinctive in lichen planus (Figure 3). Other nail changes include longitudinal grooving, ridging, and splitting and a peculiar midline fissure. Proximal onycholysis (loosening) may occur with lichen planus.

Scalp lesions are seen infrequently. When present they may produce a scarring alopecia (2). Follicular lichen planus (lichen planopilaris) is characterized by spiny erythematous follicular papules in the scalp that result in a scarring alopecia. Scalp le-



Figure 3. Absent right thumbnail plate with tenting of skin and scar.

Table 2. Disorders and other conditions associated with lichen planus

Autoimmune disorders	Chronic liver disease
Alopecia areata	Hepatitis B and C
Bullous pemphigoid	Primary biliary cirrhosis
Dermatitis herpetiformis	
Dermatomyositis	Other
Diabetes mellitus	HLA-DR1 antigen
Hashimoto's thyroiditis	Thymoma
Morphea	Urolithiasis
Myasthenia gravis	
Pemphigus (foliaceus/vulgaris)	
Pernicious anemia	
Sjögren's syndrome	
Systemic sclerosis	
Ulcerative colitis	
Vitiligo	

Table 1. Dermatological diseases in which Koebner's phenomenon occurs

Contact dermatitis	Molluscum contagiosum
Darier's disease	Pellagra
Dermatographism	Perforating disorders
Eruptive xanthoma	Pityriasis rubra pilaris
Erythema multiforme	Porokeratosis
Henoch-Schönlein purpura	Psoriasis*
Kyrle's disease	Reactive perforating collagenosis
Lichen nitidus	Rhus dermatitis*
Lichen planus*	Verrucae*
Lichen sclerosus et atrophicus	Vitiligo

*Most frequently seen

sions may occur alone or with other lesions on skin or mucosal surfaces.

Very rarely, lichen planus has been described in the esophagus and the rest of the gastrointestinal tract.

Skin biopsy specimens reveal a "saw-tooth" epidermal hyperplasia and a lichenoid interface infiltrate with vacuolar alteration along the basement membrane. Necrotic keratinocytes are present along the basal layer of the epidermis and in the dermis (Civatte bodies). Biopsy specimens of drug-induced lichen planus may have a considerable number of eosinophils. Direct immunofluorescence reveals subepidermal clumps of immunoglobulin (Ig) M (less often IgG, IgA, or C3), which correspond to the Civatte bodies (2).

Numerous diseases have been associated with lichen planus (Table 2). Most recently the association with hepatitis, particularly hepatitis C, has been stressed in the literature (5, 6). Thus, all patients with a clinical or histological diagnosis of lichen planus should be evaluated with this in mind. In addition, medications have been associated with lichen planus-type drug eruptions (Table 3) (7).

Table 3. Medications frequently associated with lichen planus*

Antimalarials	Penicillamine
Arsenic	Phenothiazine derivatives
Chlorpropamide	Propranolol
Gold	Quinidine
Methyl dopa	Spirolactone
Naproxen	Thiazide derivatives

*Compiled from reference 7.

Treatment is primarily required for symptomatic patients (i.e., those with pruritus or pain from ulcers) or patients with generalized lichen planus. Corticosteroids (topical, intralesional, and systemic) are the mainstay of therapy. Retinoids (topical and systemic) (8–11), dapsone (12), cyclophosphamide (13), cyclosporine (14–16), metronidazole, hydroxychloroquine (17), tacrolimus (18), thalidomide (19), and phototherapy have also been helpful. The recent introduction of topical tacrolimus has been valuable for patients with intractable mouth involvement (18).

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