

# Zosteriform Lichen Planus: An Unusual Clinical Variant

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## Abstract

*Lichen planus (LP) is a relatively common papulosquamous dermatosis affecting the skin and mucous membranes. It has several morphological variants. Zosteriform LP is a rare form of linear LP. Linear LP accounts for less than 1% of cases and presents in a unilateral, linear distribution.<sup>1</sup> The zosteriform type of linear LP is an uncommon variant with dermatomal or zonal distribution. Zosteriform LP is distinguished clinically from linear LP in that it forms a broader band that corresponds to specific dermatomes.<sup>1</sup> Zosteriform LP may arise de novo in previously normal skin, at sites of trauma (koebnerization), or as Wolf's isotopic response at the site of healed herpes zoster.<sup>2</sup> We report a rare case of a 52-year-old female diagnosed with zosteriform LP arising de novo in previously normal, non-traumatized skin with no history of herpes zoster.*

## Case Description

A 52-year-old woman presented with a six-week history of a pruritic eruption on her right leg. The patient had a previous history of discoid lupus erythematosus (DLE) on the scalp, which was previously treated with hydroxychloroquine 200 mg twice daily and clobetasol ointment. The patient at the time of presentation had no DLE lesions. There was no significant family or drug history. The patient had no associated comorbidities. Hepatitis C serology was negative.

On physical examination, numerous purple pruritic papules and plaques with an overlying white scale were present on her right thigh (Figure 1). The lesions were arranged in a linear pattern in the L1, L2, and L3 dermatomal distribution. A linear white patch was seen on her left buccal mucosa (Figure 2), and her genital mucous membranes were normal. There was neither scalp nor nail involvement.

A biopsy was taken from a typical lesion on her right thigh. Histopathology showed wedge-shaped hypergranulosis, acanthosis, saw toothing of the rete ridges, and a lichenoid infiltrate (Figure 3). A diagnosis of zosteriform lichen planus was made based on the clinical and pathological correlation.

The patient was given an intramuscular injection of triamcinolone acetonide 40 mg and started on tacrolimus ointment 0.1%, which was applied to the cutaneous and oral lesions twice daily.

## Discussion

Lichen planus is an idiopathic inflammatory disease affecting the skin and mucous membranes. Middle-aged individuals are most commonly affected, with the average age of onset being 50 years. The occurrence is evenly distributed worldwide with no racial predilection. Females are affected more often than males. The mucous membranes are affected in 65% of cases. In cases of oral LP, the buccal mucosa exhibits a lacy white reticulation, known as Wickham's striae, and may develop into squamous cell carcinoma in 0.2% of cases per year.<sup>3</sup> The nails may be affected along

with the scalp, causing a scarring alopecia.

Clinically, zosteriform LP is characterized by flat-topped, pruritic, violaceous papules or plaques. The eruption has a predilection for the flexure surfaces of the extremities and is usually symmetric in nature. It may occur in association with certain medications and diseases such as hepatitis C, but the etiology is unknown and is thought to arise from a T-cell mediated immune response.

In addition to the classic appearance, there are more than 20 variants that are categorized based on the morphology and configuration of the lesions. There are many atypical presentations of LP that have been described, such as linear, hypertrophic, actinic, and vesicubullous.<sup>4</sup>

Linear LP refers to lichen planus with a unilateral, linear distribution. It can present in a segmental distribution corresponding to a dermatome, referred to as zosteriform lichen planus. Zosteriform LP may arise as Wolf's isotopic response at areas of healed zoster, secondary to koebnerization from trauma, or in very rare cases as a de novo eruption on previously normal skin.

The zosteriform pattern can occur without evidence of herpes zoster, and the incidence is extremely infrequent. The lesions are arranged in a band several centimeters wide and run along the course of a peripheral cutaneous nerve and its branches. It is thought that the cutaneous manifestation of the zosteriform pattern could possibly be triggered by neural factors.<sup>5</sup> It has recently been suggested that the lesions in zosteriform LP actually follow the lines of Blaschko rather than a dermatome.<sup>6</sup> Blaschko's

lines are invisible lines in the skin believed to trace the migration of embryonic cells. Still, some believe that true zosteriform LP only occurs if the lesions develop at sites of healed herpes zoster.<sup>7</sup> In our patient, there was no history of herpes zoster, and multiple dermatomes were involved. Therefore, a more appropriate term may be dermatomal LP rather than zosteriform LP.

The differential diagnosis of zosteriform lichen planus is vast and includes linear psoriasis, lichen striatus, linear epidermal nevus, linear Darier's disease, and inflammatory linear verrucous epidermal nevus, to name a few.<sup>2</sup> LP can be differentiated based on histopathologic examination, which shows a band-like infiltrate of lymphocytes at the dermal-epidermal junction. Other noticeable features include hyperkeratosis, wedge-shaped hypergranulosis, vacuolar degeneration of the basal layer, and acanthosis with saw-toothed rete ridges. Civatte bodies (colloid or cytoid bodies) are present at the junction of the dermis and epidermis and also in the papillary dermis.<sup>8</sup> On direct immunofluorescence (DIF), lichen planus shows shaggy fibrin, cytoid bodies, and deposition of IgM immunoglobulins at the dermoepidermal junction. This can distinguish LP from hypertrophic lupus erythematosus, which would show a continuous granular band of IgG, IgM, IgA, and C3 at the dermoepidermal junction on DIF.<sup>9</sup>

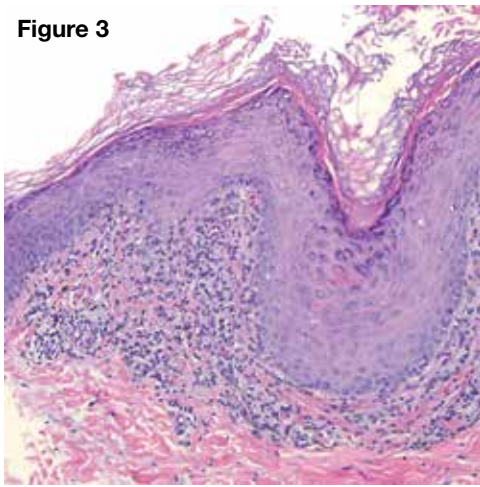


Figure 1



Figure 2

**Figure 3**



novo in normal, non-traumatized skin, as in our patient. Given that our patient had no history of herpes zoster and that multiple dermatomes were involved, a more appropriate term may be dermatomal LP rather than zosteriform LP.

Our patient is currently doing well on a treatment regimen of tacrolimus ointment 0.1% applied to her cutaneous and oral lesions. Per the current recommendations, we are closely following the oral lesion for any malignant transformation.

### **Treatment and Prognosis**

There is no definitive cure for LP, and the disease is self-limiting. There are various treatment modalities available to alleviate the associated pruritus and induce remission. Treatments may consist of topical and systemic therapies. Oral antihistamines such as diphenhydramine and hydroxyzine may be used to relieve pruritus. Topical antipruritic agents such as menthol, camphor, pramoxine or doxepin may also be used. First-line therapy for cutaneous lesions includes high-potency topical steroids applied twice daily. Systemic corticosteroids can be used as a second-line treatment or in those with more extensive disease. Alternative treatments include immunosuppressive agents such as cyclosporine, methotrexate, azathioprine, dapsone, retinoids and topical tacrolimus. Narrow-band ultraviolet-B phototherapy and psoralen plus ultraviolet A (PUVA) can also be used as adjuvant therapy.<sup>10</sup> Newer LP treatment modalities such as enoxaparin sodium and oral metronidazole have also been used with success.

In cases of oral LP, corticosteroids are the mainstay of therapy, based on their ability to reduce cell-mediated immune response. They can be given topically, intralesionally or systemically. Systemic and topical steroids, when given together, are extremely effective. A steroid mouth rinse used twice daily can be used to treat generalized oral lesions. Topical ointments can be used to treat localized oral lesions and are applied two to four times daily after meals. *Candida albicans* superinfection can occur with immunosuppressive therapy and should be managed with topical antimycotics.<sup>11</sup>

Lichen planus often resolves over an average period of 18 months, but approximately 20% of patients will have a second occurrence.<sup>12</sup> In a subset of patients, the disease may persist for many years. Oral LP is more therapy-resistant, and close follow-up is advised given the increased risk of developing squamous cell carcinoma.

### **Conclusion**

Zosteriform LP is a rare form of linear LP. It may arise secondary to trauma as Koebner phenomenon, at sites of healed herpes zoster as Wolf's isotopic response, or very rarely de

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