

Introduction

Ulcerative lichen planus (LP) is a variant of lichen planus characterized by chronic, painful erosions, bullae, and ulcerations most commonly located on the mucous membranes. Ulcerative lesions also occur on the palmoplantar surfaces, particularly on the soles. This variant is more common in female patients despite the fact that palmoplantar LP is more common in males. Ulcerative LP clinically manifests as intense pain, typically described by patients as a “burning pain”. These lesions may result in therapy-resistant ulcers and lead to chronic atrophic scarring or the development of squamous cell carcinoma within these lesions.

Histologically, the ulcerative type resembles the other forms of LP with degeneration of the basal layer of the epidermis and a lymphocytic infiltrate obscuring the dermoepidermal junction, as well as sparse scattered plasma cells. Other important histologic features include hyperkeratosis without parakeratosis and focal increases in the granular cell layer.

Syphilis is a chronic, multistage disease caused by *Treponema pallidum* that is acquired via sexual contact with active primary or secondary lesions. Syphilis can present in one of four varying stages. Syphilis is often referred to as “the great mimicker” because lesion characteristics vary to a considerable degree. The second stage of syphilis typically presents as a disseminated disease characterized by constitutional symptoms, a rash, and condyloma lata.

The rash of secondary syphilis consists of papules or patches covering the trunk and extremities, as well as the palms and soles. Condyloma lata refers to the broad, white, wart-like lesions that accompany the rash and typically occur on mucosal membranes. We report a unique case of a 24-year old African American male who presented with shallow ulcerations, which clinically and histologically appeared to share features with the lesions seen in secondary syphilis.

Case Report

A 24-year old African American male presented with a 6-month history of intensely pruritic and painful ulcerations of his fingers, fingernail beds, palms, glans penis, and lower legs. The patient has a past medical history significant for Hodgkin’s lymphoma which was treated two years earlier. The patient has not had recurrence of the lymphoma since the time of treatment. The patient was not currently on any medication regimen.

On physical examination, the patient was afebrile and had no evidence of any systemic symptoms upon review of systems. Skin examination revealed well-demarcated, 1-2 cm round to oval, shallow ulcerations of the bilateral palms, nailbeds, lower legs, and glans penis (Figures 1-

3). In addition, the right heel exhibited a larger, 5 by 3 cm deep, weeping ulceration (not shown).

Based on the patient’s clinical presentation, the differential diagnosis included syphilis, primary HIV infection, ulcerative lichen planus, lichenoid drug eruption, pityriasis rosea, viral exanthem, infectious process, paraneoplastic pemphigus, plasma cell dyscrasia, recurrent Hodgkin’s lymphoma, and ulcerative herpes simplex virus (HSV). Secondary syphilis was the main concern at initial presentation

due to the anatomical distribution of the lesions.

Several involved areas were sampled using an 8-mm punch biopsy and sent for histopathologic examination. Laboratory studies were done to further evaluate the lesions. These studies included Darkfield microscopy, Warthin-Starry staining, RPR, FTA-ABS, HIV testing, a Tzanck prep, PPD, wound culture for bacteria, HSV, fungus, and mycobacteria, testing for autoimmune diseases and recurrence of Hodgkin's lymphoma. Initial management for this patient included empirical treatment with 2 courses of acyclovir for potential HSV and 3 intramuscular doses of penicillin for potential syphilis.

Histopathologic examination revealed hyperkeratosis, focal hypergranulosis, irregular acanthosis, and a "saw-toothing" pattern of epidermis with vacuolar degeneration. There is a band-like dense lymphoplasmacytic inflammatory cell infiltrate in the dermis abating the dermoepidermal junction and extending to the deep dermis in a perivascular and periadnexal distribution (Figure 4, 5, 6). Darkfield microscopy, Warthin-Starry staining, RPR, and FTA-ABS were negative - thus ruling out syphilis. Additional studies included HIV testing, a Tzanck prep, PPD, and wound culture for bacteria, HSV, fungus, and mycobacteria were all negative. The ANA titer was 1:40, making the possibility of autoimmune disease unlikely. The patient's wife was also tested for syphilis, which resulted in a negative RPR and FTA-ABS. Immunohistochemical staining for light chain restriction and immunofixation electrophoresis studies ruled out a plasma cell dyscrasia. Bone marrow aspiration and lymph node biopsies did not reveal recurrent Hodgkin's disease. Indirect immunofluorescence excluded the diagnosis of paraneoplastic pemphigus.

A diagnosis of ulcerative LP made based on histopathological finds and exclusionary data. The patient returned for a follow-up visit two months after initial management was started and no clinical improvement was noted. The patient was then started on an oral course of prednisone for presumed ulcerative LP. The patient responded with greater than 50% healing. Discussion

This case is particularly difficult in that the lesions follow an unusual clinical pattern. The cutaneous manifestations of ulcerative LP are clinically similar to those that present in patients with secondary syphilis. Lesions located on the palms and soles are typically suggestive of secondary syphilis. The lesions of syphilis can present in multiple forms including macular, small papular, follicular, lichenoid, vesicular, pseudovesicular, or psoriasiform. Thus, it is imperative to rule out syphilis in patients that present with such lesions to prevent the

development of complications that could result from untreated syphilis. While lesions in the anogenital regions are common in syphilis, ulcerative LP uncommonly presents in this anatomic location. Thus, a thorough diagnostic workup must be done in order for proper diagnosis and sequential treatment be given. Close follow-up is required in patients with ulcerative LP due to the chronicity of this type of lesion and the potential subsequent development of squamous cell carcinoma arising within the lesions.¹

Histologically, the dermis of ulcerative lichen planus is typically characterized by a lymphocytic infiltrate with no or sparse numbers of plasma cells². Other cutaneous lesions including those of syphilis, plasmocytomas, necrobiosis lipoidica, leishmaniasis, and deep fungal infections share these features.

There have been few case reports of patients with ulcerative

lichen planus which showed a lichenoid infiltrate composed mainly of plasma cells. One report describes a patient with a toenail lesion that clinically and histologically represents LP, but also shows a band-like plasma cell infiltrate². The few cases that have been reported thus far have been in women ages 56-86 and a 78 year old male, all with associated nail involvement.²⁻⁶

Ulcerative LP has been reported in patients with immune dysfunction and several underlying malignancies including: small-cleaved cell lymphoma, Non-Hodgkin's lymphoma, Castleman's tumor (Giant lymph node hyperplasia), metastatic adenocarcinoma, and malignant fibrous histiocytoma⁶. Thus, ulcerative LP may represent an autoimmune reaction based on cross reactivity between tumor and cutaneous antigens. Underlying immune dysfunction may be responsible in part for these events and this unusual lichenoid infiltrate. One proposed mechanism involves cytotoxic T lymphocytes, presumably sensitized to tumor antigens, attacking cross-reacting antigenic structures in skin and mucosa. Although our patient had no evidence of recurrent Hodgkin's disease, tumor associated lichen planus has been reported to continue after complete remission of the underlying neoplasia. The etiology and pathogenesis of ulcerative LP has been a focus of continued research in the dermatological community. Conclusion

Ulcerative LP can present unconventionally. The clinical and histopathological features may vary from the standard presentation. Thus, this case illustrates an important concept that there should be a high index of suspicion for ulcerative lichen planus when evaluating ulcerative lesions of the palmoplantar and anogenital areas.

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