

## BRIEF ARTICLES

## Lues maligna in an immunocompetent male

Carly Dunn, BA<sup>1</sup>; Danielle S. Applebaum, MD<sup>2</sup>

<sup>1</sup>School of Medicine, Baylor College of Medicine, Houston, Texas

<sup>2</sup>Baylor College of Medicine, Department of Dermatology, Houston, Texas

## ABSTRACT

Lues maligna (malignant syphilis) is an aggressive, rapidly developing, rare variant of secondary syphilis and is characterized by flu-like prodromal symptoms followed by an eruption of irregularly scattered erythematous papules and pustules that quickly progress into well-defined necrotic ulcers. Atypical presentations of syphilis, such as lues maligna, occur more commonly in HIV-positive and immunocompromised individuals. We present a rarer case of an immunocompetent patient with lues maligna. Syphilis mimics many other conditions and it is easily treatable once diagnosed. Therefore, dermatologists should think beyond pityriasis rosea-like rashes and consider syphilis when patients present with lesions that resemble pityriasis lichenoides et varioliformis acuta (PLEVA), drug eruption, disseminated herpes or zoster infection, cutaneous T-cell lymphoma, and ulcerating vasculitis.

## INTRODUCTION

Syphilis, a sexually transmitted disease from the spirochete *Treponema pallidum*, is often referred to as the “the great imitator” because of its plethora of clinical presentations.<sup>1,2</sup> Lues maligna (malignant syphilis) is a rare, aggressive variant of secondary syphilis predominately found in patients with immunodeficiency; however, it has also been described in patients with chronic alcohol or drug abuse, tuberculosis, or diabetes mellitus.<sup>1</sup> We present a case of an immunocompetent patient diagnosed with lues maligna.

## CASE REPORT

A 29-year-old immunocompetent African American male presented with diffuse, painful, boils for two weeks. The rash

originated as a single bullous lesion and became generalized despite empiric treatment with cephalexin for ten days. He had no past medical history.

The patient was afebrile, and physical examination revealed widespread hyperpigmented papules, nodules, and ulcerated papulonodules with overlying crusts on his torso, buttocks, and extremities as shown in Figure 1. Biopsy of a lesion on his back revealed an inflammatory crust, necrosis in the epidermis and mid-dermis, and a wedge-shaped mononuclear infiltrate containing eosinophils in the deep dermis. The biopsy was initially read as consistent with PLEVA. Prior to initiation of methotrexate for PLEVA, laboratory examination revealed a positive RPR. HIV testing was negative. The patient was diagnosed with lues maligna given his

March 2019 Volume 3 Issue 2

**Figure 1:** Multiple ulcerated and necrotic papulonodules with a secondary hemorrhagic crust as well as hyperpigmented macules.



clinical, histopathologic, and positive RPR. The patient experienced complete resolution in two to three weeks with enduring post-inflammatory hyperpigmentation after treatment with a single dose of Benzathine penicillin G 2.4 million units intramuscularly.

## DISCUSSION

Syphilis progresses from primary to secondary to tertiary syphilis; however, at each stage the presentation can deviate. Secondary syphilis can be variable, including, but not limited to: diffuse maculopapular rash that involves the palms and soles with lymphadenopathy, highly infectious condyloma lata, syphilitic hepatitis, mild constitutional symptoms, and/or bone pain<sup>1</sup>. Lues maligna (malignant syphilis) is an aggressive, rapidly developing, rare variant of secondary syphilis and is characterized by flu-like prodromal symptoms followed by an eruption of irregularly scattered erythematous papules and pustules that quickly progresses into well-defined necrotic ulcers. The lesions most commonly present with a pityriasis rosea-like eruption, are polymorphic, often involve the scalp and face, and can become covered with a dark,

rupioid crust.<sup>2</sup>

Atypical presentations of syphilis, such as lues maligna, commonly occur in HIV-positive and immunocompromised individuals.<sup>2</sup> A retrospective, multicenter study of 11,368 patients with HIV revealed that 1.3% of them had concurrent syphilis and of those 7.3% had lues maligna.<sup>2</sup> Another retrospective study by Romero-Jimenez found that approximately 80% of patients with lues maligna had a concurrent AIDS-defining illness with CD4 counts less than 200 cells/mL<sup>3</sup>. However, it is thought that a functional, rather than quantitative deficit is responsive for its aggressive presentation, as many cases of lues maligna are seen in individuals with CD4 counts greater than 200 cells/mL.<sup>1</sup>

Characteristic histologic findings are similar to other presentations of secondary syphilis and include epidermal necrosis, dense perivascular and interstitial inflammation, lymphocyte and plasma cell invasion, and vascular changes.<sup>1</sup> The vessel involvement spans from endothelial swelling and/or proliferation, hyaline thrombi, and fibrinoid deposition.<sup>1</sup> Along with clinical and histologic findings, immunohistochemical staining can identify spirochetes thus aiding in diagnosis.<sup>1</sup>

The differential diagnosis for malignant syphilis includes disseminated herpes simplex or zoster, pityriasis lichenoides et varioliformis acuta, drug eruption, cutaneous T-cell lymphoma, and ulcerating vasculitis.<sup>6</sup> Lues maligna can be differentiated using the clinical presentation, serological syphilis testing, histologic correlation, Jarisch-

Herxheimer reaction, and a response to treatment.**Error! Bookmark not defined.**

The gold standard treatment for lues maligna is a single intramuscular dose of Benzathine penicillin G 2.4 million units.<sup>7</sup> In cases where the patient is allergic to penicillin, ceftriaxone can be used as an alternative.<sup>7</sup> Ulcers respond more slowly than other stages of lues maligna; however, most patients respond completely to therapy within 2 weeks to 4 months.<sup>i</sup>

**Conflict of Interest Disclosures:** None.

**Funding:** None.

**Corresponding Author:**

Carly Dunn  
Baylor Dermatology Clinic  
Houston, TX 77030  
Email: [carlyd@bcm.edu](mailto:carlyd@bcm.edu)

---

**References:**

1. Watson KMT, White JML, Salisbury JR, Creamer D. Lues maligna. *Clin Exp Dermatol.* 2004;29(6):625-627. doi:10.1111/j.1365-2230.2004.01630.x.
2. Chang WT, Hsieh TT, Wu YH. Malignant syphilis in human immunodeficiency virus-infected patients. *Derm Sinica.* 2015;33:21-25.
3. Pföhler C, Koerner R, Von Müller L, Vogt T, Müller CSL. Lues maligna in a patient with unknown HIV infection. *BMJ Case Rep.* 2011;2011:bcr0520114221. doi:10.1136/bcr.05.2011.4221.
4. Stevenson J, Heath M. Syphilis and HIV infection: an update. *Dermatol Clin.* 2006;24:497-507.
5. Schöfer H, Imhof M, Thoma-Greber E, et al. Active syphilis in HIV infection: a multicentre retrospective survey. *Genitourin Med.* 1996;72:176-81.
6. Romero-Jimenez MJ, Suarez Lozano I, Fajardo Pico JM, Baron Franco B. Malignant syphilis in patient with human immunodeficiency virus (HIV): case report and literature review. *An Med Interna.* 2003;20(7):373-6 [in Spanish].
7. Maldonado P, Sendagorta Cudos E, Zamora Vargas FX, Merino MJ, Pinto PH. Pathologically confirmed malignant syphilis using immunohistochemical staining: report of 3 cases and review of literature. *Sex Transm Dis.* 2014;41:94-97.
8. Witkowski JA, Parish LC. The great imitator: malignant syphilis with hepatitis. *Clin Dermatol.* 2002;20(2):156-163. doi:10.1016/S0738-081X(01)00249-8.
9. Yamashita M, Fujii Y, Ozaki K, et al. Human immunodeficiency virus-positive secondary syphilis mimicking cutaneous T-cell lymphoma. *Diagn Pathol.* 2015;10(1). doi:10.1186/s13000-015-0419-5.
10. Sary, A, Sary, G. Sexually Transmitted Infections. *Dermatology*, by Jean L. Bologna et al., Elsevier, 2018, pp. 1447-1469.
11. Sands M, Markus A. Lues maligna, or ulceronodular syphilis, in a man infected with human immunodeficiency virus: case report and review. *Clin Infect Dis.* 1995;20:387-390.