

The great impostor: Lues maligna in an HIV-infected male

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Abstract

Lues maligna is a rare severe cutaneous manifestation of secondary syphilis. It is also known as malignant syphilis and ulceronodular syphilis. We report a case of a 58-year-old HIV-infected male who presented with diffuse, pruritic, non-tender, maculo-papular skin lesions, ulcerated nodules and plaques surrounded by an erythematous base. The disseminated skin lesions were at various stages and were located on his back, chest, arms and testicles. Patient had been receiving antiretroviral therapy. Laboratory studies had demonstrated CD4 lymphocyte count of 463 cells/mm³ and an undetectable HIV viral load. Workup revealed a rapid plasma reagin of 1:256 dilutions and the skin biopsy findings were compatible with syphilis. The skin lesions resolved with intramuscular penicillin. We herein describe a rare case of lues maligna in an HIV-infected patient with a preserved immune function and viral suppression. Such skin lesions can mimic fungal or mycobacterial infections and can pose a diagnostic challenge. Even in the modern era, syphilis remains the great impostor. Clinicians must be able to recognize this condition based on clinical characteristics and risk factors to diagnose and treat this condition promptly.

Keywords

Infectious diseases, dermatology

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Introduction

Lues maligna (LM) is a rare dermatologic manifestation of *Treponema pallidum*. We describe a male infected with human immunodeficiency virus (HIV) who presented with disseminated papulopustules and ulcerated nodules at various stages. The clinical presentation of this case can be a diagnostic challenge and can mimic other diseases such as fungal, mycobacterial and herpetic infections, as well as, pityriasis lichenoides et varioliformis acuta.¹ Clinicians must be aware of this rare cutaneous manifestation of a common sexually transmitted illness (STI) in the differential diagnosis, especially in those populations who are at risk.

Case report

A 58-year-old HIV-infected man who has sex with men (MSM) presented with a 2-week history of diffuse, non-tender, pruritic skin lesions at different stages. The lesions initially appeared as red macules on his chest and arms, and evolved into raised papular, darkened violaceous lesions surrounded by an

erythematous base. More lesions later developed on his back, legs, feet and testicles (see Figure 1). The patient had a medical history of HIV and prior STIs. He was diagnosed in 1987 and has been compliant with his antiretroviral therapy (ART) for the past 15 years. His ART medications included emtricitabine, rilpivirine and tenofovir in a once-daily combination pill. Blood tests revealed a CD4 count at 463 cells/mm³ and the HIV viral load was undetectable.

Physical examination revealed a large 4 cm crusted plaque on his right abdomen, plus scattered annular dime-sized crusted papules on chest, back, arms, legs, feet and testicles. Laboratory testing reported normal complete blood counts,

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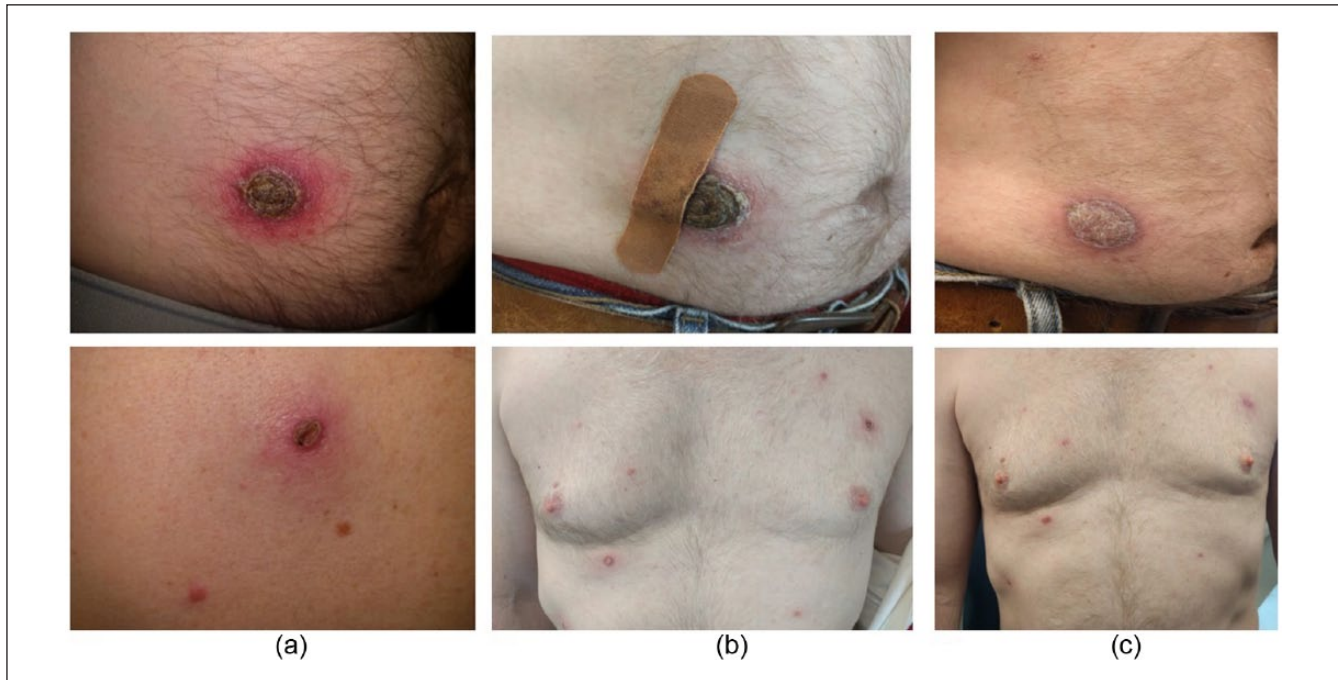


Figure 1. (a) Skin manifestations of lues maligna on presentation and response to treatment at (b) 1 week and (c) 2 weeks.

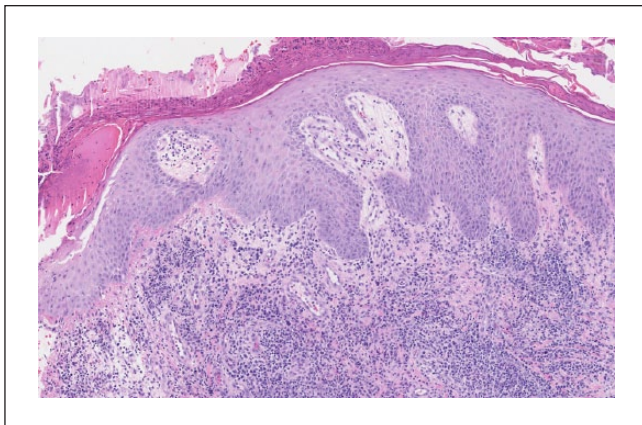


Figure 2. Dense dermal plasma cell infiltrates with an overlying purulent serum crust and reactive hyperplastic epidermal changes.

liver function tests and chemistry panel. Chest radiography was unremarkable. Urine *Blastomyces* Ag was negative. The quantitative rapid plasma reagin (RPR) was reported positive with a 1:256 dilution. The patient received one dose of intramuscular penicillin. A skin biopsy was performed 5 days after treatment. Microscopic examination showed dermal lymphoplasmacytic infiltrates with overlying cutaneous ulceration suggestive of syphilis (see Figure 2). Special stains including periodic acid–Schiff (PAS), acid-fast bacilli (AFB) and Steiner stains did not reveal fungi, mycobacteria or spirochetes, respectively. The patient was asymptomatic 3 weeks after treatment.

Discussion

LM was first described in 1859 by Bazin.² Huslund and Neisser independently classified LM as a rare and severe variant of secondary syphilis in 1896.^{2–4} Prior to the advent of HIV, LM had an estimated incidence of 0.12%–0.36% and was mostly described in patients with severe malnourishment or chronic alcoholism.^{1,5,6} Only 14 cases were reported in the literature prior to 1988.⁷ The HIV epidemic coincided with a rise in reported syphilis cases in the United States affecting the same at-risk populations.^{8,9} HIV-positive patients are more than 60 times more likely to develop LM than HIV-negative patients.¹⁰ Despite this rise, LM remains rare. To our knowledge, less than 40 cases of HIV-related LM have been reported in the literature.^{7,11} Rare cases in immunocompetent or HIV-positive individuals with adequate T cells have been described, including patients with poorly controlled diabetes.^{1,7,12,13} Most HIV-related cases occur in poorly controlled patients with CD4 counts between 100 and 350.^{7,11} Immune reconstitution inflammatory syndrome (IRIS) seems to increase the risk of LM developing in HIV-infected patients.^{6,14}

Various terms have been used to describe LM, including malignant syphilis, syphilis maligna praecox or ulcero-nodular syphilis. The hallmark of LM is the dermatological finding of multiple pleomorphic round-to-oval papules, papulopustules or nodules with ulceration and brown lamellar crusted lesions disseminated over the trunk and extremities.^{5,7} These skin lesions may be preceded by fever, malaise, myalgia, headache and weight changes over 4 weeks prior to the appearance of skin lesions.⁷ A biopsy is recommended

and is useful in excluding other causes such as bacterial, fungal and mycobacterial infections. Microscopic evaluation may reveal nonspecific findings such as perivascular congestion along with dermal infiltrate composed of plasma cells. Most reports show limited or no spirochetes.^{6,15,16} It is acknowledged that immunohistochemistry is superior in detecting spirochetes in comparison with the silver stains.^{16,17} However, this immunohistochemistry may not always be available on a routine basis at many laboratories. Thus, clinical pathologic correlation with serologic testing is key to prompt diagnosis and treatment of secondary syphilis.

Successful treatment with aqueous crystalline penicillin G has been reported, with regimens varying from single intramuscular injection, three weekly injections and 14 days of parenteral penicillin.^{1,7,10,12} Clinicians must stay vigilant for possible Jarisch–Herxheimer reaction, and HIV-infected patients on ART should continue antiretrovirals.¹⁸ Lesions respond to therapy leaving minimal residual effects.^{10,18}

Conclusion

With its multiple stages and wide clinical presentation, syphilis has been described as the great imposter. LM can be mistaken for a disseminated mycosis or mycobacterial disease. Our patient with well-controlled HIV infection was diagnosed with LM based on clinical suspicion, risk factors and serologic testing. This rare case also highlights that LM can occur in patients with HIV outside the clinical spectrum of IRIS and without profound immunosuppression. Of significance, the United States has seen an increase in reported syphilis cases, with the highest rate occurring among MSM.⁹ The risk behaviors associated with acquiring syphilis also increase the likelihood of acquiring HIV.¹⁹ More than half of newly diagnosed HIV infections in the United States are MSM.⁸ Likewise, more than 50% of MSM with syphilis are also co-infected with HIV.⁹ With these two coexistent epidemics, clinicians must be vigilant of this rare manifestation of a common condition, especially among MSM. Clinical suspicion combined with serologic testing for syphilis is key for early diagnosis and treatment.

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Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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