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Malignant syphilis: ostraceous, ulceronecrotic lesions in a patient with human immunodeficiency virus

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Abstract

We present a 36-year-old HIV-positive man with a six week history of spreading, ulcerative, and necrotic cutaneous lesions. Laboratory and histopathologic examination revealed syphilis. This case of malignant syphilis, also known as lues maligna, is an uncommon variant of this sexually transmitted infection. This case highlights the importance of including malignant syphilis in the differential diagnosis of patients presenting with a disseminated ulcerative and necrotic rash, especially in individuals with HIV.

Keywords: malignant syphilis, syphilis, doxycycline, lues maligna, HIV

Case Synopsis:

History

A 36-year-old man with a known history of human immunodeficiency virus (HIV) diagnosed in 2000, who had known non-compliance with taking highly active anti-retroviral therapy (HAART), presented with a diffuse, non-pruritic erythematous skin eruption. The eruption started on the nares as a "small sore" six weeks prior to presentation and progressively evolved into a more generalized eruption composed of large, scaly, and crusted plaques, some of which were necrotic. A few of the papular lesions were present on the palms and soles, and several lesions were tender. Associated symptoms included fever, chills, night sweats, severe fatigue, and swelling of the hands and feet, all of which began over the preceding several weeks. The patient denied neurologic or psychiatric symptoms. The patient did not report any previous genital chancre.



Figure 1. Multiple ulceronecrotic plaques with ostraceous crusting, diffusely distributed across trunk, and arm. Similar lesions were present on the face, back, and other extremities.

Physical Examination

Integumentary exam revealed multiple erythematous papules, plaques, and nodules of varying sizes with ostraceous crusting, distributed diffusely across the face, trunk, back and extremities. Among these lesions, many had central ulceration with necrosis. There were also multiple small, erythematous papulosquamous lesions scattered across the trunk, extremities, palms, and soles. No lymphadenopathy was noted. The remainder of the physical exam was unremarkable.

Laboratory and Imaging:

Pertinent laboratory studies included a positive RPR (1:64 titer), positive syphilis confirmatory test, negative CSF VDRL, a CD4 count of 57 cells/mm³, HIV viral load of 365,000 copies/mL, and white blood cell count of 3,400/mm³. Roentgenograms of the hands and feet revealed polyostotic cortical irregularities, and a bone scan revealed bilateral extensive



Figure 2. Erythematous, papulosquamous lesions on the soles. Similar lesions were present on the palms.

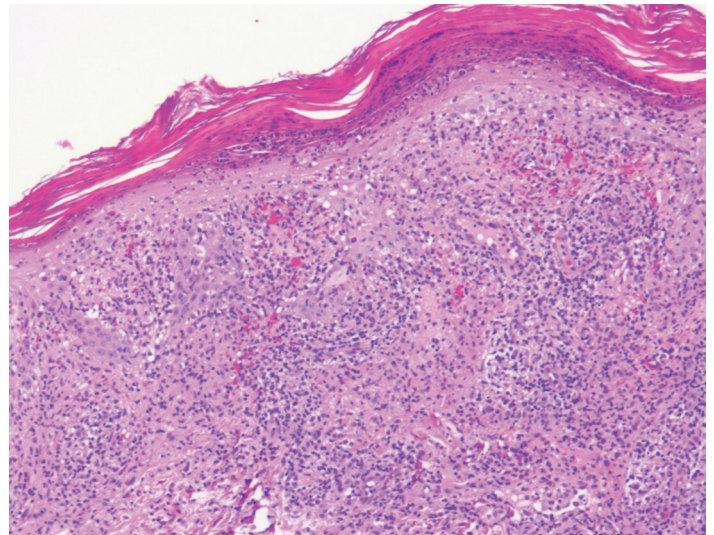


Figure 3. Photomicrograph from left arm lesion revealed a lichenoid, psoriasiform, somewhat granulomatous dermatitis. (hematoxylin and eosin, x100)

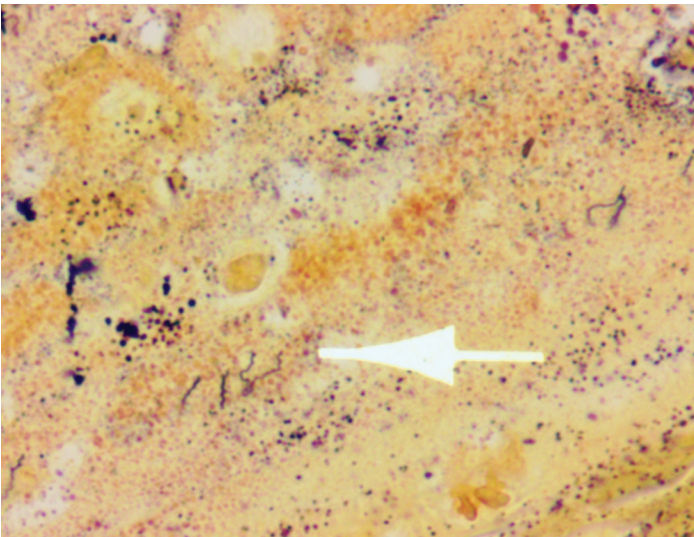


Figure 4. Steiner stain demonstrates several spirochetes (white arrow) within the epidermis. (modified Steiner, x 400)

polyostotic uptake of the forearms, tibias, hands, and feet, all findings consistent with secondary syphilis. Chest roentgenogram was negative. Punch biopsies were performed from skin lesions on the forearm and leg. See Figures 3 and 4 for histopathologic findings.

With the above clinical, laboratory, imaging and histopathologic data taken into consideration, it was determined that secondary syphilis was the most likely diagnosis. The patient was admitted to the hospital for treatment. Doxycycline 100mg twice daily was initiated to treat the secondary syphilis, in addition to treatment for HIV infection. The patient's condition improved significantly within the first week of therapy.

Discussion:

Syphilis, a sexually transmitted disease owing to infection with the spirochete *Treponema pallidum*, has 3 classic stages: primary syphilis, which presents with a chancre at the primary inoculation site (most often the genitalia), secondary syphilis, which is typically characterized by a widespread papulosquamous rash, and tertiary syphilis, which can have many manifestations including neurologic and cardiovascular symptoms [1].

We present an HIV patient with “malignant” syphilis, confirmed by laboratory, imaging, and histopathologic examinations. This uncommon variant of syphilis, also termed lues maligna, is a severe form of secondary syphilis and classically demonstrates skin lesions with more ulcerative and necrotic features than is typically seen with ordinary secondary syphilis. Prominent constitutional symptoms are often present as well, in addition to the requisite laboratory and histopathologic findings [2].

Malignant syphilis was first described before the spread of HIV [3] and can be seen in any immunosuppressed individual, including patients with a history of ethanol abuse [4]. However, malignant syphilis is most likely to be found in HIV patients, as the incidence of this variant of syphilis is reported to be up to 60 times higher in HIV patients

compared to the general population [5]. Additionally, one study found 80% of HIV patients with malignant syphilis possessed a CD4 count greater than 200 cells/mm³ [6]. Our patient's presenting CD4 count was 57 cells/mm³, making this case somewhat unusual.

The ulcerative features of the skin lesions in malignant syphilis are owing to an obliterative endarteritis caused by the infection [7]. The severely destructive nature of these lesions is likely related to the immunocompromised state of the host, but may also be due to a more virulent strain of *Treponema pallidum* [8, 9].

Treatment of choice for syphilis remains benzathine penicillin G, but in patients with penicillin allergy (such as in our case), other antibiotics may be used. Our patient was successfully treated with doxycycline. One other study also reported effective treatment of malignant syphilis with doxycycline [10].

Rates of syphilis have increased in HIV patients in recent years [11]. Up to two-thirds of new syphilis cases can be found in patients also infected with HIV, and the course of syphilis tends to be accelerated in HIV patients [7, 12]. This case of florid, malignant syphilis demonstrates the importance of including this variant of syphilis in the differential diagnoses of patients presenting with a disseminated ulcerative and necrotic rash, especially in individuals infected with HIV, particularly those with CD4 counts below 200. The evaluation of patients with this type of skin eruption and clinical presentation should include a skin biopsy with stains for spirochetes, and laboratory testing for syphilis [13]. Early diagnosis of syphilis is paramount, as prompt treatment can prevent severe, systemic complications of the infection.

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