Malignant syphilis in an immunocompromised female: A case report from Northeast India

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Abstract

Malignant syphilis (lues maligna) is a rare form of secondary syphilis, first described by Bazin in 1859, frequently associated with HIV infection. The resurgence of syphilis in the recent times has been attributed to rise in HIV infection. Malignant syphilis is characterized by the presence of pleomorphic multiple round-to-oval papules, papulopustules, or nodules with ulceration, without central clearing, and occasionally exhibit a lamellate brown to black rupioid crust with prodromal symptoms. We herein report a case of early malignant syphilis in a young immunocompromised patient who was HIV positive and noncompliant to antiretroviral therapy, from Northeast India. The HIV/AIDS epidemic is rising at an alarming rate in this part of the country which has thus led to an increase in the number of other sexually transmitted infections.

Key words: HIV, malignant syphilis, Northeast India, rupioid crusts

Introduction

Malignant syphilis (MS) (lues maligna) is a rare form of secondary syphilis, first described by Bazin in 1859, frequently associated with HIV infection. However, rarely, it can occur in immunocompetent individuals as well as those with chronic alcoholism, malnutrition, prolonged corticosteroid therapy, and coexistent debilitating diseases.[1]

Since the beginning of the HIV epidemic, the incidence of MS has been steadily rising, making it a disease of vital recognition for any patient with suspicious cutaneous lesions.[2] The resurgence of syphilis in the recent times has been attributed to rise in HIV infection. A study from Germany revealed that 7.3% of HIV-positive patients had MS.[1]

MS, or syphilis maligna praecox, lues maligna, or rupioid syphilis, is defined as the presence of pleomorphic multiple round-to-oval papules, papulopustules, or nodules with ulceration, without central clearing, and occasionally exhibit a lamellate brown to black rupioid crust. It is characterized by marked prodromal constitutional symptoms, such as fever, malaise, myalgia, headache, and weight changes over the span of 4 weeks before the appearance of skin lesions.[2]

We herein report a case of early MS in a young immunocompromised patient who was HIV-positive and noncompliant with antiretroviral therapy, from Northeast India. The HIV/AIDS epidemic is rising at an alarming rate in this part of the country which has thus led to an increase in the number of other sexually transmitted infections.

Case Report

A 28-year-old female patient presented to the emergency department of our hospital with a 4-week history of fever, loss of appetite, and myalgia, followed by the onset of skin lesions.

The lesions first began on the trunk and gradually involved the face and the extremities. She also gave a history of weight loss over the past 6 months. She is married with two children, but the husband has been estranged for the past 1 year.

The patient did not give a history of any genital lesion in the past.

The physical examination revealed that the patient was afebrile, conscious, and well-oriented to time, place, and person. She was severely underweight with a BMI of 16.33 (weight –36 kg and height –1.49 m).

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There was no pallor/icterus/cyanosis; a few enlarged lymph nodes were seen in the bilateral inguinal region; peripheral edema was present over the bilateral lower limbs extending up to the knee.

The cutaneous examination revealed multiple nodules over the face, the trunk, and the upper and lower limbs. The nodules were covered with thick, yellowish, rupioid-like crusts, and some of them were ulcerated. There was mutilation of the nasal tip, and scarring was present wherever the necrotic crusts were present [Figure 1a-c].

On examination of the oral mucosa, thrush was seen on the tongue and over the buccal mucosa. The genital mucosa appeared normal. There were lesions over the eyelids; however, the ophthalmological examination revealed no significant abnormality.

The neurological, gastrointestinal tract, musculoskeletal, and cardiovascular examinations were all within the normal limits.

From the cutaneous presentation, a differential diagnosis of pemphigus vegetans, pyoderma gangrenosum, lupus vulgaris, deep fungal infection, rupioid psoriasis, and secondary syphilis was thought of.

The patient was admitted and investigated further.

The serum investigations revealed a positive Venereal Disease Research Laboratory (VDRL) Rapid Plasma Reagin (RPR) (TITER-1:4) and a positive Treponema pallidum hemagglutination assay status. After further enquiry, the patient revealed that she had been diagnosed as HIV positive in the past 6 months, had been started on antiretroviral treatment, but had not continued taking the treatment.

A CD4 cell count was 90 cells/mm^3. The serology for hepatitis B and C was negative. A complete blood count revealed pancytopenia with a total white blood cell count of 21,000 and hemoglobin –7.7 gm%, and the liver function test revealed a serum albumin of 2.2 gm% with a total protein level of 6.6. The Mantoux test was negative.

A histopathological examination of the skin revealed pustule formation, i.e. neutrophil collection in the stratum corneum, ulceration, pseudoepitheliomatous hyperplasia, inflammatory cells consisting of plasma cells, and histiocytes was seen in the dermis [Figure 2a-c].

Stains for fungal elements were negative. The Ziehl–Neelsen stain was negative. No granulomas or clefting were seen.

Considering the VDRL positivity, HIV positivity, and histopathology revealing plasma cells, a diagnosis of MS was made.

The patient was treated with 2.4 million units of benzathine penicillin in a total of three series with an interval of 1 week between the series. The patient showed a significant improvement after each dose [Figures 3-5]. Owing to the low CD4 counts – cotrimoxazole (800/160) and fluconazole (100 mg) prophylaxis was also started. Antiretroviral therapy was started after the completion of the course of benzathine penicillin in view of preventing immune reconstitution inflammatory syndrome). The lesions healed completely with scarring.

Discussion

After the first description in 1859, for many decades, doubts remained if MS was a part of the secondary

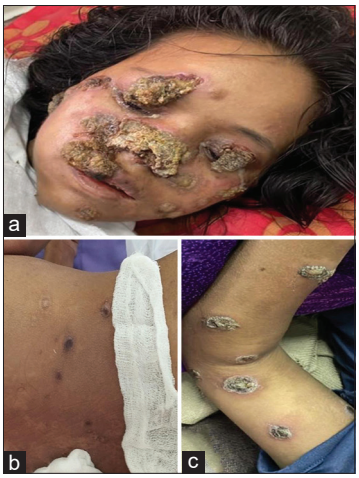

Figure 1: (a-c) Nodules covered with thick, yellowish, rupioid crusts, ulceration, scarring, and mutilation

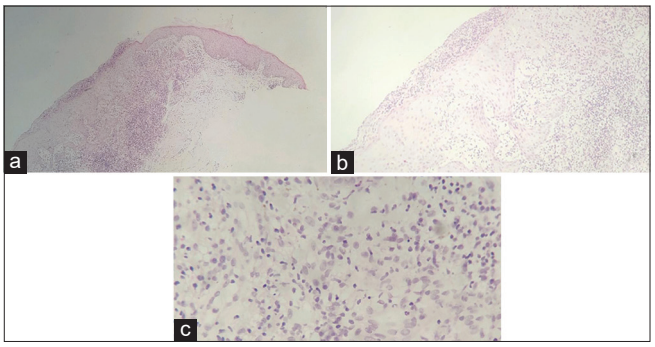

Figure 2: (a-c) Neutrophil collection in the stratum corneum, ulceration, pseudoepitheliomatous hyperplasia, plasma cells, and histiocytes in the dermis

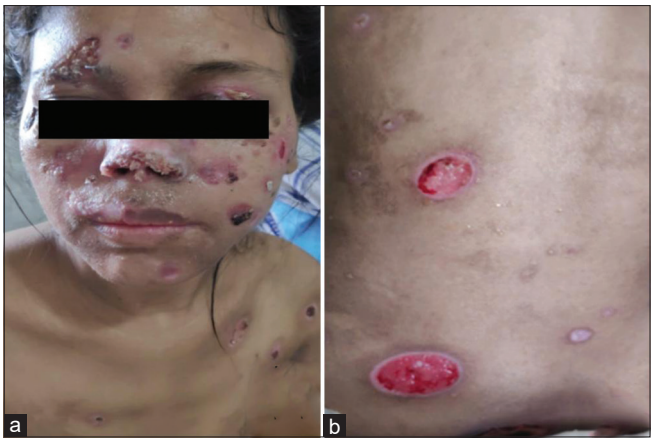

Figure 3: Lesions improved after 1st dose of Benzathine Penicillin

syphilis spectrum or a manifestation of tertiary syphilis. This issue was clarified by studies conducted by Haslund and Neisser, published in 1897. MS is distinguished from the classical secondary syphilis by the general clinical picture, more exuberant and severe, pleomorphic lesions and fundamentally by the presence of ulceronecrotic lesions. It is distinguished from the tertiary syphilis by the larger number of lesions, repercussion in the general health condition, and for being morphologically distinct from chronic gummas, both clinically and histologically.[3]

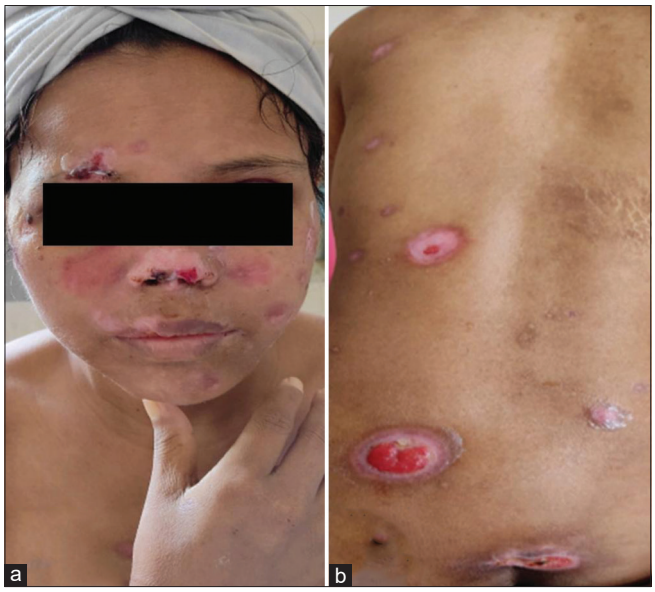

Figure 4: Lesions improved significantly after 2nd dose of Benzathine Penicillin

MS is most likely to be found in HIV patients as the incidence is 60% higher in those with HIV infection.[4]

HIV infection may make clinical manifestations of syphilis more severe and/or atypical. Humoral and cellular immunity against Treponema pallidum depends on the stage of HIV infection and host defense; impairment in immunity may lead to changes in the clinical presentations and natural course of syphilis, increased number of syphilis lesion and degree of infectiousness, and shortened incubation time. A treponemal infection may also act as a facilitator or aggravator for HIV transmission from coinfected patients. Coinfection of syphilis and HIV alters the course of both diseases. The clinical impact of HIV and syphilis coinfection is bidirectional; HIV alters the course of syphilis, and syphilis also appears to adversely impact the HIV disease progression and transmissibility. It is also associated with decreased CD4+ T-cell counts and increased HIV viral loads. Immunologic events that facilitate the development of MS are unknown, but it is reasonable to postulate that the loss of helper T cells is responsible.[2]

One study found that 80% of HIV patients with MS had a CD4 count >200 cells/mm.[5]

Our patient's presenting CD4 count was 90 cells/mm^3, which makes our case somewhat unusual.

Fisher *et al*. defined the classical diagnostic criteria for MS as compatible macroscopic and microscopic skin lesions, a high serology titer, JHR on starting antibiotic treatment, and a rapid clinical resolution with treatment.[6]

Even though our patient did not develop a Jarisch–Herxheimer reaction after starting treatment, the classical presentation of lesions, skin pathology findings, a high VDRL titer, and a rapid resolution of lesions after treatment confirm the diagnosis of MS.

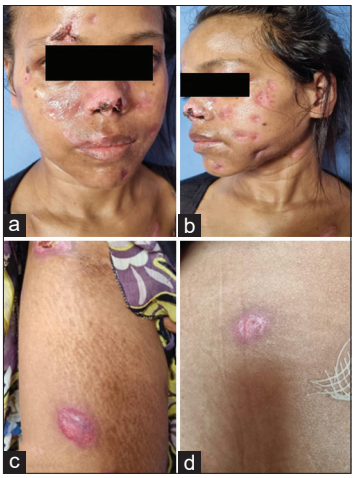

Figure 5: Lesions heal with scarring after 3rd dose of Benzathine Penicillin

Although a few cases have been reported from other parts of India, they have all been reported in immunocompetent individuals. This case is being reported considering the very high prevalence of HIV/AIDS in Northeast India, and no such cases have been reported from this part of the country.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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