Malignant Syphilis in an Immunocompetent Adult Male
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Abstract
The occurrence of malignant syphilis in an immunocompetent individual is rare. We present malignant syphilis in a 35-year-old immunocompetent male who presented with a 1-month history of noduloulcerative lesions on the torso. Examination revealed multiple pustules, nodules, and deep-seated ulcers distributed on the trunk, face, and upper and lower limbs. Characteristic morphology of lesions, positive serological tests for syphilis, characteristic histopathology, and resolution of lesions following institution of penicillin therapy confirmed the clinical diagnosis of malignant syphilis.

Key Words: Endarteritis obliterans, malignant syphilis, nodules, treponemal hemagglutination, ulcers

Malignant syphilis (lues maligna), is a rare form of secondary syphilis, was first described by Bazin in 1859. It is frequently associated with HIV infection; however, rarely, it can occur in immunocompetent individuals.[1] Chronic alcoholism, malnutrition, prolonged corticosteroid therapy, and coexistent debilitating diseases are other predisposing factors.[2] The clinical manifestations of malignant syphilis are different from classical secondary syphilis as these are characterized by pleomorphic pustules, nodules, and deep ulcers with thick crusts.[3]

Case Report
A 35-year-old farmer presented with multiple ulcers over the trunk and upper limbs of 1-month duration. The episode started with the appearance of few papules and pustules on the trunk and arms which subsequently ulcerated to form large deep-seated ulcers. There was associated fever and loss of weight. He denied extramarital sexual exposure. Generalized lymphadenopathy was noted. Examination revealed multiple ulcers of varying sizes ranging from 1 to 3 cm, circular and oval in shape, distributed on the face, front and back of trunk, arms, forearms, thighs, legs, and genitalia [Figure 1]. The ulcers were well circumscribed punched out, deep-seated covered with thick black and brownish yellow laminated crusts with a zone of erythema around [Figure 2]. The ulcers were found healing with atrophy and hypopigmentation [Figure 3]. There was a single indurated nontender, linear ulcer localized to prepuce [Figure 4]. There were few split papules on the upper lip [Figure 5]. Palms showed hyperkeratotic plaques and hyperpigmented patches [Figure 6]. Based on these features, he was provisionally diagnosed as malignant syphilis. However, erythema necroticans and ecthyma gangrenosum were considered in the differential diagnosis. Routine hematological investigations, chest skiagram, and ultrasonography of abdomen were normal. Serological tests for syphilis were positive (venereal disease research laboratory [VDRL] test 1:32 dilutions and treponemal hemagglutination [TPHA] 1:160 dilutions)). HIV serology was nonreactive. Antinuclear antibody test was negative. Staphylococcus aureus was isolated in culture. Slit skin smear for acid-fast bacilli was negative. Histopathological examination of the biopsy taken from the margin of the representative ulcer showed a dense collection of neutrophils, lymphocytes, and few plasma cells in the dermis [Figure 7]. Blood vessels in the dermis showed swollen endothelial cells occluding lumen with the permeation of polymorphonuclear cells in the walls (endarteritis obliterans) [Figure 8]. Examination of the spouse did not reveal clinical evidence of syphilis. Nonetheless, VDRL test was positive

What was known?
Malignant syphilis is known to occur in immunocompromised individuals.

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in 1:32 dilutions. Both were administered injection benzathine penicillin 2.4 mega units intramuscularly and are under follow-up.

**Discussion**
The term “malignant” is used to describe the clinical features and not related to the pathological entity.
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The exact pathogenesis of malignant syphilis in immunocompetent is not known. However, it was propounded that the infection may be due to the preponderance of virulence of Treponema in the agent-host challenge. Histopathologically, the ulcers were found to be sharply circumscribed anemic infarctions of the dermis and epidermis, due to thrombosis of cutaneous vessels, secondary to obliteration of medium-sized vessels at the border of dermis and subcutaneous tissue.

Malignant syphilis is differentiated from classical secondary syphilis by severe, exuberant pleomorphic lesions, mainly by the presence of ulceronecrotic lesions. The absence of pink, fleshy growths, characteristic of gummatous syphilis, and also the typical round, oval shape of lesions of malignant syphilis helped in differentiating from tertiary syphilis. In pyoderma gangrenosum, the ulcers are usually situated on legs and trunk which are painful and rapidly progressing with raised inflammatory border and boggy necrotic base.

It has been reported that one-third of patients with secondary syphilis have primary chancre at the time of presentation; incidentally, index case also had primary chancre on the prepuce along with nodulo-ulcerative lesions of malignant syphilis. Systemic involvement involving liver and eye has been reported in malignant syphilis. However, there was no evidence of systemic involvement in the index case.

The resurgence of syphilis in recent times has been attributed to rising in HIV infection. A study from Germany revealed that 7.3% of HIV-positive patients had malignant syphilis. Conversely, our patient had no association with HIV infection.

Fisher propounded criteria for the diagnosis of malignant syphilis which include strong serological test, a severe Jarisch–Herxheimer reaction, and a good response to antibiotic therapy. Similarly, strong serological test (positive VDRL test and TPHA test) and characteristic nodulo-ulcerative lesions and response to penicillin therapy confirmed the diagnosis of malignant syphilis in the index case. However, index case did not experience Jarisch–Herxheimer reaction. It was found to be more common among patients with HIV co-infected with syphilis (34.6%) than HIV-uninfected with syphilis (25.2%) who received penicillin treatment.

In conclusion, although malignant syphilis is most commonly associated with HIV infection, it should be strongly suspected in a widespread nodulo-ulcerative presentation in an immunocompetent also. Early diagnosis and treatment will help in alleviating morbidity and thereby arrest the progression of the disease. In addition, every effort should be made to trace and treat the contacts so as to prevent transmission of syphilis in the community.

**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 anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**What is new?**

Malignant syphilis presenting in an immunocompetent individual is rare.
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