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Case Report

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Syphilis Maligna (Lues Maligna): A Case Report

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INTRODUCTION

Syphilis is a sexually transmitted disease (STD) produced by *Treponema pallidum*, an anaerobic filamentous spirochete, which has a tropism for several organs and tissues in the body. A rare form of detructive syphilide, with deeply ulcerative covered with thick crust lesions was described under the name malignant syphilis (lues maligna).[1] Most of the cases, resulting from human immunodeficiency virus (HIV)-induced suppression of cell mediated immunity, have been reported in HIV positive patients[1-20] but some cases occur in individuals with poor health, alcoholic or immunocompetent patients.[21,22]

CASE REPORT

A 49-year-old promiscuous man presented to our department with multiple nodular ulcerative lesions on his arms and legs (Figure 1), and a healing genital ulcer. Laboratory tests including syphilis serology showed a Venereal Disease Research Laboratory (VDRL) titre of 1:32 and a positive treponema pallidum hemagglutinatoin (TPHA). HIV (ELISA method) was positive. His CD^{4+} count was 425/mm³ and his HIV viral load 63,420 copies/ml. Histopathological study of skin lesion revealed an epithelial hyperplasia, and perivascular infiltrate containing plasma cells and endothelial thickening of blood vessels throughout dermis extending into the subcutaneous tissue. Periodic acid-Schiff, Grocott´s and Warthin-Starry stains were all negatives in cerebrospinal fluid (CSF) examination showed 1 cell/mm³; protein 35 mg/dL; glucose 57 mg/dL; VDRL negative. Patient was diagnosed to have malignant syphilis associated with HIV infection and he was given injection benzathine penicillin 2.4 per week for 3 weeks. A Jarisch-Herxheimer reaction occurred in the course of therapy. Lesions healed in a month with hypopigmented macules. Viral load dropped to undetectable levels following antiretroviral treatment.

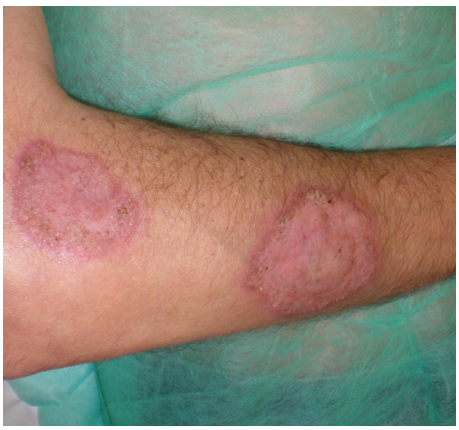

Figure 1: Rupioid plaques on the right arm.

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DISCUSSION

Early syphilis (recently acquired or less than 2 year's duration) is the more contagious stage and includes the primary and secondary forms and the early latent period. Primary syphilis is often asymptomatic and the initial lesion-chancre is extragenital in much cases. Secondary syphilis and latent infection is the most usual forms of presentation in HIV positive patients. Lues maligna is a severe form of secondary syphilis. In these patients syphilis presents an atypical clinical course with severe constitutional symptoms and unusual nodules, necrotic or rupiod skin lesions, organ involvement and a great tendency to develop neurosyphilis and ocular involvement.[20] Serology for syphilis in HIV infection can be falsely negative and biopsy is commonly needed to establish the diagnosis.[6-9]

Histological findings in malignant syphilis are similar to those of secondary syphilis but lymphocytic predominance superficial and deep perivascular infiltrate containing plasma cells, epithelial hyperplasia, perineural plasma cellular infiltrate and thickening of lamina propia blood vessels have been seen in lues maligna in HIV-infected patients. The abundance of plasma cells is a good indicator of malignant syphilis on skin histological analyses, in some cases, the plasma cell count may be very low and cutaneous T-cell lymphoma could be misdiagnosed.[14] No differences in a comparative immunohistologic study were observed between HIV patients and patients who were HIV negative.[12]

Diagnosis of malignant lues should be considered in all HIV-infected individuals who have nodules.[1] CD^{4+} cell count is not the only determinant factor for the occurrence of lues maligna and propably interaction between *T. palidum* and HIV lead to defects of both cell-mediated and humoral immunity.

After the effective control of acquired immune deficiency syndrome (AIDS) in the US and Europe, preventive measures relaxed and in the last years a resurgence of syphilis has been reported in several countries as US, Spain, England, France, Eastern Europe, Russia and China. The main reason for the increase in prevalence is unprotected anogenital and oral sex. Epidemiological changes are related to sexual promiscuity, prostitution, drug abuse, increased travel and migration. New cases occur especially among men who have sex with men (MSM) and are strongly associated with HIV coinfection.[8,9]

A high clinical index of suspicion should be maintained to prevent development of late syphilis or tertiary disease characterized by skin, cardiovascular, neurological, liver, spleen, bones or other organs manifestations. Syphilis is usually diagnosed on the basis of serology test, as detection of treponemes by dark-field microscopy tends to be unreliable. Non-treponemal tests (VDRL, rapid plasma reagin (RPR)) are inexpensive, rapid and commonly used for screening but develop late in primary syphilis. Treponemal test (FTA-ABS y TPHA) are specific antibody test.[2,9] Diagnostic criteria for malignant syphilis include strongly positive serologic test titre, a severe Jarisch-Hersheimer reaction, characteristic microscopic morphology and excellent response to antibiotics therapy.[3,6]

Benazthine penicillin G or aquous procaine penicillin G remains the drug of choice for all forms of syphilis. Oral tetracycline, or doxycycline 100 mgr orally twice, for 14 days are also effective for allergic to penicillin patiens with early syphilis.

HIV patients with primary and secondary syphilis can be treated in the same way as seronegative patients but some authors recommend a more aggressive treatment (as 3 doses of 2.4 million units of benzathine penicillin intramuscularly at weekly intervals) and an accurate follow-up.[9,13]

CONCLUSIONS

Because of increasing syphilis rates the importance of recognizing the early clinical manifestations needs to be re-emphasized.

Diagnosis of syphilis in HIV patients is based in a clinical-pathological correlation together with serological studies. Screening is simple and inexpensive and treatment is highly effective.

CONFLICTS OF INTEREST

The author declares that there are no conflicts of interest.

CONSENT

Author has recieved oral informed consent from the patient whose photograph is involved in this manuscript.

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