

Case Report

*Corresponding author

Rosa Giménez-García, MD

Clinical Assistant

Department of Dermatology

Hospital Universitario Río Hortega;

Associate Professor

Faculty of Medicine

Calle Carabela 115, Boecillo

Valladolid 47151, Spain

E-mail: rosagim@hotmail.com

Volume 1 : Issue 3

Article Ref. #: 1000DRMTOJ1116

Article History

Received: October 18th, 2016

Accepted: November 4th, 2016

Published: November 7th, 2016

Citation

Giménez-García R. Syphilis maligna

(lues maligna): A case report. *Dermatol Open J.* 2016; 1(3): 60-62.

doi: [10.17140/DRMTOJ-1-116](https://doi.org/10.17140/DRMTOJ-1-116)

Syphilis Maligna (Lues Maligna): A Case Report

Rosa Giménez-García, MD^{1,2*}

¹Clinical Assistant, Department of Dermatology, Hospital Universitario Río Hortega, Valladolid, Spain

²Associate Professor, Faculty of Medicine, Valladolid, Spain

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 destructive 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 patients¹⁻²⁰ 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 CD⁴⁺ 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.

Copyright

©2016 Giménez-García R. This is an open access article distributed under the Creative Commons Attribution 4.0 International License (CC BY 4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

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-chancres 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 rupoid 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 propria 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 CD4⁺ cell count is not the only determinant factor for the occurrence of lues maligna and probably interaction between *T. pallidum* 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 aqueous 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 patients 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 received oral informed consent from the patient whose photograph is involved in this manuscript.

REFERENCES

1. dos Santos TR, de Castro IJ, Dahia MM, Creamer D. Malignant syphilis in an AIDS patient. *Infection*. 2015; 43: 231-236. doi: [10.1007/s15010-014-0698-x](https://doi.org/10.1007/s15010-014-0698-x)
2. Watson KMT, White JML, Salisbury JR, Creamer D. Lues Maligna. *Clin Exp Dermatol*. 2004; 29: 625-627. doi: [10.1111/j.1365-2230.2004.01630.x](https://doi.org/10.1111/j.1365-2230.2004.01630.x)
3. Passoni LFC, de Menezes JA, Ribeiro SR, Sampaio EC. Lues maligna in an HIV-infected patient. *Rev Soc Bras Med Trop*. 2005; 38: 181-184. doi: [10.1590/S0037-86822005000200011](https://doi.org/10.1590/S0037-86822005000200011)
4. Pérez-Pérez L, Cabanillas M, Ginarte M, Sanchez-Aguilar D, Toribio J. Malignant syphilis in an HIV-infected patient. *Actas Dermosifil*. 2007; 98: 351-354. doi: [10.1016/S1578-2190\(07\)70460-2](https://doi.org/10.1016/S1578-2190(07)70460-2)
5. Monteagudo-Sánchez B, Rodríguez-González L, León-Muiños E, Ortiz-Rey JA. Sífilis secundaria en paciente infectado

- por VIH [In Spanish]. *Enferm Infecc Microbiol Clin.* 2005; 23: 175-176. doi: [10.1157/13072169](https://doi.org/10.1157/13072169)
6. Tucker JD, Shah S, Jarell AD, Tsai KY, Zembowicz A, Kroshinsky D. Lues maligna in early HIV infection case report and review of the literature. *Sex Transm Dis.* 2009; 36: 512-514. doi: [10.1097/OLQ.0b013e3181a2a946](https://doi.org/10.1097/OLQ.0b013e3181a2a946)
7. Gregory N, Sanchez M, Buchness MR. The spectrum of syphilis in patients with human immunodeficiency virus infection. *J Am Acad Dermatol.* 1990; 22: 1061-1067. doi: [10.1016/0190-9622\(90\)70153-9](https://doi.org/10.1016/0190-9622(90)70153-9)
8. Viñals-Iglesias H, Chimenos-Küstner E. The reappearance of a forgotten disease in the oral cavity: Syphilis. *Med Oral Patol Oral Cir Bucal.* 2009; 14: 416-420.
9. Ficarra G, Carlos R. Syphilis: The renaissance of an old disease with oral implications. *Head and Neck Pathol.* 2009; 3: 195-206. doi: [10.1007/s12105-009-0127-0](https://doi.org/10.1007/s12105-009-0127-0)
10. Modjarrad K, Vermund SH. Effect of treating co-infections on HIV-1 viral load: A systematic review. *Lancet Infect Dis.* 2010; 10(7): 455-463. doi: [10.1016/S1473-3099\(10\)70093-1](https://doi.org/10.1016/S1473-3099(10)70093-1)
11. Prasad PV, Paari T, Chokkalingam K, Vijaybushanam V. Malignant syphilis (Lues maligna) in a HIV infected patient. *Indian J Dermatol Venereol Leprol.* 2001; 67(4): 192-194. Web site. <http://ijdv.com/article.asp?issn=0378-6323;year=2001;volume=67;issue=4;spage=192;epage=194;aulast=Prasad>. Accessed October 17, 2016.
12. McBroom RL, Styles AR, Chiu MJ, Clegg C, Cockerell CJ, Radolf JD. Secondary syphilis in persons infected with and not infected with HIV-1: A comparative immunohistologic study. *Am J Dermatopathol.* 1999; 21(5): 432-441.
13. Blank LJ, Rompalo AM, Erbeding EJ, Zenilman JM, Ghanem KG. Treatment of syphilis in HIV-infected subjects: A systematic review of the literature. *Sex Transm Infect.* 2011; 87: 9-16. doi: [10.1136/sti.2010.043893](https://doi.org/10.1136/sti.2010.043893)
14. Yamashita M, Fujii Y, Ozaki K, et al. Human immunodeficiency virus-positive secondary syphilis mimicking cutaneous T-cell lymphoma. *Diagn Pathol.* 2015; 8: 10: 185. doi: [10.1186/s13000-015-0419-5](https://doi.org/10.1186/s13000-015-0419-5)
15. Hanson C, Fischer R, Fraga G, et al. Lues maligna praecox: An important consideration in HIV-positive patients with ulceronodular skin lesions. *Dermatol Online J.* 2014; 14: 21(3). Web site. <http://escholarship.org/uc/item/5mj272nw>. Accessed October 17, 2016.
16. Wang H, Wang X, Li S. A case of lues maligna in an AIDS patient. *Int J STD AIDS.* 2012; 23(8): 599-600. doi: [10.1258/ijsa.2012.011413](https://doi.org/10.1258/ijsa.2012.011413)
17. Rajan J, Prasad PV, Chockalingam K, Kaviarasan PK. Malignant syphilis with human immunodeficiency virus infection. *Indian Dermatol Online J.* 2011; 2(1): 19-22. doi: [10.4103/2229-5178.79864](https://doi.org/10.4103/2229-5178.79864)
18. Pfoehler C, Koerner R, von Müller L, Vogt T, Müller CS. Lues maligna in a patient with unknown HIV infection. *BMJ Case Rep.* 2011; 27: 2011. doi: [10.1136/bcr.05.2011.4221](https://doi.org/10.1136/bcr.05.2011.4221)
19. Kelly JD, LeLeux TM, Citron DR, Musher DM, Giordano TP. Ulceronodular syphilis (lues maligna praecox) in a person newly diagnosed with HIV infection. *BMJ Case Rep.* 2011; 20: 2011. doi: [10.1136/bcr.12.2010.3670](https://doi.org/10.1136/bcr.12.2010.3670)
20. de Socio GV, Simonetti S, Tomasini C, Ansidei V, Pasticci MB, Baldelli F. Malignant syphilis with ocular involvement in an HIV-infected patient. *Int J STD AIDS.* 2011; 22: 298-300. doi: [10.1258/ijsa.2010.010389](https://doi.org/10.1258/ijsa.2010.010389)
21. Bayramgürler D, Bilen N, Yildiz K, Sikar A, Yavuz M. Lues maligna in a chronic alcoholic patient. *J Dermatol.* 2005; 32(3): 217-219. doi: [10.1111/j.1346-8138.2005.tb00749.x](https://doi.org/10.1111/j.1346-8138.2005.tb00749.x)
22. Alves J, António AM, Matos D, Coelho R, Cachão P. Malignant lues in an immunocompetent patient. *Int J STD AIDS.* 2015; 26: 518-520. doi: [10.1177/0956462414544162](https://doi.org/10.1177/0956462414544162)