

## Case Report

# Rare presentation of the great imitator: a rare case report

Ravindranath Brahmadeo Chavan, Vasudha Abhijit Belgaumkar, Aarti Sudam Salunke,  
Sharada Shivaji Chirame\*

Department of Skin & VD, BJGMC & Sassoon General hospital, Pune, Maharashtra, India

**Received:** 06 July 2017

**Revised:** 08 August 2017

**Accepted:** 10 August 2017

**\*Correspondence:**

Dr. Sharada Shivaji Chirame,

E-mail: [drsharadachirame@gmail.com](mailto:drsharadachirame@gmail.com)

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

### ABSTRACT

Syphilis is often thought to be a disease of the past, largely eradicated in modern health care; however, the rates are still extremely high in certain populations. The diagnosis of syphilis may be overlooked by primary-care clinicians due to the presence of nonspecific signs and symptoms that may be indistinguishable from other diseases. Left undiagnosed and untreated, life-threatening complications, including hepatitis, stroke, and nervous system damage, may occur particularly in immuno-compromised individuals. We present a case of lues maligna an extremely rare presentation of syphilis.

**Keywords:** Lues maligna, Malignant syphilis, Great imitator

### INTRODUCTION

Syphilis is a chronic disease with a waxing and waning course, the manifestations of which have been described for centuries. Malignant syphilis is a severe and rare manifestation of secondary syphilis and in recent times has been reported mostly in patients with HIV infection. Clinically, the condition is characterized by ulcerated skin lesions covered by thick crusts associated with severe constitutional symptoms. The diagnosis is often difficult and is based on clinical, histological and laboratory parameters.<sup>1</sup> We report an HIV-seropositive female presenting with multiple papulosquamous and ulcerative lesions diagnosed as malignant syphilis on serology and histopathology.

### CASE REPORT

A 30 years old married female labourer was brought to out-patient department of Sassoon general hospital, Pune with multiple non-healing ulcers over trunk, extremities (Figure 1 and 2), face since one month (admitted from 10

April 2017 to 1 May 2017). There was history of asymptomatic genital ulcer few months back which healed spontaneously. Patient was separated from her husband and offered history of multiple unprotected penovaginal sexual exposures with a known male. There was no history of similar lesions in partner. She denied history of fever, joint pain, abdominal pain, bowel disturbances, behavioural changes, seizures. There was history of appearance of small raised reddish lesions which developed into ulcers over upper limb, face, trunk, external genitalia and trunk. Patient neglected these lesions which resulted in crusting over each ulcer. Patient was cachexic also having pallor but without lymphadenopathy. On dermatological examination there were multiple round to oval deep ulcers with raised borders over face, extremities and trunk, few of them showing keratotic heaped up brownish crusts with seropurulent discharge. She also had few erythematous annular and gyrate scaly plaques over extremities. Ulcers over eyelids led to destruction and eversion of lower eyelid. Genitals showed single non-tender ulcer with crust over labia minora. Oral cavity showed staining of teeth and

poor hygiene. Her systemic and ocular examination was unremarkable.



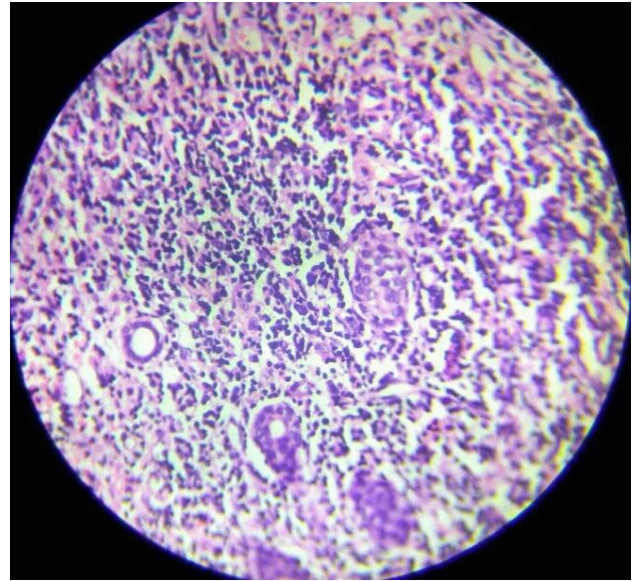
**Figure 1: Multiple ulcers with heaped up crusts.**



**Figure 2: Multiple ulcers with rupioid crusts.**

Patient was investigated considering differential diagnosis of secondary syphilis, Reiter's disease, pyoderma gangrenosum and chronic cutaneous leishmaniasis. On investigations, HIV- ELISA was positive with CD4 count of  $150/\text{mm}^3$ . Her VDRL and TPHA were positive in high titres of 1:256 and 1:20480 respectively. All other routine investigations were within normal limits. Dark ground illumination from genital and cutaneous ulcers were negative. Histopathology from edge of ulcer revealed hyperkeratosis with basal cell layer vacuolisation. Deeper dermis and subcutaneous tissue showed diffuse and dense infiltration of lymphocytes and plasma cells with occasional epithelioid cell granuloma which was consistent with secondary syphilis CSF VDRL was negative (Figure 3). Thus, a final diagnosis of secondary syphilis (lues maligna variant) in a HIV seropositive host was reached. She was started on antiretroviral therapy (Tab. tenofovir, lamivudin, nevirapine), simultaneously she was given intramuscular inj. benzathine penicillin 1.2 mU in each buttock after sensitivity testing and written informed consent. Total

three such injections were given at weekly interval. In view of her HIV seropositive status with low CD4 count high VDRL and TPHA titres, and non-availability of crystalline penicillin, intravenous inj. ceftriaxon (known to cross blood brain barrier) 1gm twice daily was administered for ten days. Improvement in her lesions was noted within four to five days after first injection of benzathine penicillin. Ulcers healed completely with scarring and hypopigmentation (Figure 4) within 15 days. VDRL titres repeated at 8 weeks demonstrated a two-fold decline (from 1:256 to 1:128). She is under regular clinical and serological monitoring.



**Figure 3: Dense infiltration of lymphocytes and plasma cells with occasional epithelioid cell granuloma.**



**Figure 4: Healed ulcers with scarring and hypopigmentation.**

Commonest site for metastasis was regional lymph node. 8 patients had secondary deposits in liver, 2 were having deposit in anterior abdominal wall and two female were having secondary deposits in both ovaries.

## DISCUSSION

Syphilis occurs worldwide, and the incidence varies significantly with geographic location. The total number of cases of syphilis (P&S, early latent, late, late latent, and congenital) reported to CDC increased 17.7% during 2014–2015 (from 63,453 cases to 74,702 cases). Transmission is mainly by sexual contact. The causative organism, *Treponema pallidum*, was first described in 1905, but because of the inability to culture the organism and the limitations of direct microscopy, serologic testing is the mainstay of laboratory diagnosis. The disease has been arbitrarily divided into several stages. The early stages (primary, secondary, and early latent) are potentially infectious. Secondary stage has varied manifestations like roseola syphilitica, popular, papulosquamous, lichenoid syphilids, moth eaten alopecia, nickels and dimes, split papules, condyloma lata, follicular, pustular, frambosiform and corymbose eruptions.<sup>2</sup>

Lues maligna, is one of the rare manifestations of secondary syphilis was first described by Bazin and Dubuc, who applied this term based on the bizarre clinical features and progressive course of this variant.<sup>3,4</sup> In contrast to tertiary syphilis, the lesions of lues maligna are multiple, have a round or oval configuration, with no tendency to central healing, and exhibit a lamellated, brown-black rupioid crust as seen in our case. Moreover, the early onset of necrotic ulcers in the disease is in contrast to the later occurring gummas of tertiary syphilis. The criteria for diagnosis of lues maligna listed by Fisher et al include strongly positive serological test results, a severe Herxheimer reaction, and an excellent response to antibiotic therapy. In the past, serological anergia was one of the characteristic features of the disease, a concept no longer supported. Extensive clinical differential diagnosis of lues maligna, includes pyoderma gangrenosum, vasculitis, lymphoma, leishmaniasis, leprosy, yaws, and mycobacterial or fungal infections. HIV-infected patients are at greater risk for developing malignant syphilis which may mimic various opportunistic infections and malignancies. However, no other disease process could explain the morphology of the lesions and the rapid involution with penicillin, as observed in our case.

A rise in the sero-prevalence of syphilis has been observed in various studies in India and other countries.<sup>5</sup> The clinical presentation of syphilis varies in minor ways between HIV-infected and HIV-uninfected persons. HIV-infected persons are more likely to present with more than one chancre and with larger and deeper chancres in primary syphilis, and are more likely to manifest signs of secondary syphilis while a chancre is present. Because of its effect on the immune system, HIV infection is thought to increase risk of neurosyphilis. False-negative serologic test results in the setting of the prozone phenomenon and

seronegative syphilis have been reported. Persistently reactive non-treponemal test results, even following appropriate treatment can occur in up to 40%. The CDC recommends CSF examination in all patients with syphilis and neurological symptoms and those with serological and/or clinical evidence of treatment failure. However, a recent study found greater success in detecting asymptomatic neurosyphilis in HIV-seropositive patients by applying serological criteria (CD 4 < 350/cmm and/or RPR titres > 1:32) instead of stage-based criteria.<sup>6</sup> The low CD4 count (150 cmm) and high VDRL titres (1:256) in our patient prompted us to perform CSF examination even in the absence of neurological symptoms or signs.

## CONCLUSION

Malignant syphilis or lues maligna, commonly reported in the pre-antibiotic era, has now seen resurgence with the advent of human immunodeficiency virus (HIV). Immunosuppression and sexual promiscuity set the stage for this deadly association of HIV and *Treponema pallidum* that can manifest atypically and prove to be a diagnostic challenge. The potentially fulminant course of this disease and the dramatic response to penicillin underline the importance of timely diagnosis and treatment institution.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

1. Fisher DA, Chang LW, Tuffanelli DL. Lues Maligna Presentation of a Case and a Review of the Literature. Arch Dermatol. 1969;99(1):70-3.
2. Crissey JT, Denenholz DA. Clinical picture of infectious syphilis. Clin Dermatol. 1984;1:39-61.
3. Rutowitsch MS. Sifilis maligna precoce. Anais brasileiros de Dermatologia. 1980;55:147-50.
4. Armignacco O, Antonucci G, Croce GF, Grillo LR, Valenzano L. Lue ed infezione da HIV. Considerazioni su un caso di sifilide maligna. Recenti progressi in medicina. 1988;79:132-4.
5. CDC. Sexually Transmitted Disease Surveillance 2012. Atlanta: U.S. Department of Health and Human Services, 2013.
6. Ghanem KG, Moore RO, Rampalo AM, Erbeling EJ, Zenilman JM, Gebo KA. Lumbar puncture in HIV infected patients with syphilis and no neurological symptoms. Clin Infectious Dis. 2009;48(6):816-21.

**Cite this article as:** Chavan RB, Belgaumkar VA, Salunke AS, Chirame SS. Rare presentation of the great imitator: a rare case report. Int J Res Dermatol 2017;3:453-5.