JCRMHS

Journal of Clinical Case Reports, Medical Images and Health Sciences

Volume 4 Issue 4, 2023

Article Information

Received date: 10/04/2023
Published date: 08/06/2023

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*Key Words:

HIV, syphilis, skin ulcers, penicillin

Case Report

Ulcerative syphilis (malignant syphilis) in a patient with human immunodeficiency virus infection. About a case

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Abstract

We present the case of a 59-year-old male patient diagnosed with HIV infection since 2008 on intermittent antiretroviral therapy with dolutegravir+lamivudine/abacavir, stage C3 (viral load 29,855 and CD4 count 40) who was admitted for clinical picture of two months of evolution consisting of the appearance of non-pruritic nodular ulcerated cutaneous lesions on the face and lower limbs associated with loss of visual acuity in the left eye, on physical examination with ulcerated lesions with necrotic and hyperkeratotic edges, for which reason they were requested extension studies within which he presented reactive VDRL, which is why a skin biopsy was requested due to the suspicion of malignant syphilis, as well as lumbar puncture and ophthalmological assessment confirming Neurophilis and panuveitis. Finally, the skin biopsy report showed lesions compatible with malignant syphilis, for which he was treated with single-dose benzathine penicillin and crystalline penicillin due to involvement in the CNS and in the eye, with complete resolution of the symptoms.

Introduction

Syphilis is an infectious-contagious disease with a high prevalence worldwide, especially in homosexual patients and with a diagnosis of HIV. Its incidence is higher in Africa, Europe, the lower part of the Pacific West and the American continent, with the United States being the country with the most reported cases. (1) It is produced by the microorganism Treponema Pallidum subspecies Pallidum, belonging to the order of spirochetes, with a size of 6-9 microns, its transmission is sexual with lymphatic and hematogenous dissemination. (1,2)

Regarding the clinic, it is divided into an early stage which includes primary syphilis or hard chancre in which a single, painless, ulcerated lesion with limited edges and a clean bottom associated with inguinal adenopathies is evident; Secondary syphilis manifests as a maculopapular rash with palmoplantar involvement associated or not with systemic symptoms, elevated transaminases, nephrotic syndrome and aseptic meningitis. Latent syphilis is an asymptomatic stage of less than one year of evolution with positive serological tests. The early stage is the most contagious, but with less morbidity. (1) Finally, the late stage in which cardiovascular compromise is evidenced where the aorta is the most affected vessel, generating aortitis and endarteritis, bone involvement is mainly in the tibia, with periostitis in long bones, skin involvement in addition to the Syphilitic roseola are the gummas, these soft, necrotic lesions that can even

DOI: 10.55920/JCRMHS.2023.04.001178

appear in the central nervous system. Ophthalmological damage manifests as uveitis or retinitis and finally meningeal damage with neurophilis, syphilitic vasculitis and chronic meningoencephalitis (2, 5).

Malignant syphilis is a form of syphilitic secondaryism previously considered a manifestation of tertiary syphilis, first described in 1859 in immunocompromised people, however it was not until 1988 that it was designated a cutaneous manifestation in HIV-positive patients in whom lesions were found. ulcerated necrotic and crusted accompanied by systemic symptoms such as general malaise and fever with positive biopsies for leukocyte-clastic vasculitis with spirochetes in special stains (3).

Malignant syphilis is more prevalent in patients infected with HIV or with some degree of immunodeficiency whose incidence is unknown since the literature is scant, making it difficult to determine its frequency and pathogenesis. The first case reported was in a young man with HIV infection, however the immunological status of these patients seems not to be determined for the development of this entity given that 80% of the patients have more than 200 CD4 cells, which shows the relationship between Treponema pallidum with the HIV virus or a functional immunological defect but not a CD4 deficiency (6).

Clinically, it is distinguished by ulcerated papular and nodular lesions with a necrotic or hyperkeratotic surface with an ostraceous appearance, mainly affecting the trunk, extremities, followed by the face, palms, soles, and scalp, leaving varioliform scars, frequently associated with fever and symptoms. constitutional in cases of greater aggressiveness (6,7). Its differential diagnosis should be with other infectious dermatoses, mainly infections by the Herpes family virus, ecthyma gangrenosa, deep mycoses, mycobacteriosis and leishmaniasis, as well as dermatoses of lymphoproliferative origin such as cutaneous T-cell lymphomas, papulosis and lichenoid ptyriasis and others such as Reiter's syndrome (6,8).

The diagnosis of all stages, including malignant syphilis, is by measuring serological tests, among which we have treponemal tests, which are circulating IgG and IgM antibodies directed against the spirochete, are positive 2 weeks after acquiring the infection and are remain positive indefinitely; within these I know they have; the FTA ABS, EIA, CIA, TPPA. On the contrary, non-treponemal tests are IgG and IgM antibodies directed against lecithin, cholesterol and cardiolipin produced when the spirochete is destroyed. These tests are positive 4 weeks after infection, which are reported in dilutions and unlike treponemal tests. , become negative after treatment, examples of these tests are the VDRL (being the most sensitive in the CSF), RPR and TRUST. Since the latter are directed against lecithin, cholesterol and cardiolipin, we can find false positives in tests such as pregnancy, antiphospholipid syndrome, SLE (systemic lupus erythematosus) and even infectious causes such as malaria and brucellosis. They can also cause false negatives given the prozone effect secondary to the agglutination of Treponema in the sample (4).

The traditional diagnostic algorithm is by performing a non-treponemal test and in case of positivity it will be confirmed with a treponemal test, or the reverse algorithm can be performed starting with a treponemal test, if negative, infection is ruled out and in case of If positive, a non-treponemal test will be requested; if this is negative, we can say that it is a past syphilis or a prozone effect (3,4).

The histopathological diagnosis of malignant syphilis is variable, since spirochetes will not be obtained in all the samples, only in 40% of the cases, being similar to the findings in secondary syphilis. The epidermis can be visualized normal, with ulcers or necrosis, in the dermis an infiltrate of plasma cells, lymphocytes, histiocytes, lichenoid-type infiltrate, nodular, granulomatosis that can resemble sarcoidosis, as well as vasculitis (4). Ulcerated lesions could be caused by the affectation which can cause heart attacks in medium-caliber arteries. (7)

The classic diagnostic criteria are macroscopically and microscopically compatible skin lesions, high VDRL titers, and rapid clinical resolution with treatment. In those who have visual or neurological symptoms, a lumbar puncture should always be performed (7,8).

The treatment of choice for the early stages is benzathine penicillin at a dose of 2,400,000 IM in a single dose or ceftriaxone 1 g IV or IM for a total of 10 days. In patients with syphilis of indeterminate time, late latent, three doses of benzathine penicillin are required at a dose of 2,400,000 IM once a week or when there is involvement at the CNS, ocular or auditory level, crystalline penicillin will be started at a dose of 18-24 million each. 24 hours or continuous infusion for 14 days (6,8,9)

Clinical case

We present the case of a 59-year-old male patient diagnosed with HIV since 2008 on intermittent antiretroviral therapy with dolutegravir+lamivudine/abacavir, stage C3 (viral load 29,855 and CD4 count 40) who was admitted due to a clinical picture of two months of evolution consisting of the appearance of non-pruritic ulcerated skin lesions on the face and lower limbs associated with loss of vision in the left eye.

She presented oral cavity with whitish lesions on the tongue and palate, on the skin there were giant ulcerative lesions with hypertrophy and central necrosis on the face

(left zygomatic region, lower lip, see Figure 1), left dorsal region (shoulder, see Figure 2), lower limbs. on the back of the thigh and leg without discharge or fetid odor. Normal neurological examination.

Due to the above, given the characteristics of the lesions, their location and the visual alteration, a nontreponemal test (VDRL) was requested, which was reactive in 8 DILS, for which it was evaluated by ophthalmology who confirmed panuveitis, also with compromise in the central nervous system given lumbar puncture. with lymphocytic pleocytosis, without hypoglycorrhachia, and hyperproteinorrachia (white blood cells 10, red blood cells 1, glucose 48, proteins 74), positive VDRL in CSF, with negative FILM array ruling out common viral or bacterial infection, Cryptococcus antigen and negative studies for M. tuberculosis (ADA and PCR in CSF) for which management was started with crystalline penicillin at a dose of 4,000,000 IV every 4 hours and given the suspected diagnosis of malignant syphilis, we requested a skin biopsy. Cutaneous lymphoma was ruled out, negative tumor markers (carcinoembryonic antigen, alpha feto protein, CA 19-9, with normal contrast CT scan of the chest and abdomen.

Finally, the skin biopsy confirmed malignant syphilis (left cervical and preauricular biopsy) for which management was given with benzathine penicillin one dose every week for three doses of 2,400,000 IM and also completed 21 days of therapy with crystalline penicillin given ophthalmological compromise and in CNS with which it presents significant clinical improvement.

Subsequently, after 3 weeks of treatment, the involution of the lesion was evidenced (see Figure 4).

Figure 1: *Lesion in the left zygomatic región*

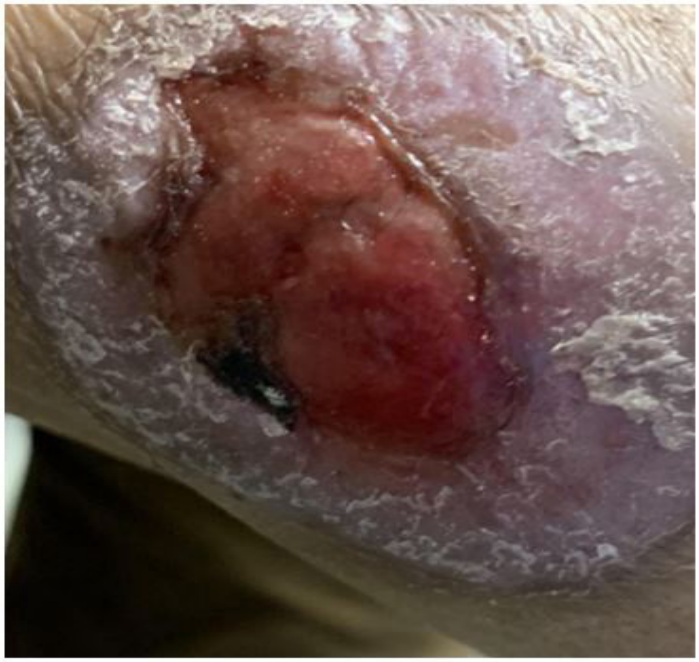

Figure 2: *Lesion in the posterior region of the left lower limb*

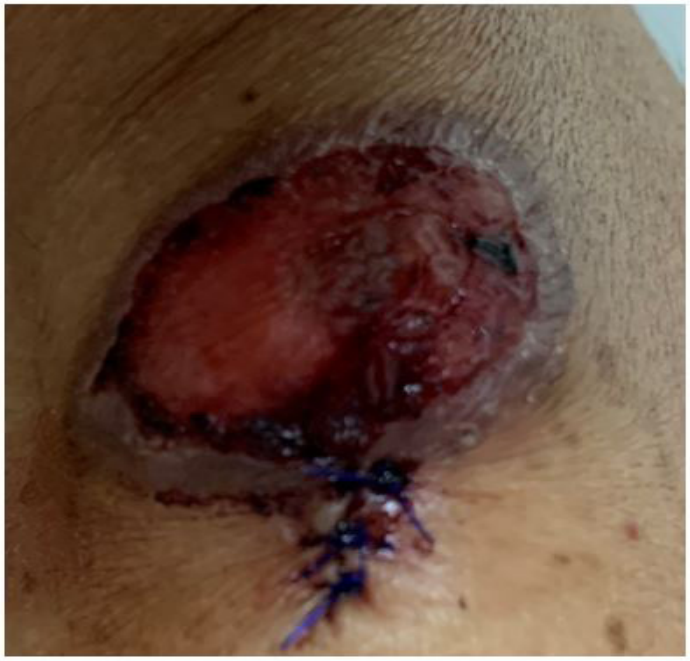

Figure 3: *Lesion in the left dorsal region*

DOI: 10.55920/JCRMHS.2023.04.001178

Discussion

Malignant syphilis is a rare form of secondary syphilis, with few data in the literature about its incidence, however it is known to be more prevalent in men, mainly MSM with HIV coinfection without racial predilection, with an incubation period of 4 weeks (11) Classically, it has various skin manifestations, beginning as papules that later become nodules with pustules to finally develop central necrosis with extensive ulceration, affecting the face, scalp, trunk, and extremities (11-12).

Patients may present systemic symptoms such as fever, arthralgia, myalgia, generalized adenopathies or concomitantly present hepatitis, ocular involvement, pulmonary or central nervous system involvement with no mucosal involvement, as in the aforementioned case, the patient presented panuveitis and neurolues . It is more common in patients with HIV or with some degree of immunosuppression (11-12).

Its pathophysiology is secondary to an exaggerated response of the immune system against Treponema with defects in the function of macrophages and the innate response. (9-12)

The diagnosis is clinical, serological (treponemal and non-treponemal tests) and histopathological, where edema, endothelial proliferation, fibrinoid accumulation in the vessels with occlusion is evidenced, generating infarction and necrosis of the dermis and epidermis. In the exposed case, it was confirmed by reactive VDRL associated with a skin biopsy, which confirmed the involvement of syphilis in the skin lesions.

The treatment of choice is a dose of 2,400,000 IM of benzathine penicillin; in case of allergy, ceftriaxone is recommended. When there is a compromise in the central nervous system, the indication is crystalline penicillin. Our patient presented clinical and paraclinical improvement after handling with penicillins, leaving only a residual pigmented lesion. (Figure 5)

Conclusions

In conclusion, malignant syphilis is a rare entity that is associated with immunocompromised patients and despite the severity of the lesions it has a good prognosis with treatment, so it should always be taken into account as a differential diagnosis in a patient with large ulcerated lesions. Associated with positive serologic tests for syphilis with or without involvement of the central nervous system.

References

1. Peeling, r., mabey, d., kamb, m. et al. syphilis. nat rev dis primers 3, 17073 (2017).
2. Hook, Edward W. (2017). syphilis. the lancet, 389(10078), 1550–1557. doi: 10.1016/s0140-6736(16)32411-4
3. Dalia ibarra-morales,1 natalia pérez-leal,1 dayra jiménez-mendoza,1 silvia trujillo-novoa,3 mario sierragarduño,2 paula torres-camacho,1 maría ivonne arellano-mendoza1 malignant syphilis that resembles a polymorphous erythema on a patient with acquired immunodeficiency syndrome med int mex. 2020 Jan-Feb; 36(1): 101-106
4. Plowing lasagabaster, maider; Otero Guerra, Luis (2019). syphilis. infectious diseases and clinical microbiology, (), s0213005x19300072–. doi: 10.1016/j.eimc.2018.12.009
5. Morales-múnera, c.e.; Fuentes-Finkelstein, P.A.; Vall Mayans, m. (2015). fr – syphilis: update on diagnostic and therapeutic management. Actas Dermo-Sifiliográficas, 106(1), 68–69. doi: 10.1016/j.ad.2014.06.007
6. Fustà-novell, x.; morgado-carrasco, d.; barreiro-capurro, a.; manzardo, c.; alsina-gibert, m.; fuertes-de vega, irene; white, jose-luis; González, Anna; brown, assumption; Alvarez, Miriam; bosch, jordi (2018). malignant syphilis, a syphilis presentation to consider. dermo-sifiliographic records, (), s0001731018303545–. doi: 10.1016/j.ad.2018.02.024
7. Vinay, keshavamurthy; kanwar, amrinder jit; narang, tarun; Saikia, Uma Nahar (2013). malignant syphilis. international journal of infectious diseases, 17(10), e930–e931. doi: 10.1016/j.ijid.2013.03.020
8. Malignant syphilis in a patient with human immunodeficiency virus infection María Cecilia Garbarino1 , Carla Trila2 , Leandro Heffner1 , María Eugenia Cantón1 , Florencia Quadrana3 , Marcelo Zylberman. medical clinical division, 2 pathology division, 3 dermatology unit, acute general hospital dr. cosme argerich, buenos aires, argentina. medicine (buenos aires) 2020; 80: 714-717
9. Corti m, palmieri o, maronna e. Secondary malignant syphilis, rupioid clinical form, in an HIV-positive patient. rare presentation of secondary lues. Case report and literature review. rev argent dermatol 2014; 95: 22-6.
10. Wappner, Diego; carbia, sergio; Gioseffi, Luis; Schroh, Roberto; Losso, Marcelo H. (1997). answer to photo quiz. clinical infectious diseases, 25(6), 1447–1447. doi:10.1086/516143
11. Romero-jime´nez mj, sua´rez lozano i, fajardo pico´ jm, baro´n franco b. Malignant syphilis in patients infected with the human immunodeficiency virus (HIV): description of a case and review of the literature. an internal med. 2003; 20: 373–6
12. Fernández-guarino m, fernández de la mora a, gonzález garcía c, fed up castaño a, moreno izquierdo r, jaén olasolo p. malignant syphilis in a patient infected with the human immunodeficiency virus (hiv). proceedings dermosifiliogr 2006; 97: 400-3.
13. Cid pm, cudós is, zamora vargas fx, merino mj, pinto ph. Pathologically confirmed malignant syphilis using immunohistochemical staining: report of 3 cases and review of the literature. sex transm dis 2014; 4