Nodular tertiary syphilis in an immunocompetent patient*

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Abstract: Acquired syphilis can be divided into primary, secondary, latent, and tertiary stages. About 25% of patients with untreated primary syphilis will develop late signs that generally occur after three to five years, with involvement of several organs. The authors present an immunocompetent female who developed a tertiary stage syphilis presenting with long-standing nodular plaques.

Keywords: Sexually transmitted diseases; Syphilis, cutaneous; Treponemal infections

INTRODUCTION

Syphilis is an infectious disease, sexually transmitted, caused by *Treponema pallidum*.[1] Its incidence has been reduced with the introduction of penicillin for its treatment, however it has been sharply increasing with the advent of human immunodeficiency virus (HIV) infection.[1,2] Approximately 25% of the patients with untreated primary syphilis develop symptoms of tertiary syphilis.[1-4] They usually occur three to five years after primary infection and involve several organs, mainly skin, heart and central nervous system.[3] In the antibiotic era, it is extremely rare to find cases of tertiary syphilis. This fact, added to the scarcity of reports in the literature in recent years, has caused lack of familiarity with clinical and histopathological aspects of tertiarism.

CASE REPORT

A 40-year-old woman had been presenting, in the last two years and six months, nodular lesions of progressive growth in the upper limbs, without associated symptoms or previous treatments. She denied any compatible symptoms or signs of primary and secondary phases or recurrence of syphilis. There was no history of injectable drug use, blood transfusion or convulsions. The dermatological examination showed extensive centrifugal-growth erythemato-violaceous nodular plaques in the upper limbs (Figure 1).

Anatomopathological examination of left arm biopsy showed atrophic epidermis and vacuolar interface dermatitis. The dermis and the subcutaneous were filled with a mixed, dense and diffuse inflammatory infiltrate, composed of lymphocytes, histio-

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cytes and abundant plasmocytes. The vascular alterations included vascular proliferation and clogged vessels, with tumescent endothelium and walls infiltrated by inflammatory cells, besides erythrocyte extravasation. The infiltrate was separated from the epidermis by a *grenz* band (Figure 2). Caseous necrosis was not noticed. Fite-Faraco and Grocott stainings were negative for BAAR and fungi, respectively.

Due to histological suspicion of syphilis, FTA-ABS and VDRL were requested, which resulted as reagent (VDRL in a titer of 1:32). Anti-HIV 1 and 2 were negative. Patient was submitted to treatment with benzathine penicillin (2,400,000 IU per week, for three weeks), with complete remission of lesions (Figure 3). Electrocardiogram, echocardiogram, X-ray of long bones, eye examination, ultrasound of the abdomen, blood count and tests of hepatic and renal function showed no alterations.

DISCUSSION

Tertiary or late syphilis is a rare systemic disease which may present mucocutaneous, cardiac, ophthalmic, neurological or osseous involvement.[1,3] Skin is the most affected organ, and its clinical presentation varies according to the level of involvement, which may be hypodermic (syphilitic gumma) or dermoepidermal (pso-

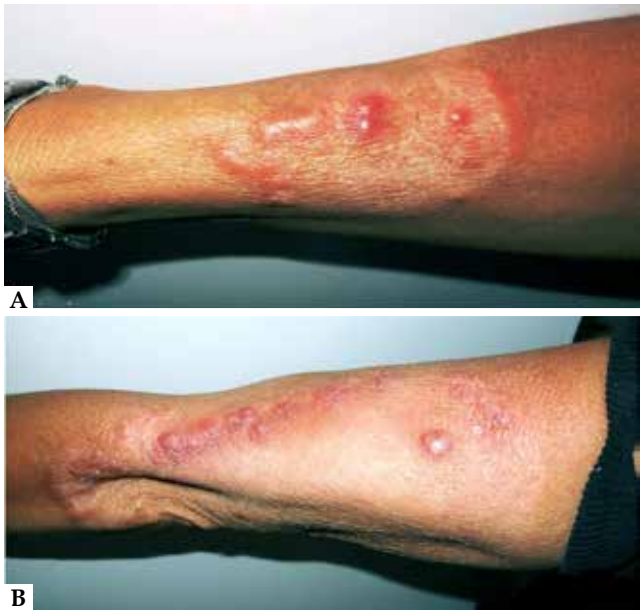

Figure 1: Arcuate papulonodular lesions in the right forearm **(A)** and left arm **(B)**

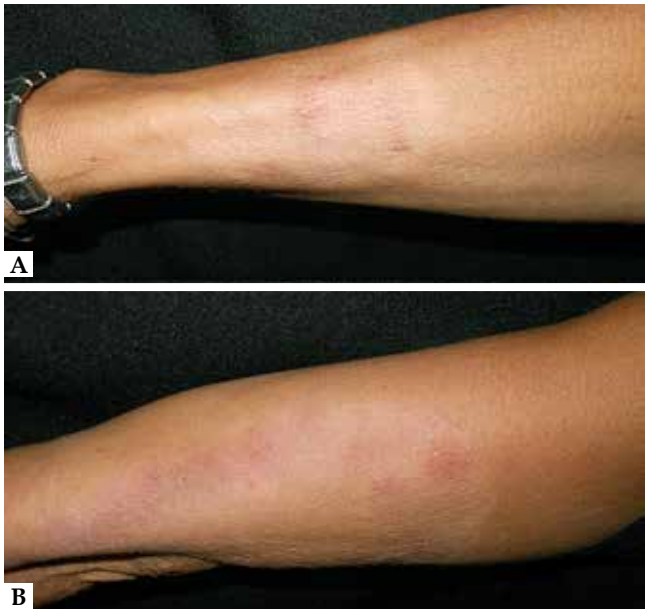

Figure 3: A and B: Clinical aspect of lesions after treatment

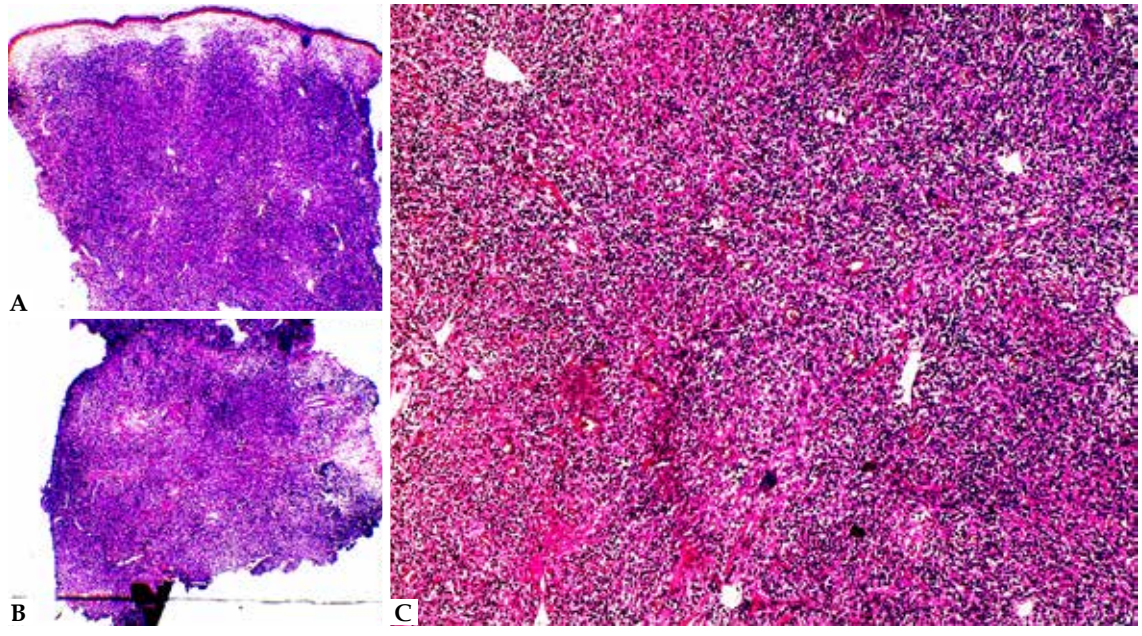

Figure 2: A and B: Anatomopathological examination showed atrophic epidermis and vacuolar interface dermatitis. The dermis and the subcutaneous were filled with a mixed, dense and diffuse inflammatory infiltrate, composed of lymphocytes, histiocytes and abundant plasmocytes, separated from the epidermis by a grenz band. C: Detail of the dense inflammatory infiltrate and vascular proliferation (HE, 400x).

riasiform plaques or nodules).[3] It is then classified into two types: gummatous and nodular. The nodular form, as in the present case, consists of painless, hardened erythematous nodules, of varying sizes, which may occur in any site on the skin. They may remain isolated or coalesce to form plaques. Multiple nodules may be distributed in an arciform pattern, with a predilection for the face, interscapular areas and extremities.[3] The gummatous form presents itself as firm, painless subcutaneous nodules which later develop ulcerations and drainage of necrotic materials.

These two forms, which show great clinical and morphological diversity and show the involvement of the dermis and the subcutaneous by the granulomatous process, may simulate several diseases.[4-6] Differential diagnosis is broad and includes infectious, inflammatory and neoplastic disorders, such as sarcoidosis, lupus vulgaris, atypical mycobacterial infections, Jorge Lobo's disease, sporotrichosis, leishmaniasis, Hansen's disease, granuloma annulare, pyoderma gangrenosum, lymphoma and pseudolymphoma.[3,5,6] Spontaneous remission is rare, and consequently, recurrences in the skin become even more destructive with time. In most cases, however, lesions are resolved quickly when treated.

The histopathologic features of tertiary syphilis lesions consist, in the nodular form, in an abundant plasmocytic infiltrate, with formation of granulomas and many times caseous necrosis. In the syphilitic gumma an important caseous necrosis occurs, surrounded by numerous giant cells and dense inflammatory infiltrate, composed of lymphocytes and plasmocytes, which extends itself to the subcutaneous.[3] In the case presented, the important density of the infiltrate is highlighted, which, associated with vascular lesions and abundant plasmocytes, led to suspicion of syphilis diagnosis.

Nontreponemal serological tests may present negative results or low titer for late syphilis, specially in cardiovascular or neurosyphilis forms.[3,7-9] Treatment is carried out with benzathine penicillin at a dose of 2,400,000 IU per week, intramuscular, for three weeks.[9] In any phase of the disease, the possibility of other sexually transmitted diseases being present must be considered. Thus, serology for HIV and screening for hepatitis B and C ought to be recommended for all patients.[3] Cases of tertiary syphilis in patients free from HIV have been rarely described in recent years.[3-8] The case presented emphasizes the need of keeping syphilis in the list of differential diagnoses for long-duration papulonodular lesions, to ensure early diagnostic and treatment, even in the absence of previous history of syphilis or immunosuppression.❑

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