Lues Maligna in an Immunocompetent Female

Sir,

French dermatologist Pierre Bazin was the first to coin the term malignant in a case of secondary syphilis. About 5 years later, it was defined as malignant syphilis by Dubue.[1] Malignant syphilis or lues maligna is an uncommon form of secondary syphilis, which usually occurs in immunosuppressed patients, mainly those who have HIV infection.[2,3] Skin lesions include pustules, nodules, and ulcers.[4] It may involve mucosa, lymph node, liver, and spleen. It is rarely reported in immunocompetent individuals. We are reporting a case of lues maligna in an immunocompetent female.

A 35-year-old married woman presented with skin lesions, fever, malaise, and arthralgia for 3 weeks. Dermatological examination revealed; skin colored to erythematous macules, papules and nodules. The relatively older papules and nodules had developed crust, collaret of scale, and few had ulceration on surface. The lesions were present all over the body with increased predilection for the extremities, trunk, face, genitalia, palm, sole, and scalp [Figure 1]. She also had curdy white vaginal discharge sticking to her vaginal wall associated with itching suggestive of candidiasis. Rest of mucosal surfaces were free of any lesions. The inguinal, axillary, and cervical lymph nodes were 1.5 cm × 1 cm enlarged and nontender. Other systemic examinations were unremarkable. Her husband denied history of extramarital sexual contact and was free of any skin and mucosal lesions suggestive of syphilis.

The hematological parameters were within normal limits. Her venereal disease research laboratory (VDRL) and Treponema Pallidum Hamagglutination Assay(TPHA) tests were reactive with titers 1:64 and 1:160 dilutions, respectively. Test for HIV was nonreactive. Her husband's VDRL test was reactive at 1:32 dilutions. Histopathology of skin lesion from leg showed the presence of plasma cells in the vessel wall and thrombosed vessels in the dermis with endarteritis [Figure 2a and b]. Based on clinical, histopathological, serological, and associated systemic features, a diagnosis of lues maligna with vaginal candidiasis was made. The patient was sensitive to test dose of penicillin and hence, she did not give consent for injectable penicillin. She was treated with Doxycyline 100 mg twice daily for 21 days for syphilis and fluconazole 150 mg stat dose for vaginal candidiasis. The skin lesions of syphilis healed by 60% within 14 days of initiation of medication without Jarisch-Herxheimer reaction and vaginal candidiasis improved completely. The spouse also was treated with doxycycline 100 mg twice daily for 21 days, as he refused to give consent for injectable penicillin. Repeat serology of patient and her husband could not be done as both were lost to follow up.

Malignant syphilis is referred to as syphilis maligna praecox, lues maligna, and rupioid syphilis. It is a rare form of secondary syphilis and differs from classical manifestations of secondary syphilis by exuberant, pleomorphic, ulcerated, and necrotic lesions and associated systemic symptoms.[4] These lesions can occur on the entire body with increased predilection for extremities and face.[5,6] Because of wide range of skin and mucosal lesion it is often confused for other diseases.

The syphilitic lesions have been reported to mimic cutaneous T-cell lymphoma, mycosis fungoides, persistent gyrate erythema, lichen planus, parapsoriasis lichenoides et varioliformis acuta, erythema exudativum, psoriasis, pustular psoriasis, histiocytoma, and sarcoidosis clinically and histopathologically.[7-9] However, the diagnosis of lues maligna is based on three criteria: clinical and histopathological characteristics, presence of high titre

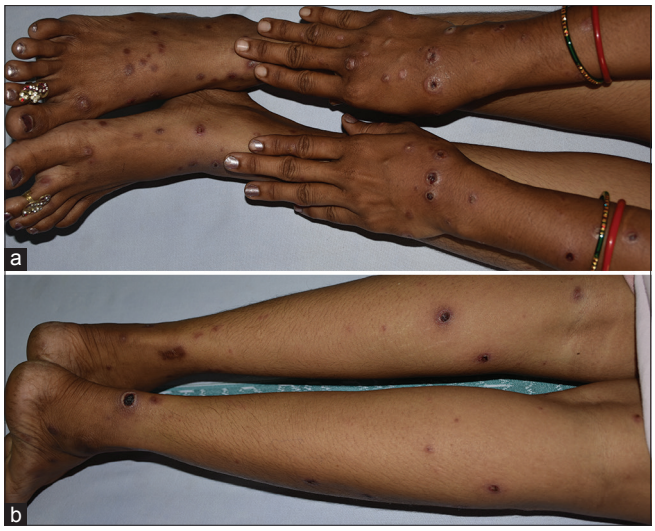

Figure 1: (a) Papule and nodules with crust on surface on extremities. (b) Nodules with crust on surface

of antibodies in VDRL or a similar test, and intense and severe Jarisch–Herxheimer reaction and rapid resolution of lesions with adequate therapy.[10]

In the present case, the skin lesions were in the form of nodules and papules with ulceration on surface of few which usually occurs due to end arteritis obliterans in syphilis. In addition, reactive VDRL, positive TPHA test, corroborative histopathology, associated systemic features, and rapid response to doxycycline confirmed the diagnosis of lues maligna. Although severe Jarisch–Herxheimer reaction after treatment is one of the criteria for lues maligna, our patient did not develop the same with treatment, which is quite unusual.

Malignant syphilis is frequently associated with HIV- co-infected patients.[2,3] It has been suggested that in patients co-infected with HIV, the immunodeficiency favors the predominance of the virulence of the agent in the agent-host contest.[2] Other predisposing factors include malnutrition, alcohol abuse, and concomitant debilitating illnesses.[11,12] Our case did not have any predisposing factor and was negative for HIV. In such cases, it is difficult to suspect and diagnose the case as lues maligna. However, widespread pleomprphic, nodulo-ulcerative lesions along with associated systemic symptoms even in an immunocompetent individual should lead to suspicion of lues maligna, so that the disease can be diagnosed and treated at the earliest. Simultaneously, the partner should be treated to prevent reinfection to the spouse and transmission to community as we did in our case. On review of literature, we found few reports of lues maligna in immunocompetent adults, hence our case will add to the pool of evidence.

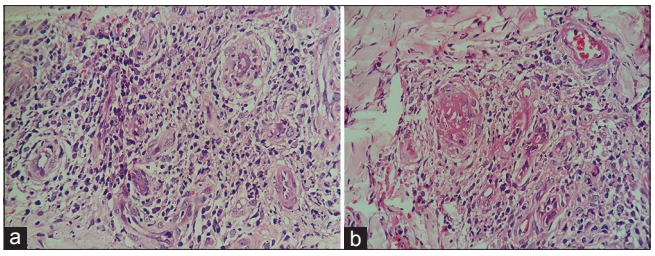

Figure 2: (a) Hematoxylin and eosin; 40×: Dermis showing endothelial swelling with infiltration of histiocytes and plasma cells. (b) Hematoxylin and eosin; 40×: Dermis showing proliferation of blood vessels with obliteration of vascular lumina with infiltration of plasma cells and histiocytes

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Swetalina Pradhan, Chandra S. Sirka, Maitreyee Panda, Manas Baisakh[1]

Department of Dermatology and Venereology, All India Institute of Medical Sciences, [1]Department of Pathology, Prolife Diagnostics, Bhubaneswar, Odisha, India

Address for correspondence:
Dr. Chandra S. Sirka,
Department of Dermatology and Venereology, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India.
E-mail: csirka2006@gmail.com

References

1. Cripps DJ, McDonald R. Syphilis mligna praecox the first patient seen at MGH in 1821. N Eng J Med 1977;296:695.
2. Corti M, Solari R, De Carolis L, Figueiras O, Vittar N, Maronna E. Sífilismalignaen un paciente con infecciónpor VIH. Presentación de un caso y revisión de la literatura. Rev Chilena Infectol 2012;29:678–81.
3. Watson KM, White JM, Salisbury JR, Creamer D. Lues maligna. Clin Exp Dermatol 2004;29:625–27.
4. Passoni LF, de Menezes JA, Ribeiro SR, Sampaio EC. Lues maligna in an HIV-infected patient. Rev Soc Bras Med Trop 2005;38:181–4.
5. Marra CM. Syphilis and HIV infection. Semin Neurol 1992;12:43–7.
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;36:512-4.
7. Yamashita M, Fujii Y, Ozaki K, Urano Y, Iwasa M, Nakamura S, *et al*. Human immunodeficiency virus-positive secondary

syphilis mimicking cutaneous T-cell lymphoma. Diagn Pathol 2015;10:185.
8. Hodak E, David M, Rothem A, Bialowance M, Sandbank M. Nodular secondary syphilis mimicking cutaneous lymphoreticular process. J Am Acad Dermatol 1987;17:914-7.
9. Gevorgyan O, Owen BD, Balavenkataraman A, Weinstein MR. A nodular-ulcerative form of secondary syphilis in AIDS. Proc Bayl Univ Med Cent 2017;30:80–2.
10. Kumar B, Muralidhar S. Malignant syphilis: A Review. AIDS Patient Care & STDs 1998;12:921-6.
11. Belda Jr W, Dias MC, Zolli CA, Santos Junior MFQ, Siqueira LFG. Sifilis maligna precoce: A proposito de um caso. An Bras Dermatol 1990;65:147-50.
12. Avelleira JCR, Rangel GCB. Syphilis: Diagnosis, treatment and control. An Bras Dermatol 2006;81:111-26.

Access this article online

Website: www.idoj.in

Quick Response Code

DOI: 10.4103/idoj.IDOJ_277_17

How to cite this article: Pradhan S, Sirka CS, Panda M, Baisakh M. Lues maligna in an immunocompetent female. Indian Dermatol Online J 2018;9:344-6.

Received: October, 2017. **Accepted:** January, 2018.