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Malignant Syphilis:
A Rare Case of Early Secondary Syphilis in an Immunocompetent Patient

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ABSTRACT
Early malignant syphilis is an uncommon form of secondary syphilis and characterized by pleomorphic multiple round-to-oval papules, some with necrosis, and associated with systemic signs and symptoms. Usually seen in immunosuppressed patients, mainly those infected with HIV, it can also be observed in immunocompetent patients.
We report a case in a young healthy woman with the characteristic features of the disease and with favourable skin lesion evolution after appropriate treatment with penicillin.

LEARNING POINTS
- Skin lesions can be caused by numerous systemic diseases.
- Syphilis is the 'great mimicker' and should always be considered as a diagnosis in those with skin lesions.

KEYWORDS
Early secondary syphilis, skin lesions, immunocompetent

INTRODUCTION
Early malignant syphilis (EMS) is a rare form of secondary syphilis which is frequently seen in patients co-infected with HIV but also presenting in those with malnutrition, alcohol abuse or debilitating illness [1, 2]. The incidence of EMS has been increasing since the beginning of the HIV epidemic and should be considered as a diagnosis in any patient with suspicious cutaneous lesions [3]. More rarely, it also occurs in immunocompetent patients. First described by Bazin in 1859 and Dubuc in 1864 as a nodular variant of syphilis [1], its presentation differs from the classic manifestations of secondary syphilis as it is more severe and has different lesion morphology. EMS, referred to in the literature as syphilis maligna praecox, rupioid syphilis or lues maligna, is characterized by pleomorphic multiple round-to-oval papules, papulopustules, or nodules with ulceration [1–4]. Cutaneous lesions are preceded approximately 4 weeks earlier by prodromal fever, headache, arthralgia, myalgia and weight changes [1, 3].
The typical morphology and histopathology of the lesions, positive serological tests for syphilis, and resolution of the lesions after treatment with penicillin support the clinical diagnosis of EMS [2, 3].
The authors report a case of EMS in a young immunocompetent woman.

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CASE DESCRIPTION

A 29-year-old woman was admitted to the emergency department with a 6-week history of fever (38.5°C), loss of appetite and myalgia followed by the appearance of an extensive erythematous nodular rash affecting the scalp, face (*Fig. 1*), neck, axilla, trunk, back (*Fig. 2*), palms (*Fig. 3*) and soles, and perineal area. Some lesions, especially in the perineal area, were ulcerated and painless. She reported a history of sexual intercourse with her husband only. Apart from hypothyroidism, she did not present any other comorbidities.

On clinical examination, the patient was afebrile and vital signs were stable. She presented in a good general condition with multiple skin lesions appearing as erythematous nodules, some coalescing into large reddish plaques, in addition to some ulcerative lesions with necrosis located in the perineal area. The palms and soles had small erythematous lesions. Oral and genital mucosa were spared. There were no ophthalmological or neurological abnormalities.

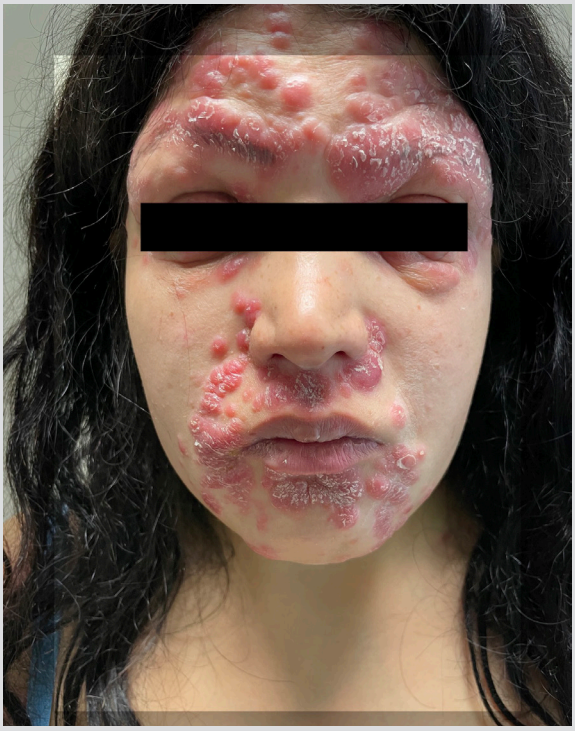

Figure 1. Erythematous nodular rash affecting the face

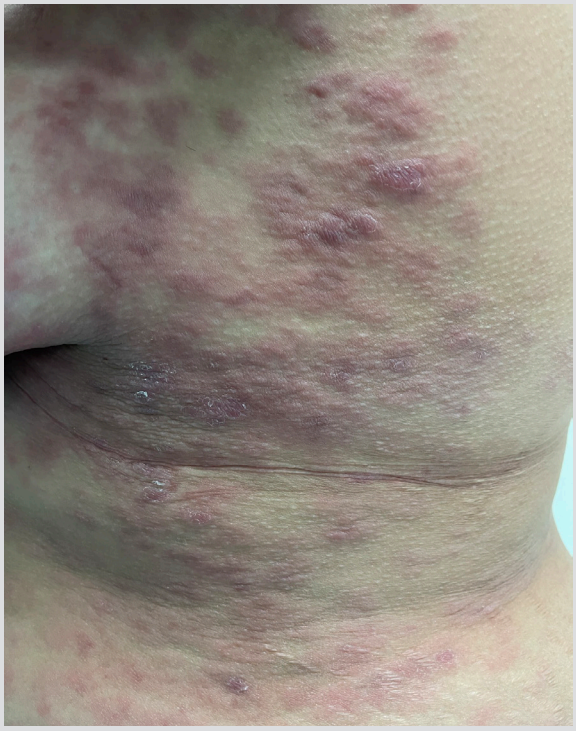

Figure 2. Erythematous nodular rash affecting the trunk

Serum investigation showed a normal complete blood count and reactive rapid plasma reagin (RPR) test (1:128) and a positive Treponema pallidum test. Serology for HIV, HCV and HBV infection was negative. An autoimmunity screen was negative.

A skin biopsy was taken from the back and histological examination with haematoxylin and eosin staining showed subcorneal pustules and non-caseating granulomas in the dermis with macrophages, Langerhans cells and a lymphoplasmacytic infiltrate.

In view of the clinical picture, the patient was treated with 2.4 million units of penicillin G benzathine, once weekly for 3 weeks.

There was no Jarisch-Herxheimer (JH) reaction and the lesions rapidly healed during the month after penicillin administration. Three months after treatment, the serological RPR titre had fallen to 1:4.

DISCUSSION

Syphilis is a systemic human disease caused by T. pallidum subspecies and classified as acquired or congenital. Acquired syphilis (primarily by sexual contact) is divided into early and late syphilis. Early syphilis includes primary, secondary and early latent syphilis [5]. EMS is an uncommon form of secondary syphilis and the exact incidence is not known [6]. EMS was described well before the HIV pandemic as an aggressive nodular variant of secondary syphilis. It was a common diagnosis in the 1700s, and usually seen in cachectic patients with tuberculosis. Its unusual clinical manifestation appears to be related to poor health condition, malnutrition, and inappropriate use of immunosuppressants or antibiotics [6].

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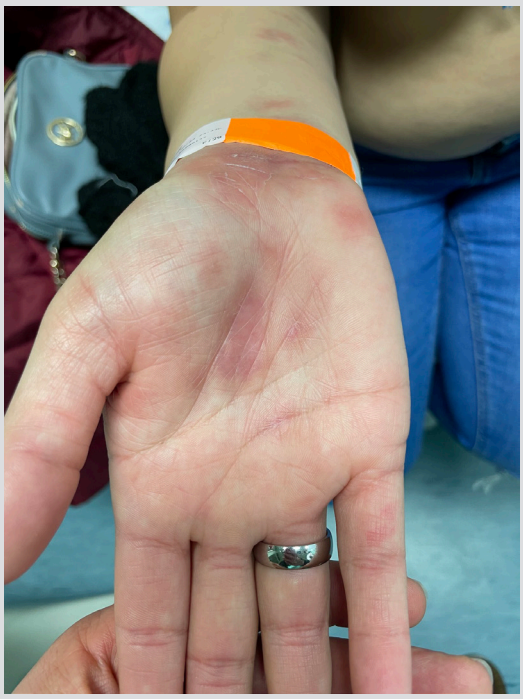

Figure 3. Erythematous rash affecting the palm

More cases have been seen recently in people with HIV infection than previously. Patients with HIV/acquired immunodeficiency syndrome are 60 times more likely to present with this form of syphilis [3]. The pathogenesis of EMS is not fully understood, but the general opinion is that in those co-infected with HIV, the immunodeficiency favours the predominance of the virulence of the agent in the agent–host contest[1,2]. This is coherent with its occurrence in patients who abuse alcohol or drugs, are malnourished or have debilitating disease. Infection with more virulent strains of Treponema, even if difficult to demonstrate, can cause EMS in immunocompetent patients [1, 2], as in the case described.

The onset of EMS is characterized by prodromal symptoms and lesions are described as erythematous-violaceous or reddish-coppery in colour and can appear as papules, nodules or blisters depending on the phase of evolution. This is usually followed by central necrosis. These lesions mainly affect the trunk and extremities, but can also be present on the scalp, face, palms and soles [1–4, 6, 7].

Clinical suspicion of EMS can be supported by three criteria: clinical and histopathological characteristics; high titres of antibodies in VDRL or similar tests; and intense and severe JH and rapid resolution of lesions [1–6].

The differential diagnosis can be difficult and must include other infectious skin diseases (from herpesviruses, ecthyma gangrenosum, deep mycosis, mycobacteriosis and leishmaniasis), lymphoproliferative skin diseases (cutaneous T-cell lymphoma, lymphomatoid papulosis and pityriasis lichenoides) and other diseases such as Reiter syndrome [7].

Although recommendations for treatment of EMS are lacking in the literature and elsewhere [1–7], the latest European guidelines recommend the use of one single dose of 2.4 M units of penicillin G benzathine [5] for the treatment of secondary syphilis.

In conclusion, syphilis is still a common disease. As it can appear with a variety of clinical manifestation and mimic several dermatoses, it should always be considered as a possible diagnosis.

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