IDCases 36 (2024) e01943

Contents lists available at ScienceDirect

IDCases

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Case illustrated

Malignant syphilis requiring differentiation from Kaposi's sarcoma

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ARTICLE INFO

Keywords:
Malignant syphilis
Rupioid syphilis
Kaposi's sarcoma
HIV infection
Atypical cutaneous lesions
Secondary syphilis

ABSTRACT

Malignant syphilis (MS) is a rare variant of secondary syphilis. Also known as rupioid syphilis, MS is characterized by the presence of multiple papules, papulopustules, black lamellate crust that may resemble an oyster shell, or nodules with ulceration lacking central clearing. MS is often associated with immunodeficiency and frequently co-occurs with HIV infection. We here report a case of MS in a patient with HIV infection. HIV infection can cause atypical clinical symptoms of syphilis. In this case, unlike previous cases, cutaneous lesions of MS were limited to the face, making the diagnosis challenging based on clinical findings alone. However, his laboratory findings, appearance of the Jarisch-Herxheimer reaction, and a dramatic response to antibiotic therapy are characteristic of MS, making the diagnosis even more certain. Our case suggests the importance of physicians considering the possibility of MS when observing black-crusted lesions.

Case report

A 40-year-old man who has sex with men with a medical history of hepatitis B presented to a hospital with a 2-month history of cutaneous lesions on his face, 2-week histories of a fever and diarrhea, and weight loss (10 kg over several months). On a physical examination, vital signs were within the normal range, and he exhibited cervical lymphadenopathy. The black-crusted lesions on his face (left side of Fig. 1) had started as pustules 60 days earlier. No lesions were observed on the oral mucosa, trunk, limbs, palms, or soles of his feet. However, the patient had ulcerative lesions on the penis and flat condylomas of the scrotum. Blood tests revealed elevated levels of rapid plasma regain (RPR) (1:256 titer) and a *Treponema pallidum* hemagglutination assay (1:40960 titer). His human immunodeficiency virus (HIV) RNA titer was 8.4×10^5 copies/mL, and his $CD4^+$T-cell count was 836 /μL. From the cutaneous presentation, tumors, fungal infections, or syphilis were considered in the differential diagnosis, but none of the diseases were atypical. A histopathological examination of the skin revealed histological findings consistent with malignant syphilis [1] (Fig. 2a, b). An immunohistochemical analysis of anti-*Treponema* antibodies showed clusters of spiral-form elements (Fig. 3). Therefore, the patient was diagnosed with malignant syphilis and an HIV infection. He received a single 2400,000 IU intramuscular injection of penicillin. He experienced a Jarisch-Herxheimer reaction (JHR). One month later, his lesions disappeared, and there had been no relapses at the six-month follow-up (right side of Fig. 1).

Malignant syphilis (MS) is a rare variant of secondary syphilis. Also known as rupioid syphilis, MS is characterized by the presence of multiple papules, papulopustules, black lamellate crust that may resemble an oyster shell, or nodules with ulceration lacking central clearing [1,2]. MS is often associated with immunodeficiency and frequently co-occurs with HIV infection [3]. Low CD4 counts may favor MS; however, there have been reports of MS in HIV patients with normal CD4 counts [4]. HIV infection can cause atypical clinical symptoms of syphilis [3]. In this case, unlike previous cases, cutaneous lesions of MS were limited to the face, making the diagnosis challenging based on clinical findings alone. However, his laboratory findings, appearance of the JHR, and a dramatic response to antibiotic therapy are characteristic of MS [2], making the diagnosis even more certain. Our case suggests the importance of physicians considering the possibility of MS when observing

Abbreviations: RPR, rapid plasma antigen; HIV, Human Immunodeficiency Virus; JHR, Jarisch-Herxheimer reaction; MS, Malignant syphilis.

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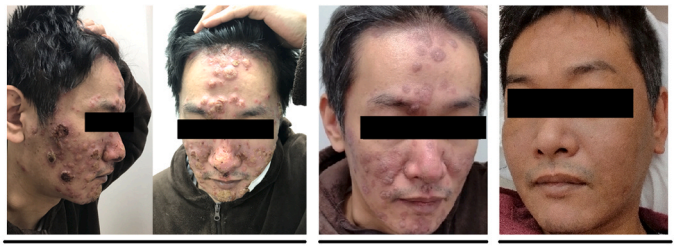
https://doi.org/10.1016/j.idcr.2024.e01943
Received 1 October 2023; Received in revised form 19 March 2024; Accepted 13 April 2024
Available online 16 April 2024



Days since treatment (injection of penicillin)	1day	10days	209days
levels of rapid plasma regain (RPR)	1:256 titer		1:2 titer
CD (cluster of differentiation) 4^+ T-cell count	836 /μL		639 /μL
HIV RNA titer	84100 copy/mL		<20 copy/mL

Fig. 1. Cutaneous lesions with black crusts. The lesions extended to the earlobe. He received an injection of penicillin. One month later, his lesions disappeared, and there had been no relapses at the six-month follow-up. He was not being treated for HIV as well as HBV. His HBs antigen was negative, HBs antibody was positive, HBc antibody was positive, HBe antigen was negative, HBe antibody was positive, and HBV-DNA was less than 1.0 log IU. Therefore, his HBV was not in an active state, and we believed it was not affecting his current symptoms. On day 43, he began taking bictegravir sodium, emtricitabine, and tenofovir alafenamide fumarate orally.

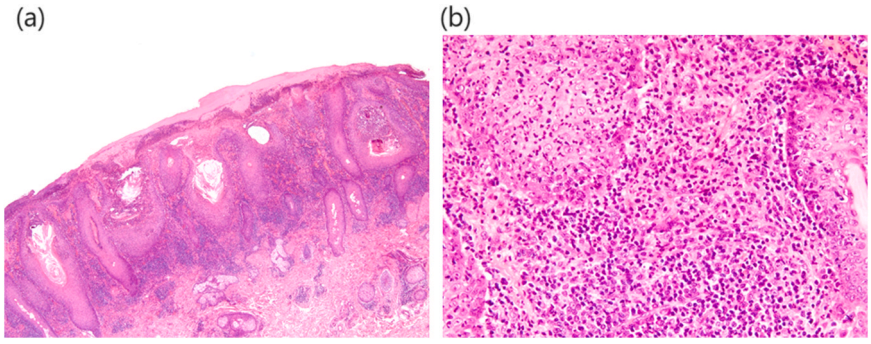


Fig. 2. Histopathological findings of the black-crusted lesion on his chin (a: ×20, b: ×200). The accumulation of neutrophils, hyperplasia, and ulceration were observed in the epidermis. In the dermis, inflammatory cells consisted of plasma cells, lymphocytes, and neutrophils.

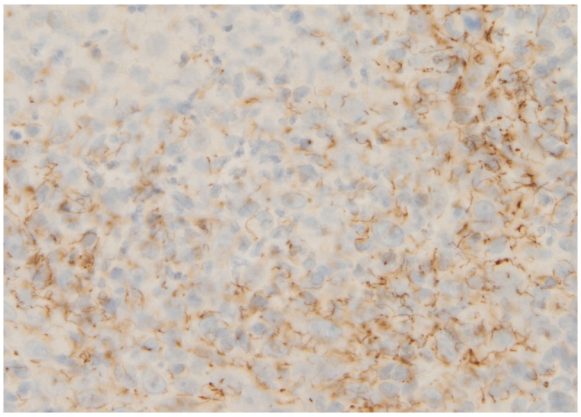


Fig. 3. Immunohistochemical staining with anti-*Treponema* antibodies (×400). Clusters of spiral-form elements were observed in the epidermis and dermis.

black-crusted lesions.

Ethical approval

Not applicable.

Funding

Not applicable.

CRediT authorship contribution statement

Shigefumi Maesaki: Supervision. **Nobuhito Okumura:** Methodology. **Fumito Inayoshi:** Methodology. **Keita Okamoto:** Methodology. **Masaaki Takeji:** Supervision. **Hiroshi Yamaguchi:** Methodology. **Kazuo Imai:** Methodology. **Norihito Tarumoto:** Supervision. **Mieko Tokano:** Methodology, Writing – original draft.

Declaration of Competing Interest

The authors have no conflicts of interest in association with the present study.

Acknowledgments

None.

Informed consent statement

Informed consent was obtained from the patient for the publication of this case report and accompanying images.

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