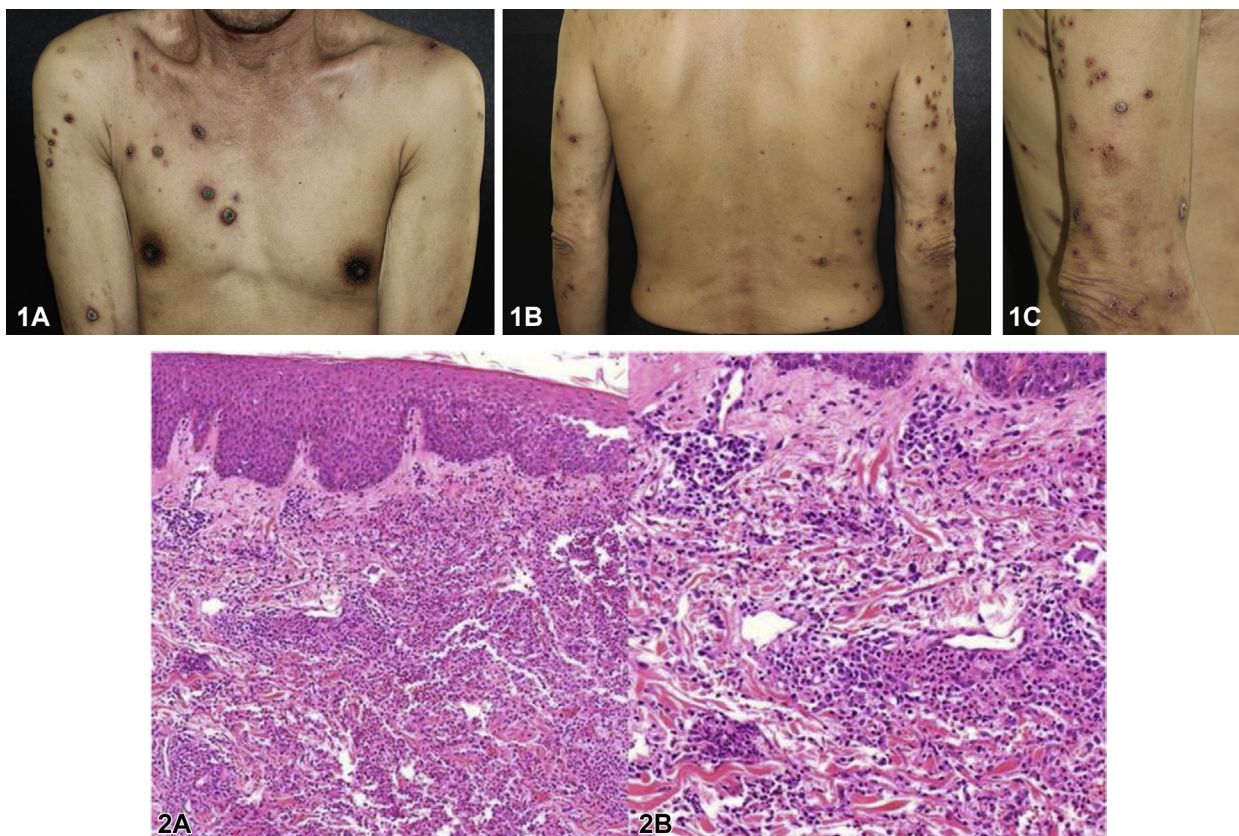


Ulcerative nodules on the entire body



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A 58-year-old man with diabetes mellitus presented with a 1-month history of nodules on his entire body. He had loss of appetite, myalgia, and fever soon followed by the onset of nodules that progressed to ulcers. Physical examination found nonpruritic, painless, multiple, variable-sized, erythematous, ulcerative nodules with central crusts on his face, trunk, and extremities (Fig 1). A biopsy specimen of the lesion was obtained (Fig 2), which showed an acanthotic epidermis and intense lichenoid lymphohistiocytic infiltrate rich in plasma cells in the dermis and extended around vessels.

Question 1. What is the most likely diagnosis?

- A. Kyrle disease
- B. Pyoderma gangrenosum
- C. Malignant syphilis
- D. Febrile ulceronecrotic Mucha-Habermann disease
- E. Disseminated herpes zoster

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Answers:

A. Kyrle disease — Incorrect. Kyrle disease is characterized by pruritic, hyperkeratotic follicular papules with a centrally cone-shaped plug. It usually involves the extensor surface of the extremities. Histopathologic findings show large parakeratotic plugs penetrating into epidermis and dermis. The plugs cause inflammatory response and foreign body giant cell reaction.¹

B. Pyoderma gangrenosum — Incorrect. Pyoderma gangrenosum is common in patients with inflammatory bowel disease. Pathergy is its most significant triggering factor. Clinical findings include a painful erythematous nodule that rapidly breaks down to form a well-demarcated ulcer with undermined edges. In histopathologic findings, nonspecific ulceration and abscess formation with dense neutrophilic infiltrates are present in the deep dermis and subcutaneous tissue.

C. Malignant syphilis — Correct. Malignant syphilis is characterized by prodrome of fever or myalgia followed by papulopustular eruption, which rapidly transforms into crusted necrotic ulcers. Histopathologic findings show dermal plasma cell and lymphocytic infiltrate. High titer positivity of serologic test for syphilis is also important in diagnosing malignant syphilis.^{2,3}

D. Febrile ulceronecrotic Mucha-Habermann disease — Incorrect. This disease is characterized by a sudden onset of ulceronecrotic skin lesions associated with systemic symptoms such as high fever and lymphocytic myocarditis. Histopathology shows acanthosis, exocytosis of lymphocytes with vacuolar degeneration of basal layer, and papillary dermal edema with perivascular lymphohistiocytic infiltrates.⁴

E. Disseminated herpes zoster — Incorrect. Disseminated herpes zoster infection can be diagnosed when 20 or more vesicles develop systemically within a week. Most cases are found in the immunosuppressed conditions such as hematologic malignancies. Histopathology findings show acantholysis with multinucleated keratinocytes.

Question 2. What is the most likely association with this disease?

- A.** Chronic renal failure
- B.** Inflammatory bowel disease
- C.** Oral ulcer
- D.** HIV infection

- E.** Hematologic malignancies

Answers:

A. Chronic renal failure — Incorrect. Chronic renal failure is an associated condition in Kyrle disease.¹ It is rare in malignant syphilis.

B. Inflammatory bowel disease — Incorrect. This disease is usually associated with pyoderma gangrenosum.

C. Oral ulcer — Incorrect. Mucosal involvement is rare in malignant syphilis, whereas it is usually observed in secondary syphilis.

D. HIV infection — Correct. Malignant syphilis is a rare and severe form of secondary syphilis. The risk of malignant syphilis has been associated with HIV and other types of immunosuppressed conditions such as severe malnutrition, chronic alcoholism, and diabetes mellitus.^{2,3}

E. Hematologic malignancies — Incorrect. Hematologic malignancies are more associated with disseminated herpes zoster infection.

Question 3. Which of the following is the most effective treatment of this disease?

- A.** A topical retinoid
- B.** Benzathine penicillin G
- C.** Systemic corticosteroid
- D.** Azithromycin
- E.** Ceftriaxone

Answers:

A. A topical retinoid — Incorrect. Topical retinoids are used to treat Kyrle disease.

B. Benzathine penicillin G — Correct. Benzathine penicillin G is the most optimal treatment of choice for malignant syphilis. Most patients show dramatic response to penicillin.⁵

C. Systemic corticosteroid — Incorrect. Although systemic corticosteroids may have a supportive role in the event of a Jarisch-Herxheimer reaction, the effect of steroids on infection is not confirmed yet.⁵

D. Azithromycin — Incorrect. Azithromycin is less effective than penicillin in malignant syphilis treatment.⁵

E. Ceftriaxone — Incorrect. When a patient is allergic to penicillin, ceftriaxone may be used, but the patient may have an adverse cross-reactivity

with β -lactams. The optimal dose and duration of ceftriaxone therapy have not been defined.⁵

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