Draining dorsal hand pustules, nodules, and ulcers in a patient with immunosuppression

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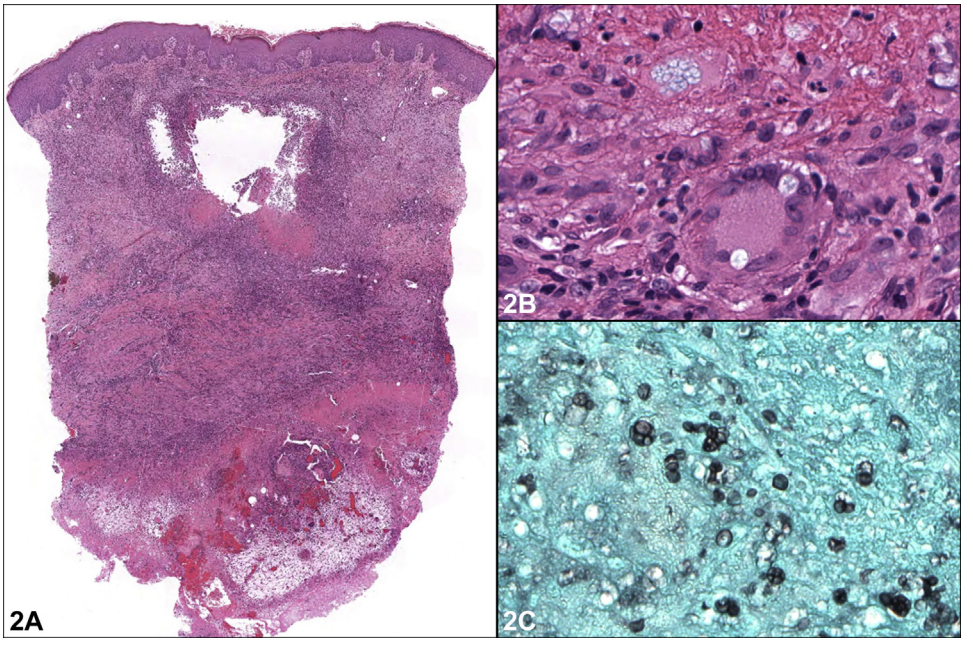

CASE PRESENTATION

A man in his 70s with type 2 diabetes mellitus and chronic obstructive pulmonary disease taking 10 mg prednisone daily presented with 2 months of right dorsal hand pustules with purulent drainage (Fig 1) after peripheral intravenous infiltration. Wound cultures grew multiple bacterial organisms, leading to antibiotic courses without resolution. Over time, the patient developed subcutaneous nodules in a sporotrichoid pattern along the forearm.

Pathology specimens from punch biopsies showed necrotizing granulomatous inflammation. Visible organisms had positive results with Grocott and periodic acid–Schiff (PAS) staining (Fig 2), with no growth on routine tissue, fungal, acid-fast bacilli, or anaerobic cultures. Broad-range fungal polymerase chain reaction testing results were negative.

Question 1: What is the most likely diagnosis?

A. Cutaneous nocardiosis

B. Mycobacterial infection

C. Cellulitis

D. Pyoderma gangrenosum

E. Algal infection

Answers:

A. Cutaneous nocardiosis — Incorrect. *Nocardia* species may cause cutaneous pustules with lymphocutaneous spread, but they was not isolated on culture and appear more filamentous in tissue.

B. Mycobacterial infection — Incorrect. Atypical mycobacterial infections may present with pustules and lymphocutaneous spread. This patient had negative acid-fast bacilli culture and histopathologic staining results.

C. Cellulitis — Incorrect. Although wound cultures grew bacterial species, the histopathologic findings are inconsistent with cellulitis.

D. Pyoderma gangrenosum — Incorrect. Pyoderma gangrenosum does not exhibit sporotrichoid spread, has a more rapid course and acutely overhanging or inflammatory border, and would not show organisms histopathologically.

E. Algal infection — Correct. The diagnosis was protothecosis, a rare algal infection.[1-3] The causative agent is an achlorophyllous alga ubiquitous in the environment that belongs to the *Prototheca* genus.[3,4] Cutaneous protothecosis is the most common clinical presentation and occurs at sites of penetrating trauma, often localized to the extremities.[1] Most patients who develop protothecosis are immunosuppressed. Chronic steroid use, chronic obstructive pulmonary disease, diabetes mellitus, malignancy, and iatrogenic immunosuppression related to history of transplantation are documented risk factors.[1,2] Cutaneous disease often presents with purulent ulcers and erosions.

Question 2: What histologic features are associated with this diagnosis?

A. Sporangia with a cartwheel-like appearance on histopathology

B. Budding yeast forms within histiocytes

C. Pseudoepitheliomatous hyperplasia and spherules containing endospores

D. Large round fungal forms with surrounding narrow-based buds

E. Yeast forms with thick gelatinous capsules

Answers:

A. Sporangia with a cartwheel-like appearance on histopathology — Correct. Histopathology associated with protothecosis often shows a mixed inflammatory infiltrate and necrotizing granulomatous inflammation.[3,4] The hallmark finding is that of morula-like structures that have a soccer ball— or cartwheel-like appearance. These structures

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represent the *Prototheca* sporangia and have positive results with GMS and PAS staining.[1,3,4] Other findings may include focal parakeratosis, hyperkeratosis, hyperplastic lymphoid tissue, and pseudoepithelialization. The organism grows easily within days on a variety of media but may be difficult to isolate if overgrown by contaminants.[4]

B. Budding yeast forms within histiocytes — Incorrect. This answer describes histoplasmosis. Most organisms are seen as intracellular yeast forms within parasitized macrophages with a rim of clearing.[5]

C. Pseudoepitheliomatous hyperplasia and spherules containing endospores — Incorrect. This answer describes coccidioidomycosis. Although organisms show positive results with Grocott and PAS staining and infection may lead to granuloma formation, spherules were not observed in this case.[5]

D. Large round fungal forms with surrounding narrow-based buds — Incorrect. This answer describes paracoccidioidomycosis. The narrow-based buds from larger forms leads to the mariner's wheel appearance.[5]

E. Yeast forms with thick gelatinous capsules — Incorrect. This answer describes cryptococcosis. Staining with mucicarmine would discriminate cryptococcosis from other infections by highlighting the characteristic capsule.[5]

Question 3: What is a potential treatment for this condition?

A. Topical corticosteroid

B. Azole antifungal or amphotericin

C. Cytotoxic chemotherapy

D. Meropenem

E. Supportive care

Answers:

A. Topical corticosteroid — Incorrect. Immunosuppression including systemic corticosteroids is a risk factor for cutaneous protothecosis. Further immunosuppression would likely exacerbate this condition.

B. Azole antifungal or amphotericin — Correct. Protothecosis can be difficult to treat and may require multiple agents or months of therapy; optimal treatment for cutaneous disease is unclear, although patients with risk for disease spread should be treated more aggressively.[3] Surgical excision or debridement is often beneficial for local cutaneous disease. For deeper or persistent infections, surgical management is combined with azole antifungals or intravenous amphotericin with a tetracycline.[1,3] Left alone, disease will persist and potentially spread, although systemic or disseminated infection is rare. The prognosis for cutaneous infection is generally positive.[1]

C. Cytotoxic chemotherapy — Incorrect. Cutaneous protothecosis is an infection and would not respond to cytotoxic chemotherapy.

D. Meropenem — Incorrect. This patient was treated with multiple antibiotic courses without resolution. Protothecosis infection is not a bacterial infection and would not respond to conventional antibiotics alone.

E. Supportive care — Incorrect. Cutaneous protothecosis requires treatment to reduce the likelihood of dissemination.

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Abbreviation used:

PAS: periodic acid—Schiff

REFERENCES

1. Tseng HC, Chen CB, Ho JC, et al. Clinicopathological features and course of cutaneous protothecosis. *J Eur Acad Dermatol Venereol*. 2018;32:1575-1583.
2. Seok JY, Lee Y, Lee H, Yi SY, Oh HE, Song JS. Human cutaneous protothecosis: report of a case and literature review. *Korean J Pathol*. 2013;47(6):575-578.
3. Hillesheim PB, Bahrami S. Cutaneous protothecosis. *Arch Pathol Lab Med*. 2011;135(7):941-944.
4. Lass-Flörl C, Mayr A. Human protothecosis. *Clin Microbiol Rev*. 2007;20(2):230-242.
5. Bolognia JL, Schaffer JV, Cerroni L. *Dermatology*. 4th ed. Amsterdam, The Netherlands: Elsevier; 2018.