Title
Disseminated cutaneous sporotrichosis presenting as a necrotic facial mass: Case and review

Journal
Dermatology Online Journal, 23(7)

Authors
He, Yong
Ma, Chelsea
Fung, Maxwell
et al.

Publication Date
2017

DOI
10.5070/D3237035747

Volume 23 Number 7 | July 2017
DOJ 23 (7): 10

Disseminated cutaneous sporotrichosis presenting as a necrotic facial mass: Case and review

Yong He MD, Chelsea Ma MD, Maxwell Fung MD, Sarah Fitzmaurice MD

Affiliations: Department of Dermatology, University of California Davis Health System, Sacramento, California, USA

Corresponding Author: Yong He MD, Department of Dermatology, University of California Davis Health System, 3301 C Street, Suite 1400, Sacramento, California 95816, E-mail: yhe@ucdavis.edu

Abstract

Sporotrichosis is a subcutaneous mycotic infection caused by Sporothrix schenckii, a group of common saprophytes of soil, plants, and organic debris. Disseminated forms may be seen in the setting of immunosuppression and are typically treated initially with intravenous lipidized amphotericin B. We report an unusual case of a 65-year-old woman who developed disseminated cutaneous sporotrichosis with extensive facial involvement in the absence of a known primary inoculation. Her cutaneous lesions completely resolved after treatment with intravenous posaconazole without amphotericin B.

Keywords: disseminated sporotrichosis, Sporothrix schenckii, posaconazole

Case Synopsis

A 65-year-old woman with history of untreated chronic lymphocytic leukemia, on chronic hydrocortisone replacement following prolactinoma resection, was admitted to the Veterans Administration (VA) hospital with failure to thrive. A dermatology consultation was requested to evaluate a progressive facial mass that was present upon admission. The patient was a nursing home resident for many years and denied travel or illnesses preceding onset of the mass, which had been present for 6 months with progressive growth and facial disfigurement. She was hospitalized at an outside facility two months earlier for presumed cellulitis associated with the mass and had received empiric treatment with sulfamethoxazole-trimethoprim, clindamycin, ampicillin-sulbactam, acyclovir, and fluconazole without improvement.

Physical examination revealed a large vegetative plaque with overlying necrotic eschar on the upper cutaneous lip, with apparent extension into the left nares and left medial cheek (**Figure 1**A). Further examination of the trunk and extremities revealed scattered crusted papules and plaques on the upper back, left arm, and left leg.

The patient was started on empiric intravenous acyclovir and posaconazole upon arrival to the VA hospital. Skin biopsies and tissue cultures were obtained from the facial plaque and a small verrucous papule on the left arm. Both biopsies showed suppurative granulomatous dermatitis with what was read as abundant parasitized histiocytes containing yeast-like organisms (**Figure 2**). A presumptive diagnosis of histoplasmosis was made. Two weeks later, tissue cultures grew dimorphic fungi with morphology consistent with Sporothrix schenckii. By then, her skin lesions had improved significantly and posaconazole was switched to oral itraconazole 200mg daily, to be continued upon discharge. Four weeks later when she presented to dermatology clinic for follow-up, all lesions had healed including the large facial mass (**Figure 1**B), leaving behind a moderate hypopigmented scar. The patient was advised to continue itraconazole 200mg daily lifelong owing to her immunocompromised state.

Case Discussion

Sporotrichosis is a subcutaneous mycotic infection caused by Sporothrix schenckii, a group of common saprophytes of soil, plants, and organic debris [1]. Infections are most prevalent in tropical regions but have been reported worldwide. In the United States, sporotrichosis has become a recognized

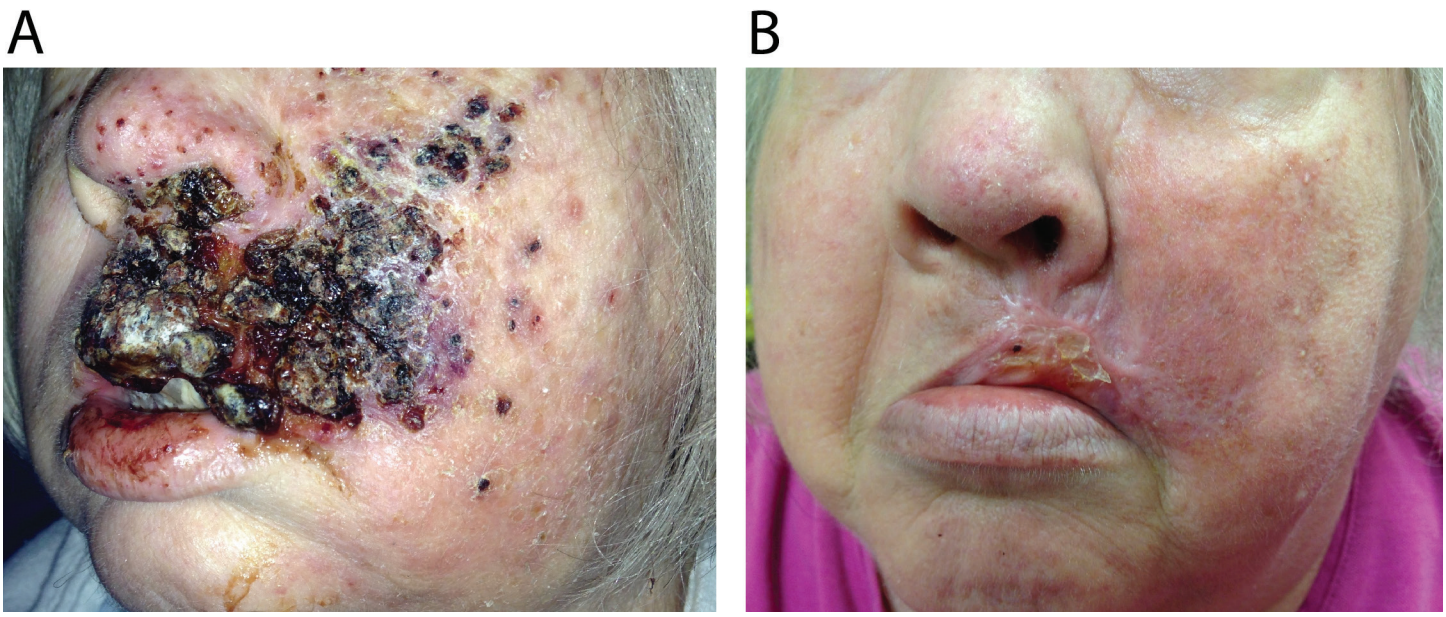

Figure 1. *A) Facial mass with necrotic eschar involving the upper cutaneous lip, left nares, left medial cheek. B) Four weeks later, all lesions have healed, leaving behind a moderate hypopigmented scar.*

opportunistic infection in patients with impaired immunity [2, 3].

Sporotrichosis is usually transmitted by traumatic inoculation and thus commonly presents on the upper extremities of individuals who handle plant material. Although rare in the United States, sporotrichosis can also be transmitted by infected animals via bites, scratches, and contact with secretions. Notably, human-to-human spread has also been reported from caretakers and health care workers handling infected dressings or wounds [4].

Clinically, sporotrichosis has a rather heterogeneous presentation – nodules, plaques, ulcers, molluscum-like lesions, draining sinuses, and subcutaneous masses have all been reported in the literature. This variability may in part relate to the method and location of inoculation, as well as differences in individual immunity. The variability of presentation has led to diagnostic difficulty, as other deep fungal infections, atypical mycobacteria, leishmaniasis, cutaneous malignancies, lymphoproliferative diseases, and inflammatory processes can present with similarly appearing skin lesions.

The case illustrated above was diagnostically challenging owing to the lack of risk factors for inoculation (patient resides in a nursing home and is predominantly indoors), extent of facial involvement, and the pseudo-dermatomal facial distribution (which initially raised suspicion for zoster). In addition, given the patient's age and immunocompromised state, the likelihood of malignancy was greater. Review of the literature showed several rare cases of adult facial sporotrichosis, the majority of which were fixed cutaneous lesions with known history of traumatic implantation or self-inoculation [5-7].

Skin biopsy can be helpful to narrow the differential diagnosis but is rarely diagnostic, as organisms are scant and infrequently detected on histopathology [8]. Histopathologic findings can also be non-specific and mimic other granulomatous disorders. Tissue cultures remain the gold standard and offer the most sensitive and specific method of diagnosis, but incubation periods vary from days to weeks.

Although the lymphocutaneous and fixed forms of sporotrichosis are the most common clinical presentations, extracutaneous and disseminated cutaneous sporotrichosis can occur in the setting of immunosuppression, as in our case. Although our patient did not exhibit extracutaneous disease, dissemination to bone, joint space, liver, and central nervous system have all been reported and can lead to significant morbidity or mortality.

There is a paucity of randomized controlled trials to guide the treatment of sporotrichosis. The Infectious

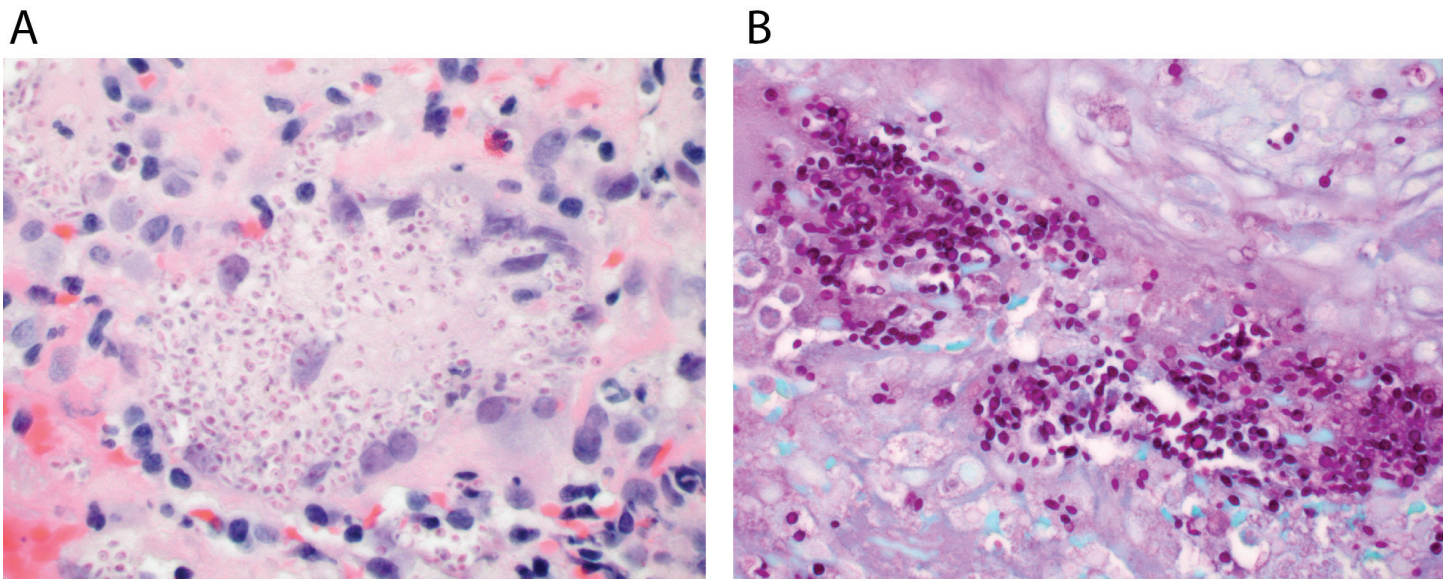

Figure 2. *A) H&E stain from biopsy showing suppurative granulomatous dermatitis and histiocytes containing yeast-like organisms consistent with sporotrichosis, 40x. B) PAS stain from biopsy highlighting budding yeast consistent with sporotrichosis.*

Disease Society of America has published treatment guidelines based on available data, most of which are derived from case reports, uncontrolled trials, and retrospective studies. The preferred treatment in the United States for non-disseminated cutaneous sporotrichosis is oral itraconazole 200 mg daily until complete clinical response is achieved, followed by an additional 2-4 weeks of treatment [9]. For patients intolerant or unresponsive to itraconazole, terbinafine 500mg twice daily has been shown to be effective.

For disseminated or extracutaneous sporotrichosis, intravenous lipidized amphotericin B at 3mg/kg/day is the preferred initial treatment with step-down to oral itraconazole 200mg twice daily, to be continued for at least 12 months after sufficient clinical response is achieved. Immunocompromised patients have difficulty clearing the organisms and should subsequently be on lifelong itraconazole 200mg daily for suppression [9]. Of note, intravenous amphotericin is also the preferred agent in the treatment of pregnant women.

In addition to the above-mentioned agents, sporotrichosis has been treated successfully with voriconazole, fluconazole, and most recently posaconazole. However, in vitro susceptibility studies have demonstrated significantly higher MICs for voriconazole and fluconazole compared to amphotericin B, terbinafine, and other azoles [10]. Treatment with these agents are thus discouraged. Posaconazole is a newer azole with limited clinical data but strong fungistatic activity against Sporothrix brasiliensis in vitro [10]. Bunce et al. reported successful treatment of disseminated sporotrichosis in a patient with hairy cell leukemia with posaconazole in combination with amphotericin B [11].

In our patient, posaconazole was used without amphotericin B because of her underlying renal insufficiency. Our patient responded well to posaconazole alone and was able to be transitioned to itraconazole prior to discharge. To our knowledge, this is the first case in which posaconazole was used to successfully treat disseminated cutaneous sporotrichosis without concurrent treatment with amphotericin B. Although isolated to one case, we speculate posaconazole may be a viable initial agent for disseminated cutaneous sporotrichosis in patients with renal disease.

Conclusion

Sporotrichosis can cause disseminated infections in immunocompromised hosts. Although this infection typically does not have a predilection for the face, our case illustrates the need to consider unusual presentations in the immunocompromised host. Whereas amphotericin B is currently the recommended initial treatment, posaconazole may have a future therapeutic role in the treatment of disseminated sporotrichosis, especially in patients

with renal impairment.

References

1. de Lima Barros M, de Almeida P, Schubach A. Sporothrix schenckii and sporotrichosis. Clin Microbiol Rev. 2011;24(4):633-654. [PMID: 21976602].
2. Gulberg R, Quintanilla A, Levin M, Williams J, Phair J. Sporotrichosis: recurrent cutaneous, articular, and central nervous system infection in a renal transplant recipient. Rev Infect Dis. 1987(9):369-375. [PMID: 3296101].
3. Rotz L, Slater L, Wack M, Boyd A, Scott E, Greenfield R. Disseminated sporotrichosis with meningitis in a patient with AIDS. Infect Dis Clin Pract. 1996(5):566-568.
4. Mahajan V, Sharma N, Sharma R, Gupta M, Garg G, Kanga A. Cutaneous sporotrichosis in Himachal Pradesh, India. Mycoses. 2005;48(1):25-31. [PMID: 15679662].
5. Dellatorre D, Lattanand A, Buckley H, Urbach F. Fixed cutaneous sporotrichosis of the face. J Am Acad Dermatol. 1982;6:97-100. [PMID: 7085959].
6. Yap F. Disseminated cutaneous sporotrichosis in an immunocompetent individual. Int J Infect Dis. 2011;15:727-729. [PMID: 21719337].
7. Pepper M, Rippon J. Sporotrichosis presenting as facial cellulitis. JAMA. 1980;243(22):2327-2328. [PMID: 7373802].
8. Quintella LP, Passos L, Regina S, et al. Histopathology of cutaneous sporotrichosis in Rio de Janeiro: a series of 119 consecutive cases. J Cutan Pathol. 2011;38(1):25-32. [PMID: 20883452].
9. Kauffman C, Bustamante B, Chapman S, Pappas P. Clinical practice guidelies for the management of sporotrichosis: 2007 update by the infectious diseases society of america. Clin Infect Dis. 2007;45:1255-1265. [PMID: 17968818].
10. Borba-Santos L, Rodrigues A, Gagini T, et al. Susceptibility of Sporothrix brasiliensis isolates to amphotericin B, azoles, and terbinafine. Med Mycol. 2015;53:178-188. [PMID: 25394542].
11. Bunce P, Yang L, Chun S, Zhang S, Trinkaus M, Matukas L. Disseminated Sporotrichosis in a patient with hairy cell leukemia treated with amphotericin B and posaconazole. Med Mycol. 2011;50(2):197-201. [PMID: 21612561].