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Rapidly progressing ulcer and a urine drainage bag

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Abstract

Primary cutaneous mucormycosis is an opportunistic fungal infection caused by the order *Mucorales*, most frequently by the *Rhizopus* species. Both systemic factors, such as diabetes mellitus or malignancies and local factors disrupting the skin barrier are implicated in development of this entity. The initial manifestation is a red-to-black papule rapidly progressing to a necrotic and painful ulcer. Diagnosis is obtained by identification of fungal forms in a skin biopsy, typically showing branching and non-septate hyphae. The clinical course is highly variable and depends mostly on the fungal invasion of deep tissues. However, an early diagnosis is essential for implementation of prompt and optimal treatment, based upon antifungal therapy and aggressive surgical debridement.

Keywords: mucormycosis, ulcer, fungal infection

Introduction

Cutaneous mucormycosis is a relatively uncommon infection caused by the fungal order *Mucorales*. It is less known than other presentations of mucormycosis, such as rhinocerebral and pulmonary types. Cutaneous infections have been associated with healthcare-related factors, such as surgical procedures. In this report, we present a case of cutaneous mucormycosis acquired by a urine drainage bag attached to a patient's leg.

Case Synopsis

A 70-year-old man, with a history of rectal cancer undergoing treatment with FOLFOX-cetuximab for 3 months, was admitted to the hospital owing to obstructive atelectasis with metastases. He was given systemic corticosteroids for dyspnea since admission. He had been diagnosed with diabetes mellitus in the past year. During his hospital stay he developed a black papule on the right lower limb (**Figure 1**), which progressed in the next 7 days to an extremely painful, 4cm, necrotic plaque, surrounded by a large purpuric halo (**Figure 2**).

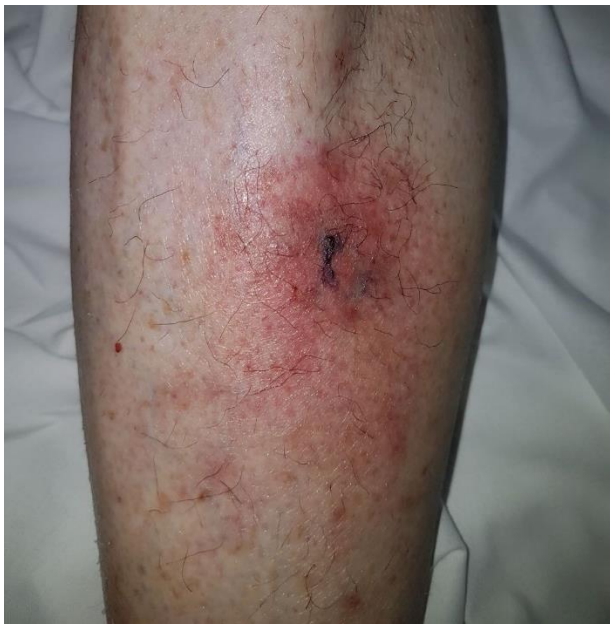

Figure 1. *Initial lesion shows a black papule surrounded by an erythematous halo.*

The history and physical examination did not reveal fever or any other significant signs or symptoms. No portal of entry could be clearly identified. The only plausible link was a urine drainage bag attached to his leg that he had worn close to the primary lesion.

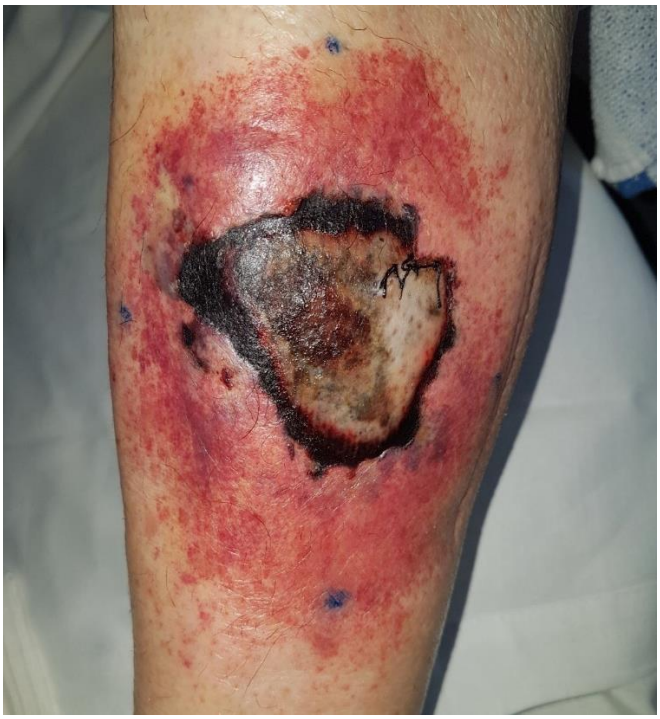

Figure 2. *Progression of initial lesion after 7 days into ulcerated plaque with necrotic and irregular edges, surrounded by a large purpuric halo.*

Laboratory tests did not show any significant findings. An incisional biopsy, including the central portion and periphery of the ulcer, was performed. Histopathological examination showed multiple foci of ischemic necrosis and several branching tubular structures invading the vascular walls and underlying tissue (**Figure 3**) that were consistent with fungal forms. Broad, non-septate hyphae with irregular walls and acute branching were observed and identified as a *Mucor* species. Later, *Rhizopus oryzae* was isolated in culture. A thorough workup ruled out systemic dissemination, establishing the diagnosis of primary cutaneous mucormycosis.

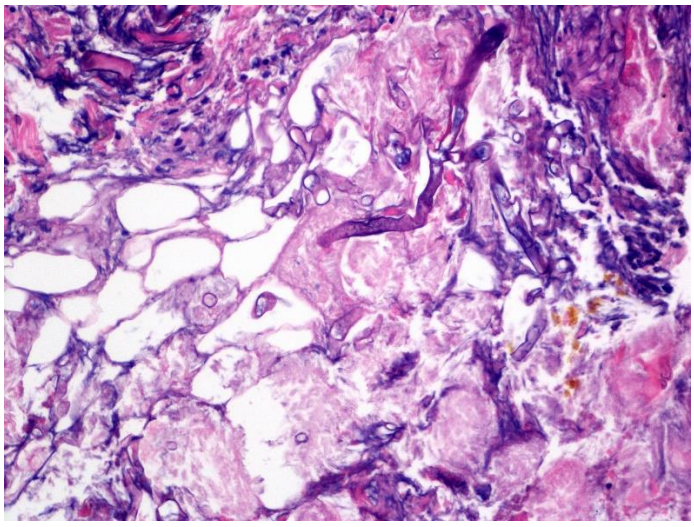

Figure 3. *Histopathological examination shows foci of ischemic necrosis. Presence of broad, non-septate, branching hyphae invading vascular walls. H&E, 40×.*

Liposomal amphotericin B was administered intravenously and surgical debridement with subsequent skin grafting was performed; the patient recovered satisfactorily.

Case Discussion

Cutaneous mucormycosis is an infection caused by the fungi of the order *Mucorales*. It is the third most common form of mucormycosis, after rhinocerebral and pulmonary manifestations, accounting for 19% of all cases [1]. Secondary cutaneous mucormycosis, produced by dissemination from other organs (mainly the rhinocerebral type) to the skin is rare. Instead, most cases of mucormycosis affecting the skin are acquired by direct inoculation and are thus called primary cutaneous mucormycosis [2, 3]. Penetrating trauma, skin abrasions, burns, and motor vehicle accidents are frequent ways of inoculation since these fungi are ubiquitous and usually isolated from soil [1, 3]. However, mucormycosis is also a nosocomial infection and the incidence has increased in recent years [4]. In a review of healthcare-associated mucormycosis [4], the skin was the primary localization of infection in 57% of the cases, in comparison with 19% of overall cutaneous mucormycosis described previously [1]. In these cases, two factors are usually present, major systemic risk factors and local risk factors. Diabetes mellitus, malignancy (especially hematological types), solid organ transplants [1-3], prolonged steroid therapy or other immunosuppressive regimens [4], malnutrition, AIDS, or prematurity [3] are common underlying risk factors associated with this disease. Local factors causing disruption of the skin barrier, most commonly a previous surgical procedure [1, 4], can also be associated with the development of this disease, but a variety of other procedures are implicated, mostly in isolated cases or outbreaks. Contaminated adhesive tapes and bandages, ostomy bags, wooden tongue depressors,

and subcutaneous or intramuscular injections have all been found to predispose individuals to the development of cutaneous mucormycosis [1-4].

Despite the aforementioned risk factors, there are cases in which no underlying disease or portal of entry is found [1]. In our case, the patient had clear predisposing risk factors such as diabetes mellitus, malignancy, and immunosuppressive therapy, but no typical local trauma. Therefore, the urine bag was the only plausible cause of a subtle skin barrier disruption, leading to fungal inoculation.

The clinical presentation of this disease includes a broad spectrum of lesions. It usually starts with a reddish-to-purple papule or plaque, more frequently located on the limbs, that rapidly progresses to a necrotic ulcer with black discoloration surrounded by an erythematous or purpuric halo. Subsequent fungal invasion of cutaneous vessels can eventually involve deeper tissues such as fascia, muscles, or bones, resulting in disseminated mucormycosis [1-4].

The diagnosis is obtained by identification of the microorganism in a tissue biopsy, typically showing areas of inflammation and necrosis with the presence of hyphae with irregular branching at acute angles, usually close to 90º. Unlike *Aspergillus*, these hyphae are broader and have irregular and non-parallel walls [1, 2]. Cultures are positive in 50-89 % and *Rhizopus* is the most frequently isolated species in cutaneous mucormycosis [1, 4].

The clinical course is highly variable and depends mostly on the fungal invasion of deep tissues, but overall mortality is much lower than other forms of mucormycosis [2]. The three cornerstones of treatment are management of underlying conditions, extensive surgical debridement, and antifungal therapy. First-line treatment is amphotericin B, usually a liposomal formulation because of its safety profile, followed by posaconazole, and other azole derivatives as second-line therapies [1,2].

Conclusion

Cutaneous mucormycosis is an uncommon entity with increasing prevalence, especially healthcare-acquired types. A high index of suspicion based on both local and underlying risk factors is necessary in order to establish an early diagnosis and optimal treatment.

References

1. Skiada A, Rigopoulos D, Larios G, Petrikkos G, Katsambas A. Global epidemiology of cutaneous zygomycosis. *Clin Dermatol.* 2012;30(6):628-32. [PMID: 23068150].
2. Castrejón-Pérez AD, Welsh EC, Miranda I, Ocampo-Candiani J, Welsh O. Cutaneous mucormycosis. *An Bras Dermatol.* 2017;92(3):304-11. [PMID: 29186239].
3. Bonifaz A, Vázquez-González D, Tirado-Sánchez A, Ponce-Olivera RM. Cutaneous zygomycosis. *Clin Dermatol.* 2012;30(4):413-19. [PMID: 22682190].
4. Rammaert B, Lanternier F, Zahar JR, et al. Healthcare-associated mucormycosis. *Clin Infect Dis.* 2012;54 Suppl 1:S44-54. [PMID: 22247444].