An Uncommon Case of Mucormycosis

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Introduction

• Mucormycosis is caused by a family of ubiquitous saprophytic fungi known as mucoraceae. *Rhizopus* spp is by far the most common pathogen. These are angiotrophic pathogens that will grow within and eventually occlude blood vessels ultimately resulting in necrosis of affected tissue.1

• Mucoraceae are opportunistic pathogens that typically infect patients with diabetes mellitus, but it can also infect other immunocompromised hosts, such as transplant or burn patients. Due to an increase in incidence, mucormycosis has been identified as an emergent disease, likely due to a rise in susceptible hosts.2

• There are five distinct clinical presentations of mucormycosis; rhinocerebral, cutaneous, pulmonary, gastrointestinal, and cardiac mucormycosis.

• Rhinocerebral mucormycosis may present initially with banal symptoms such as sinusitis, which progresses to necrotic discharge or even visibly necrotic mucous membranes. Symptoms of the infection will be specific to the structures and extent of involvement. Bilateral involvement is uncommon.1

• Cutaneous Mucormycosis can be seen as a primary infection or represent secondary spread from another site. Primary cutaneous mucormycosis spreads via direct inoculation either by trauma such as an IV site or via a contaminated dressing of an open wound. Cutaneous mucormycosis can present as papules, plaques, or nodules. These lesions will characteristically progress to necrotic lesions. Other presentations can include hemorhagic bullae or non-healing ulcers.3

• Clinical suspicion and early diagnosis is essential as this is a rapidly progressing infection with a high mortality rate. Treatment is emergent surgical debridement of all necrotic tissue; performed in an attempt to limit the spread of the infection. Liposomal amphotericin B is the antifungal of choice.1,5 It can be combined with other antifungals, such as posaconazole or caspofungin, for salvage therapy in whom surgical treatment has failed.1,5

Case Description

A 55 year-old female with past medical history of uncontrolled diabetes with a below the knee amputation and chronic kidney disease presented with acute onset necrosis of her forehead and scalp. Ten days prior to admission, she was admitted for intractable headache and periorbital edema. Her symptoms had been present for at least 3 days (13 days prior to presentation total). A work up for angioedema and acute kidney failure was negative and the patient improved on steroid therapy. The patient was discharge on a two week steroid taper.

At the time of admission, patient was awake and alert with a black dry necrotic plaque with edematous edges present over the forehead and extending onto the scalp. A CT of the head was preformed revealing changes consistent with scalp necrosis and mucosal thickening of the ethmoid sinuses. The patient was started on Liposomal amphotericin B for suspected angioinvasive fungal infection. The patient underwent surgical debridement twice, revealing active budding yeast in the temporal region during the second surgery.

Surgical debridement progressed to involve the entire scalp with continued extension into orbital tissue. Surgery was halted due to the aggressive extension into the face, likely requiring enucleation, as well as extension into cranial vessels. The H&E staining of the tissue revealed extensive necrosis with non-septate broad branching hyphae within dermal blood vessels consistent with Mucormycosis. Fungal cultures did not reveal the causative organism. The patient died in hospice three days after admission.

Discussion

• This case also represents an unusual presentation of Mucormycosis due to the subacute timeframe for the initial presentation of her symptoms and for the stepwise fashion which the infection progressed.

• Her initial symptoms, headache and periorbital edema, had been present for 13 days prior to the onset of scalp necrosis. This likely represented the first cutaneous indication of the fungus spreading outside the sinuses.

• These symptoms persisted for nearly two weeks before the supratrochlear and supraorbital arteries were compromised resulting acute onset necrosis.

• This case highlights progression of Mucormycosis from non-descript skin and clinical findings to a more characteristic presentation of the disease.

Conclusions

• Mucormycosis is an opportunistic infection caused by *Rhizopus, Mucor, and Absidia.*

• Rhinocerebral and cutaneous mucormycosis result in necrotic or ulcerated lesions

• Treatment of choice is urgent surgical debridement and Liposomal Amphotericin B

• Rhinocerebral Mucormycosis can present with bilateral periorbital edema

References


