

## What Is Your Diagnosis?



A 56-year-old woman presented with painful, erythematous to violaceous patches with necrosis of the left eye and periorbital area of 1 day's duration. She reported headaches and periorbital pain in the 3 weeks prior to presentation. She was being treated for hypertension, type 2 diabetes mellitus, and end-stage renal disease. The patient denied prior trauma to the area.

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## The Diagnosis: Rhino-orbital-cerebral Mucormycosis

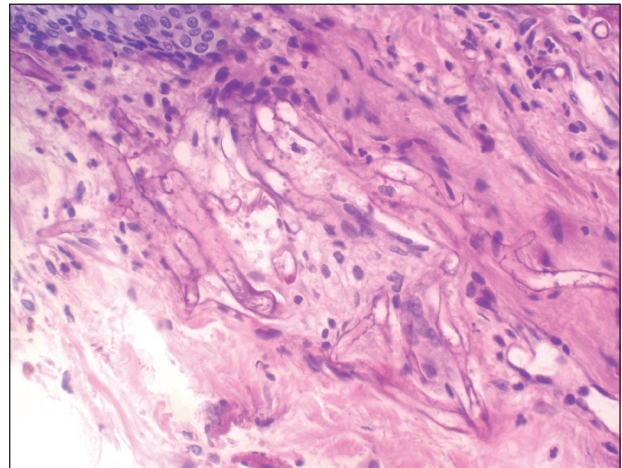
Cutaneous examination revealed a dusky, erythematous to violaceous patch with a necrotic center involving the left eye and periorbital area (Figure 1). The differential diagnosis included herpes zoster, cellulitis, and fungal infection. We obtained patient consent for a punch biopsy. Histopathologic examination revealed irregularly shaped, broad, nonseptate hyphae with right-angle branching (Figure 2). Magnetic resonance imaging of the orbit and head showed involvement of the periorbital soft tissues; the ethmoidal, sphenoidal, and maxillary sinuses; and the left medial temporal lobe. The patient was started on an empirical antifungal treatment of amphotericin B deoxycholate 50 mg daily but died 4 days later due to multiorgan failure.

Mucormycosis is a rare but fatal infection that may rapidly progress.<sup>1</sup> Risk factors include defects in host defense such as malignancy, immunodeficiency from bone marrow or solid organ transplantation, diabetes mellitus, malnutrition, abnormal metabolic states, and deferoxamine use.<sup>1,2</sup> Rhino-orbital-cerebral mucormycosis usually starts with eye or facial pain and unilateral facial swelling.<sup>3,4</sup> Visual impairment, fever, and mental status changes may follow.<sup>1,3,4</sup> Skin findings may progress from erythema to violaceous color changes and lastly to a black necrotic eschar resulting from tissue infarction.<sup>5</sup>

Radiologic imaging may be helpful but rarely is diagnostic in mucormycosis, and reliable serologic tests are lacking.<sup>1</sup> Therefore, suspicion of mucormycosis based on clinical and histopathologic factors followed by immediate initiation of empirical antifungal treatment is critical. The key factors in treating mucormycosis include early diagnosis, correction of underlying risk factors, prompt antifungal therapy, and surgical debridement.<sup>1</sup> Amphotericin B deoxycholate and its lipid derivatives (eg, amphotericin B lipid complex, liposomal amphotericin B) are the standard antifungal agents used in the treatment of mucormycosis.<sup>6,7</sup> Posaconazole is an extended-spectrum triazole with in vitro activity against *Mucorales*. Posaconazole may be useful as salvage therapy; however, strong clinical evidence to support its role as a primary therapeutic agent is lacking in the literature.<sup>6,7</sup> Blood vessel thrombosis and tissue necrosis can result in poor penetration of antifungal agents to the infection site; therefore, surgical debridement also may be critical for complete eradication of the disease.<sup>6</sup> Confirmative diagnosis of mucormycosis can be made based on histopathologic findings.



**Figure 1.** A dusky, erythematous to violaceous patch with a necrotic center on the left eye and periorbital area.



**Figure 2.** Histopathologic examination revealed irregularly shaped, broad, nonseptate hyphae with right-angle branching (periodic acid-Schiff, original magnification  $\times 400$ ).

Our case highlights the importance of clinician awareness of the typical presentation of rhino-orbital-cerebral mucormycosis to ensure prompt diagnosis and initiation of immediate treatment of this possibly fatal infection.

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