



## CASE REPORT

### PRIMARY CUTANEOUS MUCORMYCOSIS IN AN IMMUNOCOMPETENT PATIENT: A CASE REPORT

Deepak Paudel<sup>1</sup>, Raunak Dahal<sup>2\*</sup>, Dhan Kesar Khadka<sup>3</sup>, Yamuna Agrawal<sup>4</sup>

<sup>1</sup>Department of Otorhinolaryngology, BPKIHS, Dharan, Nepal

<sup>2</sup>Department of Otorhinolaryngology, Koshi Zonal Hospital, Biratnagar, Nepal

<sup>3</sup>Department of Dermatology and Venereology, BPKIHS, Dharan, Nepal

<sup>4</sup>Department of Pathology, BPKIHS, Dharan, Nepal

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*\*Correspondence to: Raunak Dahal, Department of Otorhinolaryngology, Koshi Zonal Hospital, Biratnagar, Nepal.*

*Email: [raunakdahal11@gmail.com](mailto:raunakdahal11@gmail.com)*

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#### ABSTRACT

Mucormycosis is an infrequently encountered locally invasive, aggressive fungal infection that frequently occurs in patients with an underlying immunodeficiency. It is usually diagnosed on histopathology and treated with systemic antifungals. We report a case of 16 years old female who presented with swelling on the dorsum of nose with overlying redness and tenderness. As she did not respond to antibiotics and conventional treatment, computed tomography scan and fine needle aspiration cytology were performed which revealed an inflammatory lesion. However, her histopathological examination was a surprise for all of us as it came out to be mucormycosis. She was then started on oral antifungal, and responded well. Although a rare entity, primary cutaneous mucormycosis should always be kept in mind while evaluating any swelling if it doesn't improve on conventional treatments and dealt accordingly.

#### INTRODUCTION

Mucormycosis is a locally invasive and aggressive infection due to zygomycetes that usually occurs in patients with an underlying immunodeficiency such as diabetes or neutropenia or in transplant patients.<sup>1</sup>

Five major forms of invasive mucormycosis have been described based on organ(s) involvement. This includes rhino-orbito-cerebral, cutaneous, pulmonary, gastrointestinal, and disseminated, although rarer forms of mucormycosis, osteomyelitis and endocarditis, have been described.<sup>2</sup> Primary cutaneous mucormycosis is associated with traumatic inoculation of the skin with zygomycetes while the rarer form is secondary involvement in disseminated mucormycosis.<sup>3</sup> The erythema and induration of mucormycosis are non-specific and hence indistinguishable from those of other cutaneous infection. If necrotic eschars are present then immediate biopsy of the lesion should be done. Especially in patients with low neutrophil count, zygomycetes tend to extend rapidly along tissue planes, and this phenomenon is associated with poor outcome.<sup>2</sup> Biopsy of the lesions is the mainstay of diagnosis and blood cultures have low yield.<sup>4</sup> In typical cases, the infection progresses rapidly, and death is highly probable

unless the underlying risk factors are adequately and promptly reversed, all necrotic areas are removed with wide surgical resection and amphotericin-based antifungal therapy is administered.<sup>5</sup> We present a primary cutaneous mucormycosis in an immunocompetent patient and successfully managed medically.

#### CASE REPORT

A 16 years old female presented to otorhinolaryngology outpatient department (OPD) of BPKIHS with chief complaint of bilateral nasal obstruction and bilateral nasal discharge for 3 months. She had no other co-morbid condition. On examination, she had bilateral inferior turbinate hypertrophy and structures posterior to it were not visualized.

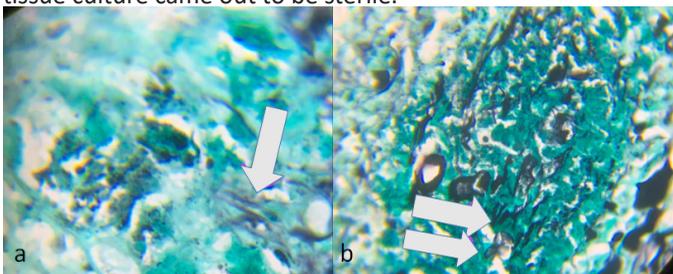
We diagnosed her as a case of bilateral inferior turbinate hypertrophy and started treatment with topical intranasal corticosteroids followed by oral corticosteroid. Turbinate reduction was performed as she did not respond to medical treatment. She was discharged on topical steroid and nasal douching and her symptoms had improved initially. However, 2 months later she again presented to the OPD with complaint of swelling over the dorsum of nose and adjacent mid face.

On examination, the nasal cavity looked normal and there was hard, red and tender swelling over the dorsum of nose and adjacent mid-face (Figure 1).



**Figure 1: Diffuse erythematous plaque on dorsum of nose**

We prescribed her a course of oral antibiotic suspecting a superficial skin infection but even after 1 week of treatment the swelling further increased in size. Then we performed fine needle aspiration cytology (FNAC) which revealed an inflammatory lesion and computed tomography (CT) scan of nose and paranasal sinuses showed cellulitis at lower part of the nose predominantly on the left side extending inferiorly towards the nasolabial folds bilaterally and upper lip. As diagnosis was not clear punch biopsy from the lesion was taken, and it showed dense inflammatory infiltrate along with multinucleated giant cells. Areas of infarction, thrombosed vessels and fibrinoid necrosis were also present. Periodic acid schiff and silver methamine stain highlighted hyphae (Figure 2) and hence diagnosis of mucormycosis was made. However, tissue culture came out to be sterile.



**Figure 2: Blackish hyphae visible in Silver Methenamine stain**



**Figure 3: Complete resolution 3 months after treatment**

The patient was treated with oral itraconazole 100 mg twice daily as it was easily available drug in our institute. She had complete resolution of symptoms after 3 months of therapy (Figure 3) and itraconazole was continued for total 6 months. In her last follow up, one year after the completion of treatment, she was still disease free.

## DISCUSSION

Mucormycosis is a rarely encountered fungal infection caused by fungi of phylum zygomycota. It occurs mainly in patients with uncontrolled diabetes, haematological malignancies and other immunosuppressed conditions.<sup>4</sup> Its clinical types are rhinocerebral (most common), cutaneous, pulmonary, gastrointestinal, disseminated, and miscellaneous.<sup>1</sup>

Cutaneous mucormycosis has two forms: primary cutaneous and secondary cutaneous. Primary cutaneous, starts with trauma, is frequent in immunosuppressed patients and has a good prognosis with early diagnosis. Secondary cutaneous mucormycosis, is usually a dissemination to the skin from a rhinocerebral region, occurs usually in uncontrolled diabetics and immunocompromised individuals, and has a poor prognosis.<sup>6</sup> Although our patient doesn't give definite history of trauma, she didn't have any systemic problems hence a diagnosis of primary cutaneous mucormycosis was made. Furthermore, she was an immunocompetent person.

The clinical presentation of cutaneous mucormycosis varies from gradual onset and slow progression to fulminant course leading to gangrene and haematogenous dissemination.<sup>7,8,9</sup> In a report of cutaneous mucormycosis caused by *Mucor hiemalis* in an immunocompetent child, the disease manifested as superficial annular, scaly lesions resembling tinea corporis and was misdiagnosed for 5 years.<sup>7</sup> Rubin et al. described a case of superficial mucormycosis caused by *Rhizopus arrhizus*, which consisted of targetoid plaques with echymotic necrotic centres and was named 'bull's eye infarct of cutaneous mucormycosis' by the authors.<sup>10</sup>

Our case was unique because the patient didn't have initial cutaneous lesion instead, she came with features of nasal obstruction and discharge with inferior turbinate hypertrophy. Later, she developed a woody indurated plaque over the dorsum of nose and adjacent midface suggestive of an inflammatory lesion which didn't respond to antibiotics. This made us suspicious of alternative pathology.

Biopsies from lesions are essential, especially in the primary cutaneous cases. The histopathology shows an inflammatory reaction with polymorphonuclear infiltration, plasma cells, and a few eosinophils. Edema and necrosis are also present. Thick, hyaline, non-septate and bifurcated hyphae, are better highlighted with periodic acid-Schiff and Grocott stains. Thrombosis and infarctions are also seen. Similar findings were also present in our case which led to diagnosis of mucormycosis. Furthermore, hyphae tend to invade the blood vessels walls, owing to their angiotrophic properties, this explains the tendency of this disease to disseminate.<sup>6</sup>

Standard treatment for mucormycosis is surgical

debridement and administration of antifungal agent.<sup>11</sup> First line therapy for mucormycosis is amphotericin B, followed by posaconazole. Itraconazole is less active than either of these drugs, whereas fluconazole, voriconazole, caspofungin and flucytosine have poor or no activity.<sup>5</sup> Our patient was treated with oral itraconazole 100 mg BD for 6 months which led to dramatical improvement.

Early initiation of treatment is a crucial factor for good prognosis. A multidisciplinary approach consisting of otorhinolaryngologists, dentists, neurologist and ophthalmologist is necessary for successful management of mucormycosis. Hence, the general approach is early and

aggressive treatment with all modalities available.<sup>12</sup>

## CONCLUSION

Although a rare entity, primary cutaneous mucormycosis should always be kept in mind while evaluating any swelling if it doesn't improve on conventional antibiotics and supportive management. Hence, each case should be dealt according to individual circumstances.

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