Rhinoentomophthoromycosis: An uncommon but not rare fungal infection of the nose

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Abstract

Entomophthoromycosis is a group of fungal infections caused by fungus belonging to the order Entomophthorales. The two clinical forms of entomophthoromycosis identified are Basidiobolomycosis and Conidiobolomycosis. Rhino-facial conidiobolomycosis mainly affects the mucosa of the nares, nasal passages, nasal sinuses, nasopharynx, mouth and spreads to adjacent tissues causing disfigurement of face. Histological examinations and mycological cultures are the gold standard for confirmation of entomophthoromycosis. In our middle aged female patient, histopathology revealed fungal elements with broad aseptate hyphae surrounded by splendor hœppli material and surrounding tissue showed dense inflammatory infiltrate with numerous eosinophils and foreign body granulomatous reaction. Treatment include both surgical and medical management, which includes systemic antifungals. Patient was initiated on itraconazole to which she responded. A significant reduction in the swelling and restoration of patency of the nostrils was noted on review after one month. Knowledge of rare fungal infections such as entomophthoromycosis is significant in clinical practice.

Keywords: Basidiobolomycosis, conidiobolomycosis, nasal rhinoentomophthoromycosis

INTRODUCTION

Entomophthoromycosis is a group of fungal infections caused by a fungus belonging to the order Entomophthorales.[1] The name Entomophthorales was coined from the Greek word “Entomon” meaning insect implicating their pathogenic nature in insects. Due to polyphyletic characteristics, the two orders, namely Mucorales and Entomophthorales which were classified in the phylum Zygomycota got redistributed under phylum Glomeromyctea with four other subphyla of uncertain placement. As a result, new subphylum of Entomophthoromycotina was developed, which encompasses two genera that cause human infection; Basidiobolus and Conidiobolus. These are saprophytic fungi in the soil and on decomposing plant matter in moist, warm climates, and sometimes, seen as parasite in insects.[2] Rhinofacial conidiobolomycosis affects the subcutaneous tissues of the face, especially the paranasal sinuses as well as the deeper organs.[3] It originates in the nasal sinuses and spreads to the adjacent subcutaneous tissue of the face, causing disfigurement. We report the case of a middle-aged female who presented to us with rhinofacial conidiobolomycosis who was managed successfully with oral antifungals.

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CASE REPORT

This 54-year-old female, who was apparently well till 8 months back, noticed a small pea-sized swelling inside her left nare. The swelling gradually increased over a period of 8 months, obstructing the left nare. This was associated with coryza and nasal obstruction. She also noticed a diffuse swelling over the left side of the midface. There was no history of trauma prior to this. She did not have pain or epistaxis. She also noticed the change in symmetry of the external appearance of the nose due to the diffuse swelling in the left alar region. The patient did not have fever, weight loss, vomiting, or decreased vision. There were no other comorbidities.

Anterior rhinoscopy revealed swelling over the left inferior turbinate [Figure 1]. There was no tenderness of maxillary, frontal, or ethmoid sinuses. Systemic examination was unremarkable. She was further evaluated for the same. Hematological and biochemical parameters were within normal limits. Computed tomography of the nose and paranasal sinuses showed nonenhancing soft-tissue density lesion with polypoidal margins which were projecting into the choana and anteriorly to the left nasal vestibule [Figure 2].

Biopsy was done under local anesthesia. Diagnosis of rhinoentomophthoromycosis was derived on histopathology which revealed fungal elements with broad aseptate hyphae surrounded by Splendor–Hoeppli material [Figure 3]. Surrounding tissue showed dense inflammatory infiltrate with numerous eosinophils and foreign body granulomatous reaction. Fungal hyphae were periodic acid-Schiff’s (PAS) positive.

She was initiated on itraconazole 200 mg twice daily for 6 months,[4] to which she responded. A significant reduction in the swelling and restoration of patency of the nostrils was noted on review after 1 month [Figure 1].

DISCUSSION

Entomophthoromycosis is a term used to describe a group of fungal infections that affect immunocompetent individuals who live in the tropical and subtropical regions.[5] It is most commonly seen in tropical rain forests of Africa, South and Central America, and Southeast Asia. A few cases have been reported from India.[6] The two clinical forms of entomophthoromycosis identified are basidiobolomycosis and conidiobolomycosis.

The name *Entomophthorales* was coined from the Greek word “*Entomon*” meaning insect implicating their pathogenic nature in insects. Formerly, the two orders, namely *Mucorales* and *Entomophthorales*, were classified in the phylum *Zygomycota*. Hibbett *et al.* suggested a comprehensive phylogenetic classification of the kingdom Fungi, and the phylum *Zygomycota* was eliminated as a result of polyphyletic
characteristics. Therefore, the taxa belonging to Zygomycota were distributed among the phylum Glomeromycota and four subphyla of uncertain placement (incertae sedis). Entomophthorales and Mucorales as well as two other orders (Kickxellales and Zoopagales) were raised to the rank of subphyla and renamed as Entomophthoromycotina, Mucoromycotina, Kickxellomycotina, and Zoopagomycotina. Entomophthoromycotina encompasses two genera that cause human infection; Basidiobolus and Conidiobolus.

These are saprophytic fungi in the soil and on decomposing plant matter in moist, warm climates. Infections are thought to develop due to traumatic inoculations such as thorn pick or insect bite. Furthermore, it may occur by inhalation of spores, or their introduction into the nasal cavities by soiled hands. Most cases affect men with agricultural or outdoor occupations. It is rare in Kerala, due to the fact of less agriculturists in Kerala as compared to other states in India. These fungi develop in the dermis causing ulcerative granulomas.

Entomophthoromycosis is a slowly progressive disease which has a predilection for areas with adipose tissue, possibly, because these organisms thrive on fatty substances. Basidiobolomycosis (subcutaneous zygomycosis) is a chronic infection which affects the lateral aspect of the head, neck, trunk, and limbs, more commonly seen in children and adolescents than in adults. The lesions are usually single, large nodular type, which erode to form discharging ulcerative granulomas. It is associated with severe pruritus.

Rhinofacial conidiobolomycosis (rhinofacial zygomycosis) mainly affects the mucosa of the nares, nasal passages, nasal sinuses, nasopharynx, mouth, and spreads to adjacent tissues causing disfigurement of the face. The lesions are single or multiple large nodules causing ulcerative pyogranulomas which cause mechanical obstruction of the airway resulting in dyspnea. Neither of these two forms occurs preferentially in patients with underlying disease or defective immunity.

Histological examinations and mycological cultures are the gold standard for confirmation of entomophthoromycosis. Biopsy of skin lesions is preferred for diagnosis than pus, as the chances of positive identification with potassium hydroxide preparation and culture are better with tissue specimens. Entomophthoromycosis can be easily differentiated from other fungi by their characteristic hyphal morphology. The hyphae are broad, aseptate, or sparsely septate, with right-angle branching. The histological inflammatory reaction shows infiltration with lymphocytes, plasma cells, epithelioid cells, multinucleate giant cells, and histiocytes with an area of central necrosis that is surrounded by eosinophilic infiltration. This phenomenon is called Splendore–Hoepli phenomenon. This finding is quite typical of entomophthoromycosis but not pathognomonic of it, as it may also seen in infections such as sporotrichosis and schistosomiasis. PAS stain is useful to demonstrate the fungal hyphae. Examination under fluorescent microscopy using fluorescent dye (Blankophor) wet mount preparation increases the sensitivity of diagnosis. Definitive diagnosis requires culture, polymerase chain reaction testing, and immunohistochemistry. Entomophthoromycosis produces characteristic colonies on Sabouraud dextrose agar, potato dextrose agar, or corn meal agar.

Treatment for endomophthoromycosis is medical and surgical management. Systemic antifungal therapy with surgical debridement is the major treatment. Several antifungal agents are used for the treatment of endomophthoromycosis such as cotrimoxazole, ketoconazole, itraconazole, amphotericin B, and potassium iodide. Higher doses than usual may be required for effective treatment, and prolonged daily therapy for months, if there is resistance to antifungals. Our patient responded to itraconazole. Disfigurement of the face can be managed surgically with facial reconstructive surgery.

CONCLUSION

Knowledge of rare fungal infections such as entomophthoromycosis is significant in clinical practice. Rhinoentomophthoromycosis mainly affects the mucosa of nasal cavity, nasopharynx, and sinuses even in immunocompetent individuals. Prompt diagnosis of this infection requires a high index of suspicion. Despite the clinical features, the disease requires biopsy for diagnosis. Histological examinations and mycological cultures are the gold standard for confirmation of entomophthoromycosis. Treatment includes both surgical and medical management, which include systemic antifungals.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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