

# Rhinofacial Entamophthoromycosis- Unravelling of a Puzzle

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**Abstract:** A 40-year-old gentleman presented with complaints of gradually progressive left sided nasal obstruction and swelling over the nose. CECT Nose and Paranasal sinuses showed a lobulated soft tissue density filling left nasal cavity extending to subcutaneous tissue on left side. Histopathology revealed chronic inflammatory lesion due to fungal infection which was morphologically consistent with Entomophthoromycosis. The patient was started on antifungal medication and he was symptomatically improved.

**Keywords:** Entamophthoromycosis, Rhinofacial, Nasal Mass, Fungal infection

## 1. Introduction

Rhinofacial entomophthoromycosis is a rare subcutaneous zygomycosis that is caused by the fungus *Conidiobolus coronatus*. It is seen mainly in the tropical and subtropical regions of the world. It primarily affects the upper respiratory mucosa and adjacent subcutaneous tissue. It can infect nasal mucosa and submucosa forming subcutaneous nodules and has the potential to spread to adjacent tissues.<sup>1</sup>

## 2. Case Report

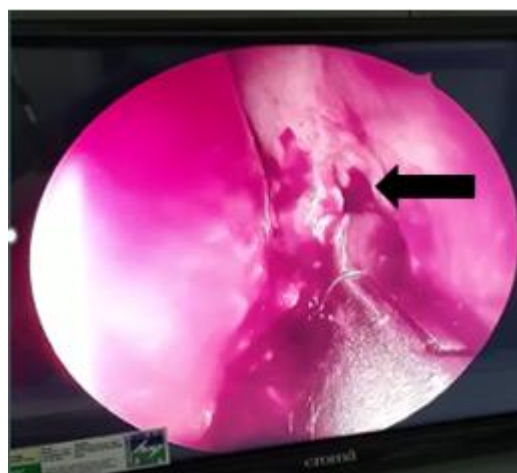
A 40-year-old male who is a diabetic for 15 years and a known case of renal cell carcinoma post nephrectomy, presented with left sided nasal obstruction for 9 months and swelling over nose for 2 months duration. The symptoms were progressively increasing in severity. On examination, there was a diffuse swelling over the dorsum of nose with erythematous and thickened skin overlying it. Also, there was bilateral periorbital oedema and oedema over left premaxillary area (Fig 1).



**Figure 1:** Picture showing diffuse swelling over dorsum of nose and periorbital oedema

Diagnostic nasal endoscopy showed a pinkish smooth mass in the left nasal cavity extending up to just behind the vestibule which was firm, sensitive to touch, non-tender and not bleeding on touch (Fig 2)

His routine blood investigations were within normal limits and peripheral smear showed normocytic normochromic picture with few reactive lymphocytes.



**Figure 2:** Intraoperative nasal endoscopy picture showing the lesion

CECT nose and paranasal sinuses showed lobulated soft tissue density filling the left nasal cavity extending to the subcutaneous tissue on left side and mucosal thickening in the left maxillary, ethmoid and frontal sinuses (Fig 3).



Figure 3: CECT Nose and PNS

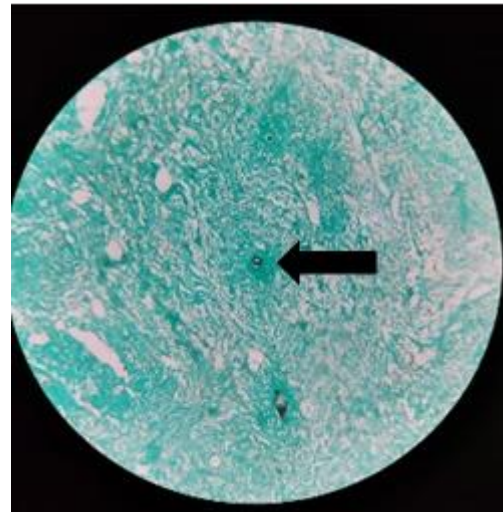


Figure 5: Giemsa stain showing fungal hyphae

Biopsy samples from the swelling were sent for microbiological and histopathological evaluation. Bacterial culture revealed *Klebsiella* infection, favouring the possibility of Rhinoscleroma. But later MALDI-TOF study identified the species as *Klebsiella aerogenes*. Antibiotics were started according to the culture and sensitivity report. Cytological examination showed scattered inflammatory cells only. Fungal Staining & Culture were negative. Histopathological examination was suggestive of only chronic inflammation. His ANA profile and both tissue and sputum CB NAAT for tuberculosis were negative.

The patient was not improving with conservative management using antibiotics, steroids and other anti-inflammatory medications for many weeks. To find out the etiological agent, repeated biopsies and fungal cultures were sent. Finally, we got a histopathology report showing chronic inflammatory lesion due to fungal infection which was morphologically consistent with Entomophthoromycosis. (Fig 4 & 5).

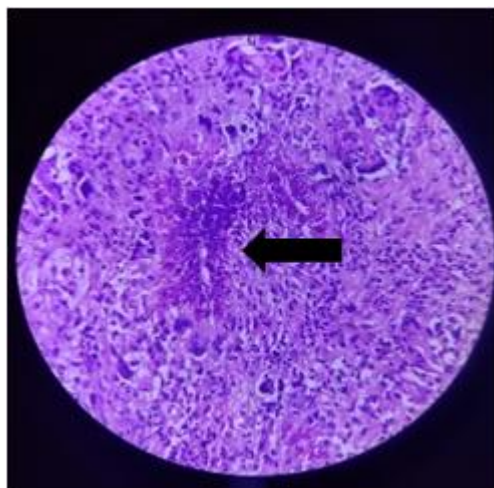


Figure 4: Histopathology showing Splendore-Hoeppli phenomenon

Treatment: Initially he was treated with Inj. Cefoperazone sulbactam and T. Minocycline according to the sensitivity of *Klebsiella aeruginosa* isolated in bacterial culture. Intravenous steroid was also started in view of periorbital and premaxillary oedema. After getting histopathological report of Entomophthoromycosis, he was started on T. Itraconazole 200mg BD and T. Methyl prednisolone in tapering doses. Then the patient was symptomatically improved within a few days (Fig 6). Itraconazole was continued for 3 months.



Figure 6: Patient after 1 month of initiation of antifungal therapy

### 3. Discussion

Entomophthoromycosis is a rare fungal infection that may affect immunocompetent hosts.<sup>2</sup> It is caused by fungi belonging to the order Entomophthorales including conidiobolus and basidiobolus species. Rhinofacial entomophthoromycosis is caused by *Conidiobolus* species while *Basidiobolus* mainly affects the thorax, trunk, limbs, and intestinal tract. *Conidiobolus* infection was first described in humans in 1961.<sup>3</sup>

Rhinofacial entomophthoromycosis is transmitted by inhalation of the fungal spores. It is a slow growing, locally infiltrative disease of the nasal cavity, paranasal sinuses and soft tissues of the face. Host defence can result in chronic granulomatous reaction. Sometimes it may be mistaken for malignancy or tuberculosis. It is seldom life-threatening but may cause severe facial disfigurement. The presenting complaints

will be nasal discharge, nasal obstruction, swelling of nose, upper lip and face. It should be differentiated from the more fulminant infection caused by mucormycosis which spreads rapidly with widespread necrosis and angioinvasion.

Histopathological examination is gold standard for diagnosis. It is characterized by the presence of hyphae surrounded by an eosinophilic halo known as Splendore-Hoeppli phenomenon which is due to antigen-antibody reaction within the tissue. Fungal stain and culture can be done for diagnosis. Findings on KOH staining are short, thick hyphae with absent or few septae. Cultures are performed in Sabouraud dextrose agar medium without cycloheximide or chloramphenicol, at 30° to 37°C. Colonies are fast growing and are white, beige, or brown, glabrous and folded in appearance.<sup>4</sup> The fungus may be rare in tissue sections and when present, is often fragmented. Also, hyphae may appear in only a part of the specimen. So the diagnosis of this disease remains difficult and may initially be missed as in our case.<sup>2</sup> Molecular methods for diagnosis are, Polymerized Chain Reaction, DNA probes, and fungal primers.

The treatment of choice for rhinofacialentomophthoromycosis is surgical debridement followed by antifungal medication. Prolonged antifungal therapy is needed. Itraconazole may be given up to 3 months. Other medical management options are Potassium iodide, Amphotericin B and Imidazoles. Cryotherapy has been tried with limited success. Relapses are common, even after successful treatment.<sup>5</sup>

## References

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