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Case Study

RHINOFACIAL ENTOMOPHTHOROMYCOSIS - A CASE TREATED SUCCESSFULLY WITH ITRACONAZOLE

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ABSTRACT
Rhinofacial Entomophthoromycosis (RFE) is an uncommon mycotic disease caused by Conidiobolus Coronata. We report a case of REF in a 21 year old, female with involvement of rhinofacial area. Diagnosis was made on resected lesion. Stains used were Hematoxylin & Eosin (H&E), Periodic acid-Schiff (PAS) and Methenamine Silver stain.
RFE caused by fungi Entomophthorales and class Zygomycetes. The Entomophthorales has two genera Conidiobolus and Basidiobolus (Ric. 2003). This occurs predominately in immunocompetent patients and presents as a slowly progressive swelling involving the head and face. RFE is occasionally mistaken for malignancy and sometimes as in this case for Tuberculosis (TB).

Key Words: *Rhinofacial Entomophthoromycosis, Entomophthorales, Conidiobolus*

INTRODUCTION
Rhinofacial Entomophthoromycosis (RFE) is an uncommon mycotic disease in humans occurring in the tropical and subtropical parts of the world. RFE comprises of various diseases caused by fungi Entomophthorales and class Zygomycetes. The Entomophthorales has two genera Conidiobolus and Basidiobolus. Conidiobolus produces RFE. This occurs predominantly in the immunocompetent patients and involves the head and face.

Case
A 21 year old female living in Pune (India) presented with a firm slowly growing swelling over right maxilla destroying orbital wall and palate, over a period of one year. No history of trauma or insect bite was present. Physical examination showed a mass over the right maxilla. We received multiple fragments of the resected lesion aggregating to 5x5x1cm. All the tissue was processed and slides were stained with H&E, PAS and methenamine silver stain. Other laboratory findings included Hemoglobin 10.6gm/dl, White cell count 9800/µl, Platelet count 4, 20,000/µl. HIV negative, HBsAg negative and fungal culture was negative.

Radiological Findings
Multislice CT scan of paranasal sinuses showed hyperdense mass seen in the right maxilla – zygomatic region with extension into the retro-orbital zone and infratemporal region. The mass measures 6x4x4.2cm. Erosion of the right maxillary antrum, pterygoid plates, superior orbital fissure and right alveolar margin is seen. Mass is seen involving the left alveolar margin with erosion. Mild proptosis of the right eye is noted. No intracranial extension is noted. Soft tissue mass in right maxilla-zygomatic region with intraorbital extension and erosion of bony structures.

Microscopy
Revealed fibroadipose tissue infiltrated by granulomas composed of epithelioid cells and giant cells. Background infiltrate was present and was composed of eosinophils and lymphocytes. Giant cells showed presence of broad septate hyphae measuring between 2 to 6 micrometer in diameter. These hyphae were highlighted on PAS and Methenamine silver stain. This lesion was diagnosed as RFE. No Splendore hoeppli material noted.

Treatment included

Itraconazole 150mg for 16 weeks.

The patient has been followed up for a period of 8 months and the mass has reduced in size and no recurrence is seen.

DISCUSSION

Entomophthoromycosis is a chronic infectious disease found predominantly in the tropical and subtropical regions. RFE comprises of various diseases caused by fungi Entomophthorales and class Zygomycetes. The Entomophthorales has two genera Conidiobolus and Basidiobolus. Conidiobolus produces RFE. The first case was reported by Bras in 1965 in Jamaica (Tho. 2006). The age range varies from 15 months to 79 years (Leo. 2010). This occurs predominantly in immunocompetent patients. Infection has been suggested to result from percutaneous inoculation of Conidiobolus coronatus via fungal spore inhalation or insect bite. It is a locally progressive infection of the nasal cavity, paranasal sinuses and soft tissues of the face that usually does not extend to the intracranium. It presents with a nasal mass with multiple subcutaneous nodules. These nodules represent granulomas. It can spread from nose and involve the paranasal sinuses, upper lip, forehead and cheeks and causes extensive facial deformity. Rare clinical presentations include nasal polyps and epistaxis. The lesion is occasionally mistaken clinically for malignancy such as squamous cell carcinoma, lymphoma or angiocentric immunoproliferative lesion (Leo. 2010)

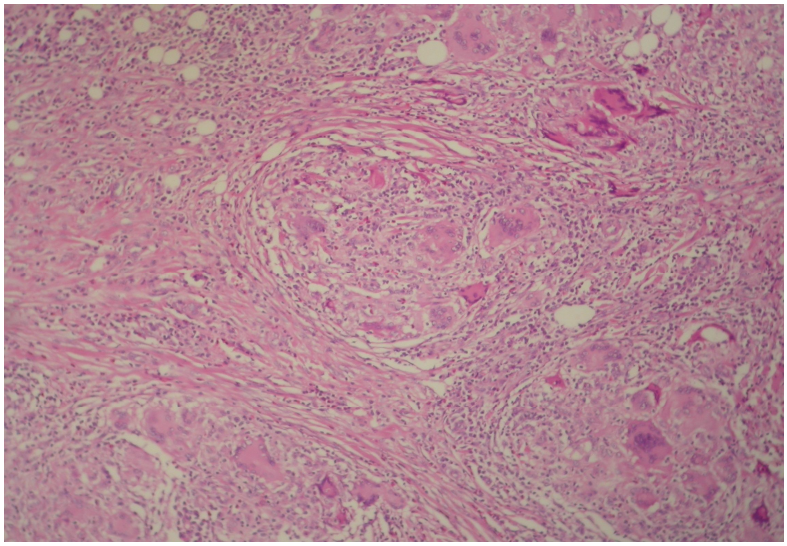

Figure 1: Fibroadipose tissue infiltrated by multiple granulomas composed of epithelioid cells and multinucleated giant cells (H & E X 100)

Histology in the acute phase shows eosinophilic, lymphocytic and plasma cell infiltrate. In the chronic phase granulomatous infiltration with giant cells and histiocytic inflammation is seen. Fungal elements in the tissue lie singly or in clusters. Their diameter varies between 4µ to 10µ. The hyphae are usually septate and thin walled. In sections stained with hematoxylin and eosin the fungi are surrounded by a granular eosinophilic material, described as Splendore-Hoeppli phenomenon and is thought to be immune precipitate around the parasites in tissue. Phagocytosis of fungal hyphae by giant cells is seen. An important point of distinction from mucormycosis is that the hyphae are not seen in the walls or within the lumen of blood vessels and vascular thrombosis is not seen. The differential diagnosis on histology is Kimura disease and

angiolymphoid hyperplasia with eosinophilia [ALHE]. Kimura disease and ALHE lack eosinophilic Splendore – Hoeppli material and have negative results on fungal stains.

The diagnosis is based on histological examination as cultures for the causative organisms are positive in only 15-20% of cases (Ram, 2000).

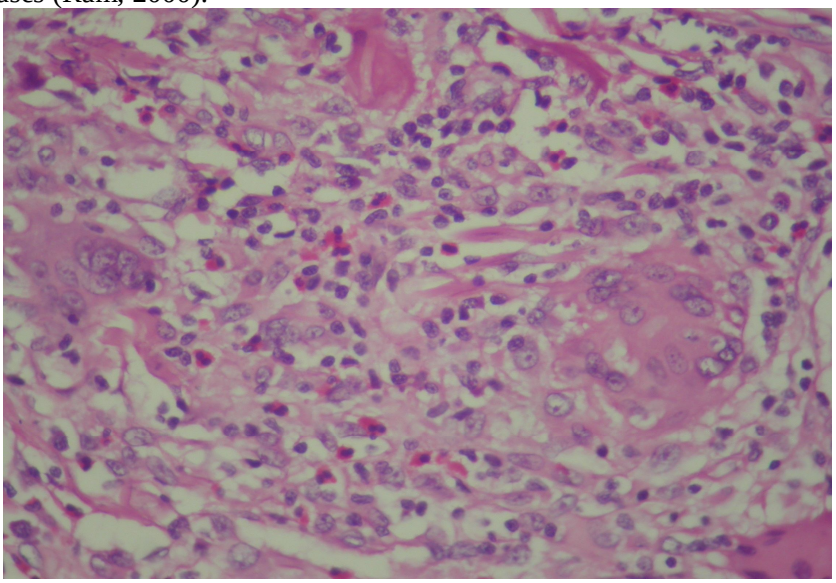

Figure 2: Inflammatory infiltrate composed of granulomas, eosinophils, lymphocytes and giant cells showing broad hyphae. (H & E X 400)

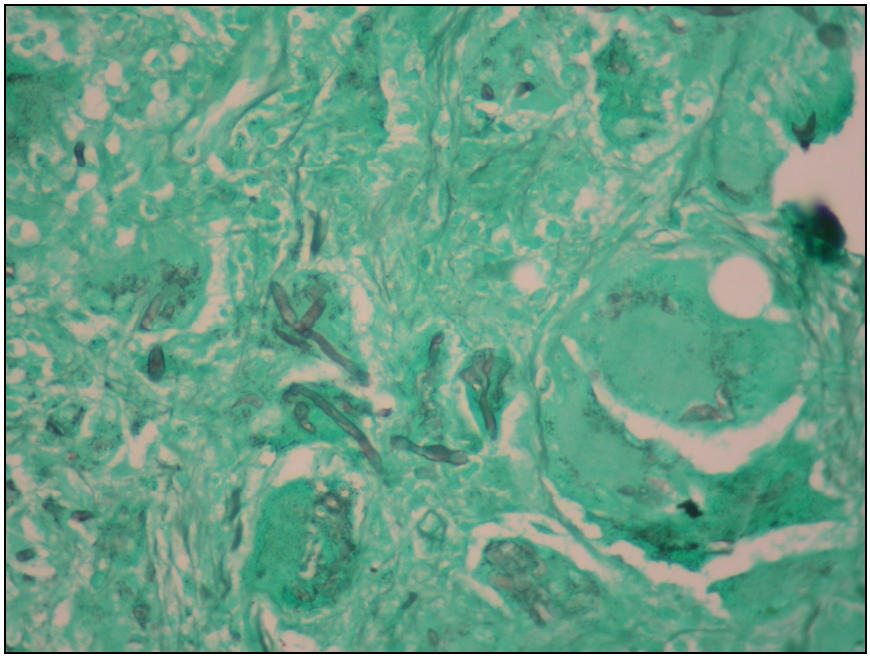

Figure 3: Fungal hyphae highlighted on Gomori Methenamine Silver staining (X 400)

Various drugs are used to treat this disease like itraconazole, fluconazole, cotrimoxazole, amphotericin and potassium iodide (Ram, 2000). These drugs have been used with varying success.

Case Study

This case highlights the need to suspect entomophthoromycosis in granulomatous inflammation of nose and paranasal sinus in non-immunocompromised patients.

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