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Case report/Cas clinique

Non-sinonasal isolated facio-orbital mucormycosis – A case report

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ABSTRACT

Mucormycosis is a rare clinical entity, often affect immunocompromised patients. It is an emergency situation and has poor prognosis. Prompt diagnosis with tissue biopsy, local control of the disease by aggressive surgical debridement and appropriate systemic antifungal treatment improve the prognosis and survival of the patients. Treatment of mucormycosis needs antifungal agents such as Amphotericin B and wide surgical debridement. Early diagnosis and treatment is often needed for survival of the patients. We describe a rare case of mucormycosis affecting facio-orbital area without involving sinonasal cavity.

1. Introduction

Mucormycosis is a rare and dreaded disease caused by a fungus of the order Mucorales. It has high morbidity and mortality [1]. Most commonly Rhizopus species are causative organisms for the mucormycosis. Diabetic ketoacidosis and neutropenia are common predisposing conditions. This is a life-threatening infection which has remarkable affinity for arteries [2]. This fungus often dissects internal elastic lamina from the media of the blood vessels, resulting extensive damage to the endothelium and lead to thrombosis. Mucormycosis is classified into different forms as per anatomic sites like rhino maxillary, central nervous system, cutaneous, pulmonary, disseminated and miscellaneous. The rhino-orbito-cerebral is the most common variety of mucormycosis [3]. The most common sites of mucormycosis infections are paranasal sinuses (39%), lungs (24%), skin (19%), brain (9%), gastrointestinal (7%) forms and other miscellaneous types are extremely rare [4]. The ideal treatment need correction of underlying risk factors, antifungal treatment with Amphotericin B and aggressive surgery. Here we are reporting a case non-sinonasal mucormycosis which only affecting the orbit and facial area.

2. Case report

A 56-year-old lady attended the outpatient department of Otorhinolaryngology with a swelling (3 cm × 3 cm) in the right facio-orbital area (Fig. 1) since 2 months. She was known case of diabetic mellitus and hypertensive patients. She was under treatment with oral hypoglycemic agents like metformin and glipizide but her blood sugar was poorly controlled (HbA1c 10.6%). Diagnostic nasal endoscopy showed normal nasal cavity and nasopharynx. Ophthalmological consultation revealed loss of the vision in the right eye. Computed tomography (CT) scan of the nose and paranasal sinuses revealed normal nasal cavity but a mass involving right orbit and adjacent facial area (Fig. 2). A small piece of biopsy sample was taken from the facio-orbital mass (Fig. 3) which showed the picture of mucormycosis with some foci of non-septate fungal hyphae with right angled hyphal branches (Fig. 4). She had undergone radical excision of the mass along with orbital exenteration followed by parenteral liposomal Amphotericin B (5 mg/kg). Patient follow-up showed no evidence of recurrence after six months of surgery.

3. Discussion

Mucormycosis is caused by saprophytic fungi of many genera related to phycomycetes (zygomycetes) and order Mucorales [5]. The fungus has great affinity towards to arteries and adheres to the arterial wall. It grows along the internal elastic lamina of blood vessels causing thrombosis, ischemia and necrosis of the surrounding tissues. Mucorales are abundantly seen in soil, decaying vegetables, animal excreta and foodstuffs. They grow rapidly in humid environment and the sporangiospores are released and spread as airborne propagules. In India, the air borne spores are more during transition from summer to rainy season as it may be ideal for fungal growth [6]. Mucormycosis is a rare clinical entity and often affect immunocompromised patients.

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Fig. 1. A mass (3 cm × 3 cm) present at the right side facio-orbit region.

The organisms causing Mucormycosis is a fungus, primarily of the genus Rhizopus or Mucor. Mucormycosis may be of rhino-orbital-cerebral and pulmonary infections and these types of the diseases may lead to emergency situation to the patients [7]. In our case, the disease is affecting only orbit and nearby facial area without involving nose and sinuses. These are rapidly growing organisms, characterized by ribbon like hyphae with no or only few septae. The genera affecting human are Cunninghamella, Absidia (Lichtheimia), Mucor, Rhizomucor, Rhizopus and one the basis of geographical distribution, Saksenaea and Apophysomyces [8]. This aggressive and highly destructive fungal infections more often seen in immunocompromised hosts like patients with hematological malignancies or those taking hematopoietic stem cells transplantation. Diabetic mellitus patients with ketoacidosis and transfusion/dyserythropoetic iron overloaded patients are also important risk groups. The most common predisposing factors associated with mucormycosis are uncontrolled diabetes mellitus especially with history of ketoacidosis [9]. Neutropenia, immunosuppressive therapy, acquired immune deficiency syndrome, malnutrition, dialysis, hematological malignancy and organ transplantation are often predisposing factors. Mucormycosis universally affects immunocompromised patients. The potent T-cell depleting agents used for immunosuppression as in organ transplantation are often leads to high risk for causing mucormycosis. Although mucormycosis is ubiquitous and rapidly spread, it seldom affects an immunologically competent patient. Therefore mucormycosis occurs among patients with serious underlying conditions like diabetic mellitus, leukemia, organ transplantations, acquired immunodeficiency syndrome, severe burn and immunosuppressive medications. Around 70 to 80% of mucormycosis patients have diabetes mellitus [10]. Radiological imaging will not establish but suggest the diagnosis of mucormycosis. CT or MRI are helpful for Facial and cerebral involvement of mucormycosis and determine loco-regional extension towards the orbit and brain and also identify the cerebral thrombosis [11]. Cavernous sinus involvement is best assessed by MRI [12]. Difficulties in diagnosis and followed by antifungal treatment with resistance to many commonly prescribed antifungal agents leads to high mortality in certain group of patients [13]. There is no such definitive diagnosis for the mucormycosis except histopathological confirmation by seeing the non-septate hyphae of the affected tissue [3]. Necrosis of the surrounding tissue prevents antifungal drugs like Amphotericin B to enter the site of infection [14]. Early debridement of the affected tissue gives better prognosis for successful treatment in mucormycosis [15]. Liposomal Amphotericin B treatment has better tissue penetration and considered as first line treatment [4]. However, monotherapy like surgery alone or Amphotericin B alone has limited efficacy whereas the combination therapy with

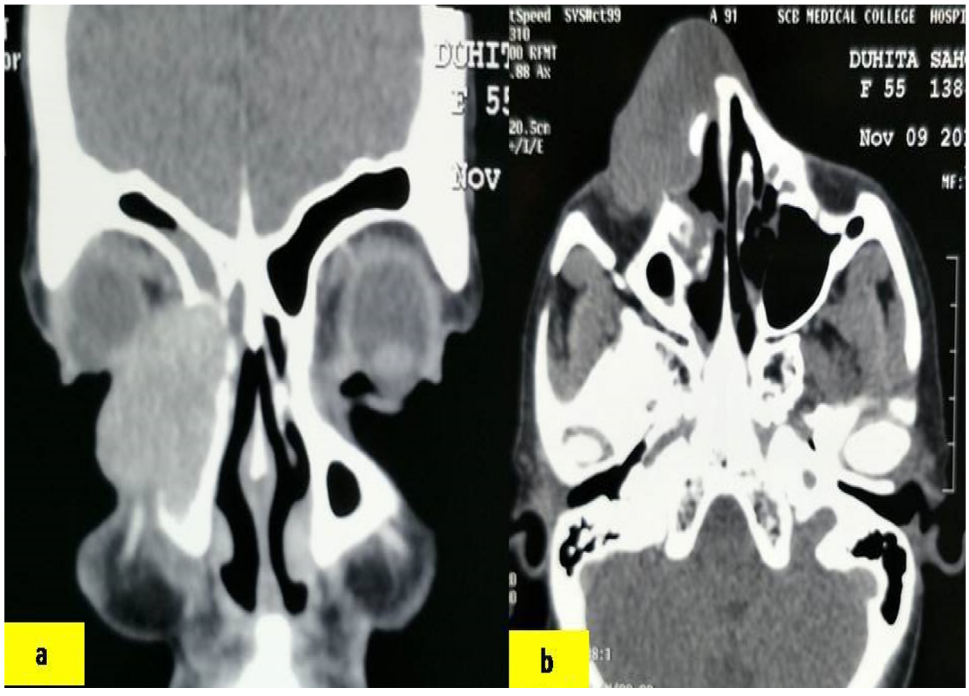

Fig. 2. CT scan (Fig 2a coronal cut and Fig 2b axial cut) picture showing mass at right facio-orbit area without involving nose and paranasal sinuses. a: coronal cut; b: axial cut.