Case Report

Giant orbitofacial basidiobolomycosis: diagnostic and management challenges in a resource limited environment.

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Abstract

Basidiobolomycosis, a rare disease caused by the fungus *Basidiobolusranarum*, an environmental saprophyte, member of the class *Zygomycetes*; order *Entomophthorales*, found worldwide is an opportunistic infection that can be life-threatening in immune compromised patients. This report presents an unusual case of a giant orbito-facial basidiobolomycosis in a 30-year-old, otherwise healthy Nigerian farmer. The diagnostic and treatment challenges peculiar in our environment were also discussed.

Key words: orbitofacial lesions, fungal infections, basidiobolomycosis, Splendore-Hoeppli phenomenon.

Basidiobolomycosis is a rare fungal disease caused by the saprophytic fungus *Basidiobolusranarum*, an environmental saprophyte that is usually transmitted by inoculation and largely restricted to tropical areas of Africa, Asia, and South America[1]. Except for their opportunistic character, little is known about its pathogenesis; hence the use of broad-spectrum and highly nephrotoxic antifungal agents like amphotericin B[2]. Also, it is characterized by the formation of firm and non-tender swellings, generally on the extremities, trunk, and rarely other parts of the body. Thotanet al[3] have described these lesions as mimicking a soft tissue tumor. To the authors' knowledge,no report of gigantic orbitofacialbasidiobolomycosisoccurring bilaterally was found in the English literature. We, therefore, share our experience on diagnostic and management challenges of a rare.

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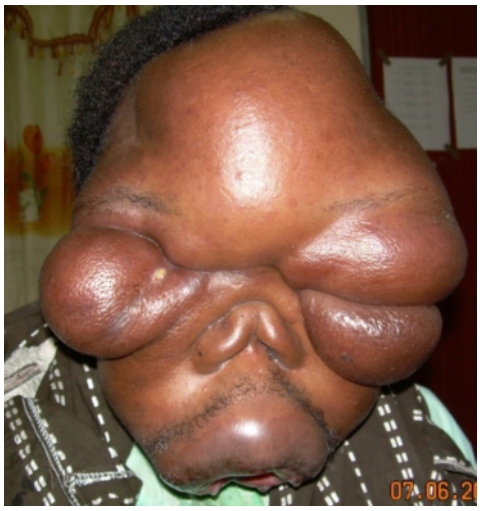

Figure1. Shows grotesque facial swelling (frontal view) of the patient.

case of gigantic orbitofacialbasidiobolomycosis in an otherwise healthy adult Nigerian farmer in a resource poor setting.

Case report:

A 30 year old male farmer presented to us with a one year history of spontaneous cephalofacialswelling that spread rapidly to involve the entire face and head regions. However, there was no history of fever, cough or trauma preceding the swelling. His past medical and social history did not reveal any significant findings, but admitted to have taken oral and herbal concoctions for two months without relief.

On admission, he was weak, mildly pale, anicteric, acyanosed and vital signs were stable. Maxillofacial examination showed bilateral gross cephalofacialdysmorphism (Fig. 1), with affectation of the fronto-parietal, ocular (with inability to part open the eyelids), nasal and orofacial regions. Swellings were smooth and rounded mostly subcutaneous, non-tender, mobile, with mixed consistency (firm and nodular); but baggy and soft around the eyes, with inability to part open the eyelids. The overlying skin was normal. Except for the presence of trismus, the intra-oral, neck and other systemic examinations were essentially normal.

All the blood investigations were normal. Plain radiograph revealed only soft tissue swellings. A CT scan was not done because of financial constraints by the patient.

A wedge shaped intraoral incisional tissue biopsy under local anaesthesia was taken and sent for microbiological and histopathological examinations.

Microbiological examination of tissue wetted with 10% potassium hydroxide revealed broad, irregular hyphae. Culture with Sabouraud's dextrose agar could not be done due to non-availability of the reagent in our centre at the time the patient was seen.

Histopathological examination with Hematoxylin and Eosin stains showed several fungal spores and hyphae surrounded by mixed inflammatory cell infiltrates containing histiocytes, multinucleated giant cells and numerous eosinophils within the stroma classical of Splendore-Hoeppli phenomenon (fig.2). There was, however, no evidence of malignancy.

GrocottGomoriMethenamineSilver was used to stain the tissue specimen which revealed elongated structure with irregular diameter, thin- walled, broad-based ribbon-like, non-septate fungal hyphae within the tissue (Fig. 3). As a result, a presumptive diagnosis of basidiobolomycosis infection was made as definitive diagnosis would require culture of the organism, serological testing via an immunodiffusion method as well as DNA

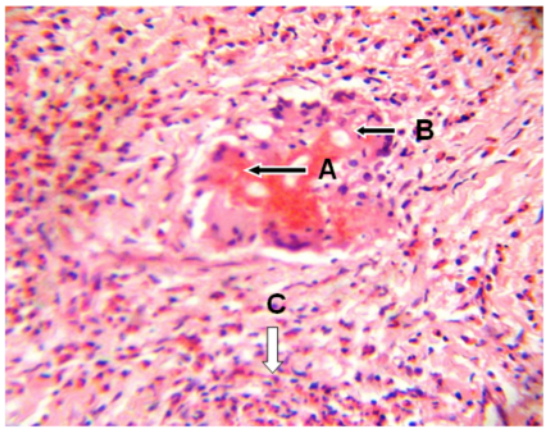

Fig. 2.shows several fungal spores and hyphae (A) surrounded by multinucleated giant cells (B) engulfing them and surrounding stroma showing numerous eosinophils (C). H and E X20.

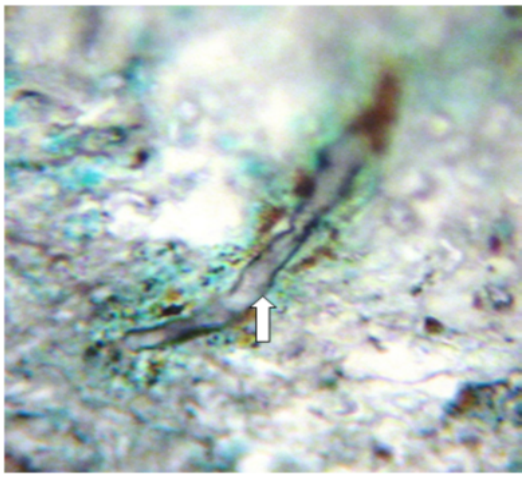

Fig. 3.shows broad-based ribbon-like, non-septate fungal hyphae (arrow) within the tissue. Grocott Gomori Methenamine Silver X100.

sequence analysis (facilities not available in our center). Consequently, the patient was commenced on long term oral antifungal therapy (fluconazole 400mg/day) with successful outcome resulting in a remarkable reduction of facial swelling after four months. Other medication given was ocular genticin for one week. Patient was followed up for a period of one year at monthly interval but was lost thereafter. Patient was located by one of the authors two years later with a residual baggy swelling and ectropion of the right lower eyelid that would require surgical excision (Figure 4).

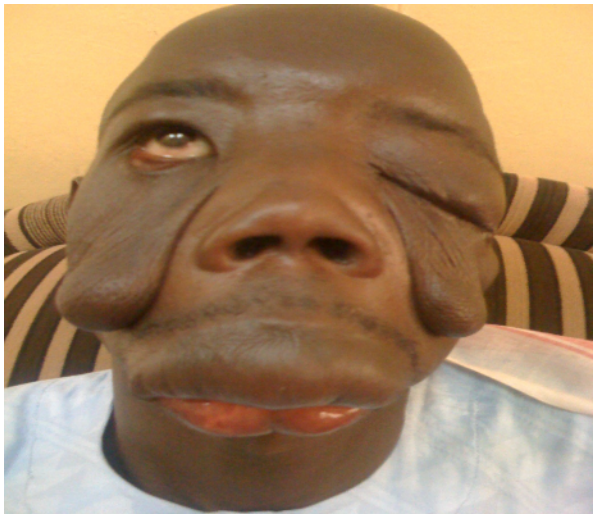

Figure 4. Residual upper lip swelling with baggy tissues and ectropion of the right lower eyelid.

Discussion:

Basidiobolomycosis is a rare fungal infection occurring almost exclusively in immunocompromised individuals[4]. To the authors' knowledge (Medline English literature search), only unilateral cases have been reported in the orbitofacial region and none in the northern part of Nigeria.

Although our patient lacked any of the known predisposing factors the fact that he is a farmer might suggest a possible aetiological factor.

As a results, traumatic implantation could probably be the route of entry in this patient as highlighted in the literature[5, 6] . Hence, the need for physicians, microbiologists and pathologists to consider basidiobolomycosis as a differential for swellings involving the orofacial region as this could increase the probability of an early and accurate diagnosis. Otherwise, these pathogens can disseminate perhaps, with significant morbidity and mortality as seen with the marked trismus resulting in limitation of mouth opening. The management of this patient has not been without its challenges, especially in a resource limited setting like ours. For instance, this patient could not afford the cost of a CT scan. Therefore, the extent of this lesion and its relationship with vital structures in this region could not be ascertained. Again, the response to treatment could not be adequately monitored since the patient was placed on long term antifungal therapy.

The diagnosis of basidiobolomycosis is based on clinical and pathologic features. Confirmation is done by culturing the microorganism on Sabouraud's dextrose agar and lactophenol cotton blue wet mount.[6] The diagnostic dilemma we had was due to nonavailability of the culture medium. Hence, our reliance on presumptive diagnosis using Grocott-gomorimethenamine silver stain technique.

Basidiobolomycosis is a potentially curable disease that mimics a tumor. Cases have been reported in the literature where basidiobolomycosis was misdiagnosed as soft tissue tumor, sarcomas and even as Burkitt's lymphoma[5, 9]. The medical treatment advocated for basidiobolomycosis is with oral saturated potassium iodide therapy, as well as azoles especially intravenous itraconazole.[8] Treatment with terbinafine and itraconazole combination has been used with successful outcome[10]. In the present case, our patient was placed on high dose oral fluconazole. It has the advantage of being safe, cost effective and has few drug interactions[11]. Interestingly in spite of the severity and the presence of poor prognosticators (orbital involvement and trismus with limitation of mouth opening), this patient made a remarkable clinical improvement with oral fluconazole. Regrettably, this patient defaulted on his follow up appointments; although, this is a common phenomenon in our practice. Perhaps, this could possibly be attributed to ignorance and poverty. As a result, the chances of recurrence and even death from this disease remain high.

In conclusion, we have presented an unusual case of orbitofacialbasidiobolomycosis highlighting some peculiar challenges in its management in our practice.

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