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SUBCUTANEOUS ENTOMOPHTHOROMYCOSIS MIMICKING SOFT TISSUE TUMOUR - A CASE REPORT.

Pathology

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ABSTRACT

Entomophthoromycosis (subcutaneous zygomycosis) is a sporadic subcutaneous infection that is largely restricted to tropical areas of Africa, Asia, and South America. It presents in two clinically distinct forms. Subcutaneous zygomycosis caused by Basidiobolus ranarum and rhinofacial zygomycosis caused by Conidiobolus coronatus. It is characterized by formation of firm, non-tender swellings, generally on the extremities, trunk but its occurrence in lip is rare which prompted us to report this case. Our case report also highlights the importance of awareness & early recognition of this entity to prevent disfigurement produced by advanced disease, misdiagnosis, & unnecessary delay in treatment in endemic areas.

KEYWORDS

Entomphthoromycosis.

INTRODUCTION:

Entomopthoromycosis is a rare fungal infection that may affect immunocompetant hosts in tropical and subtropical regions [1].It occurs in two clinical forms Subcutaneous zygomycosis caused by Basidiobolus ranarum and rhinofacial zygomycosis caused by Conidiobolus coronatus. Zygomycosis is a class of fungi which causes infections of respiratory tract and subcutaneous tissues[2]. This class contains teo orders of organisms that cause disease in humans, i.e., Mucorale and Entomophthorale[3.]The disease occurs in the age group of 20-60 years . We present a case of 35 year old farmer presenting as a large rapidly growing upper lip lesion which was initially misdiagnosed as soft tissue tumour. Histopathology is gold standard modality for diagnosis of this rare entity as the fungus is difficult to grow in culture with immunology and serology having limited basis in use for diagnostic purposes. The patient showed marked improvement with treatment of antifungal itraconzole.

CASE REPORT:

A 35 years male presented with gradually increasing painless sub cutaneous swelling on the upper lip since 5 months (Fig 1). Local examination showed that swelling was soft in consistency with smooth rounded edges and was freely mobile over the underlying structures. The overlying surface was lobulated and skin hyperpigmented . There was no regional lymphadenopathy. Based on the history and clinical presentation, a diagnosis of soft tissue tumor was considered. The rest of the general and systemic examination was normal. Routine laboratory investigations revealed an elevated total leukocyte count (21,400/cmm). His HIV status was negative. X-ray of the lip revealed a soft tissue swelling without any bone involvement. Fine needle aspiration cytology of the swelling was suggestive of a spindle cell lesion with few inflammatory cells. The swelling was completely excised and sent for histopathological examination.The excised specimen was grayish pink in multiple bits altogether measuring 8x5 cms(Fig 2). The haematoxylin and eosin stained sections revealed normal skin with subcutaneous tissue showing inflammatory granulomatous reaction with diffuse and dense infiltration by eosinophils, polymorphs, histiocytes, plasma cells, lymphocytes, and few multinucleated giant cells.(fig 3) The central portion showed fragments of aseptate hyphae surrounded by a peculiar eosinophilic mass and the Splendore-Hoeppli phenomenon (fig 4). Culture on Sabouraud's dextrose agar after three days of incubation was negative & in view of typical histopathological findings, diagnosis of entomophthoromycosis was made.

DISCUSSION

Lei-Kian Joe described the first case of entomophthoromycosis in humans in 1956 in Indonesia whose infective agent was Basidiobolus ranarum [4]. In 1965 Bras reported a case in a Jamaican native caused by Conidiobolus coronatus.

Entomophthoromycosis (subcutaneous zygomycosis) caused by Basidiobolus ranarum manifests clinically as a firm, painless, disciform nodule on the trunk or extremities, which if untreated may enlarge and spread locally, but systemic dissemination is extremely uncommon. Rhinofacial zygomycosis caused by Conidiobolus coronatus is a locally progressive infection of the nasal cavity, paranasal sinuses, and soft tissues of the face. The microscopic features of both forms of entomophthoromycosis are similar and give rise to an eosinophilic granuloma situated in the subcutaneous tissues [5].

Entomophthoromycosis has been known as a saprophytic fungus present in soil, decaying fruit, and vegetable matter as well as in the gut of amphibians and reptiles [6].The mode of infection is not known but it is assumed that traumatic implantation may play a role [7]. The infection can be transmitted by insect bites or by transepidermal inoculation with contaminated vegetable matter.

Entomophthoromycosis is a potentially curable disease which can masquerade as a neoplasm possibly due to production of protease enzyme by the fungus by which it is able to invade subcutaneous tissues. Cases have been reported in the literature, where subcutaneous zygomycosis presented as a mimicker of soft tissue tumor, synovial sarcoma, and Burkitt's lymphoma [8-10]. In our patients also, such large size of the swelling, its duration and clinical presentation misguided the clinician to diagnose this as a soft tissue tumor.

Histologically, basidiobolomycosis is associated with eosinophilic infiltration. This has been postulated to be due to a mixture of Th1 (granuloma) and Th2 type of immune response which causes the release of cytokines like IL-4 and IL-10 which in turn are helpful in recruiting eosinophils to the affected site[5]. The other histological features are presence of broad thin walled infrequently septate hyphal fragments enveloped by eosinophilic “Splendore-Hoeppli” material[1] which was prominent in our case.

In the past, clinical isolates of Basidiobolus were classified as B. ranarum, B. meristosporus, and B. haptosporus. But recent taxonomic studies based on antigenic analysis, isoenzyme banding, and restriction enzyme analysis of rDNA indicate that all human pathogens belong to B. ranarum[6].

Chiewchanvit et al.,studied eight cases of entomophthoromycosis out of which five patients were diagnosed as subcutaneous zygomycosis

(females aged 7–77 years), two rhinofacial zygomycosis (26 and 39 years, males) and one case was of gastrointestinal entomopht horomycosis (34 year, male)[11].Our case was having subcutaneous zygomycosis. In a study of ten cases of entomophthoromycosis by Krishnan et al., eight were caused by Basidiobolus haptosporus, predominantly in children below ten years of age, and thigh was the most commonly involved site[12].

CONCLUSION:

To conclude, subcutaneous zygomycosis should be considered in diffential diagnosis of subcutaneous swellings, especially in tropical countries as the condition is curable. Most patients with entomophthoromycosis respond very well to oral potassium iodide therapy as well to azoles, particularly itraconazole [6]. After complete excision and in view of the histopathological report, the patient was started on oral potassium iodide 40 mg/kg per day. After one week of treatment with potassium iodide, oral itraconazole was added in a dose of 100 mg/day and the entire treatment continued for six months. The recovery was uneventful. After six months of followup there was no recurrence. The hematological examination showed total and differential counts within normal limits indicating complete recovery.

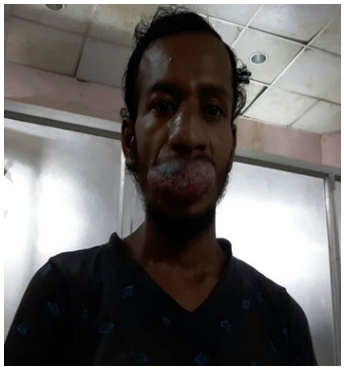

Fig 1-Soft tissue swelling in the upper lip.

Fig 2- Gross-Pinkish tissue in multiple bits together measuring 8x 5cm

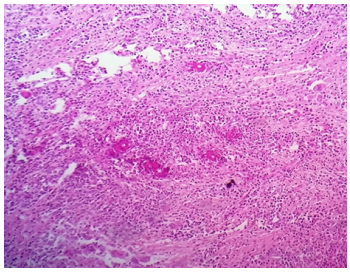

Fig 3- 40X-Showing mixed inflammatory cellular reaction

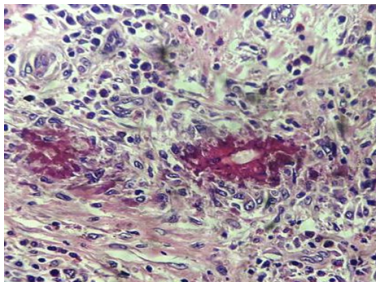

Fig 4 (a)

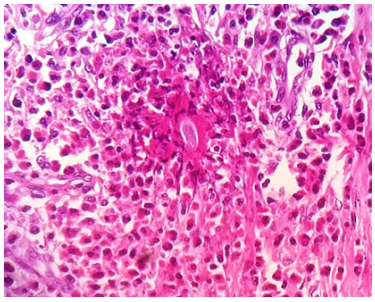

Fig 4 (b)

Fig 4 (a) (b)Showing fragments of aseptate hyphae surrounded by Splendore-Hoeppli Phenomenon

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