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Case Report

Labium superius oris mucormycosis: an enigma

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ABSTRACT

Mucormycosis is an aggressive opportunistic fungal infection affecting immunocompromised patients. It has six manifestations: rhinocerebral, pulmonary, cutaneous, gastrointestinal, disseminated and localized infection. However, isolated mucormycosis of upper lip (labium superius oris) without any bony invasion has not been reported in literature to the best of our knowledge. Hence this poses a diagnostic challenge. A 50-year-old male came with left sided nasal pain and left sided nasal obstruction associated with very mild uniform swelling of upper lip of 24 months duration. He underwent FESS in a local hospital and was misdiagnosed as granulomatous origin probably Tuberculosis. Even on ATT the symptoms didn't subside and hence presented to our hospital. On incisional biopsy deep fungal infection probably mucor was reported and he was put on tab Posaconazole 300 mg OD. On recurrent upper lip swelling, excision biopsy was done. HPE was reported as fibrocollagenous stroma interspersed by multiple non-caseating epitheloid granulomas with plenty of foreign body type of giant cells. PAS showed broad aseptate acute angled hyphal forms suggestive of mucor. The follow up clinical presentation of the patient was very satisfactory with complete regression of upper lip swelling and no damage to surrounding structures. The case report highlights the existence of localized form of mucormycosis without bony necrosis in immunocompetent patients which require minimal surgical intervention along with oral medical therapy rather than extensive debridement.

Keywords: Mucormycosis, Upper lip, Mucor, Upper lip swelling

INTRODUCTION

Mucormycosis also known as black fungus, is a rare and aggressive opportunistic fungal infection usually affecting immunocompromised patients. Mucormycosis is caused by fungi belonging to the order Mucorales of the family Mucoraceae which include rhizopus, rhizomucor, mucor, absidia and apophysomyces[1]. Mucormycosis has six manifestations: rhinocerebral, pulmonary, cutaneous, gastrointestinal, disseminated and localized infection.[2]

Some unusual sites like colon- mucormycosis of colon mimicking carcinoma colon bronchus-mimicking as endobronchial tumour have been seen.[3,4] However, isolated mucormycosis of upper lip (labium superius oris) without any bony invasion has not been reported in literature to the best of our knowledge. Hence this poses a diagnostic challenge.

CASE REPORT

A 50-year-old male came with left sided nasal pain and left sided nasal obstruction associated with very mild uniform swelling of upper lip of 24 months duration. There was no past history of diabetes mellitus or hypertension or renal failure or covid infection or any other immunocompromised conditions. There was no history suggestive of trauma or known allergies or any adverse drug reactions. There were no palpable neck lymph nodes. In October 2020, the patient presented to a local hospital with left sided nasal obstruction and he was diagnosed as left sided nasal polyposis for which he underwent Functional endoscopic sinus surgery (FESS) and the histopathological report (HPE) report of the

excised tissue was of granulomatous origin probably TUBERCULOSIS. Patient was started on Anti-tubercular therapy (ATT) for 6 months according to the Revised National Tuberculosis Control Programme (RNTCP) 2016 guidelines.

Even after completion of ATT, symptoms were persistent. Hence the patient presented to our ENT OPD with left sided nasal obstruction and mild upper lip swelling. On anterior rhinoscopy, left sided firm rubbery pale pink polypoidal mass was seen. The excisional biopsy of the nasal mass and incisional biopsy of upper lip was reported as deep fungal infection probably mucor. Tablet posaconazole 300 mg (100 mg×3 tabs) OD was given and the symptoms began to subside. He continued this treatment for 3 months.

After completion of above treatment, he had 2 months of asymptomatic period. Again, he presented with recurrent swelling of upper lip. Nasal endoscopy was done and biopsy was taken from polypoidal tissue arising from the left lateral wall of nose. HPE showed islands of hyperplastic stratified squamous and fibrous connective tissue with hemorrhage suggestive of allergic polyp while the incisional biopsy of upper lip still showed evidence of deep fungal function suggestive of mucor.

Antihistamines, steroids were added along with tablet posaconazole 300 mg (100 mg×3 tabs) OD for 1 month and the swelling regressed. After 1 month, he again presented with massive upper lip swelling and left sided nasal obstruction. On examination, a 10×6 cm firm tender indurated swelling obliterating upper gingivolabial sulcus along with obliteration of left nasal cavity was seen. Skin over swelling appeared to be normal (Figure 1). CT of paranasal sinuses (CT PNS) showed widening of left osteomeatal unit (post-operative FESS status) (Figure 2 and 3). No bony invasion or sclerosis was detected. Excision biopsy of the lesion under general anesthesia was planned (Figure 4 and 5). Horizontal incision given over upper lip (Figure 4). Dissection done all around the lesion (Figure 4). Pale dense fibrous granulomatous tissue excised (Figure 5). Wound closed in layers (Figure 4).

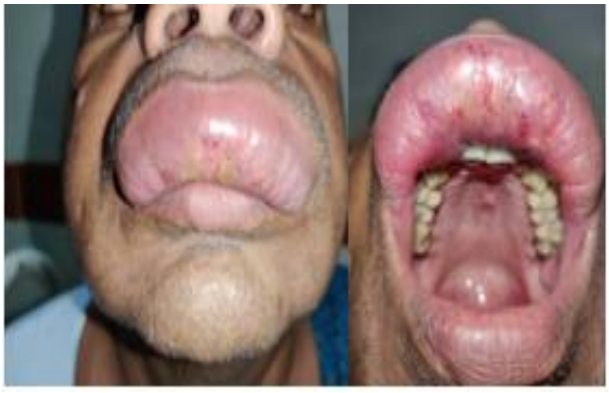

Figure 1: Upper lip swelling (A) front view; and (B) open mouth.

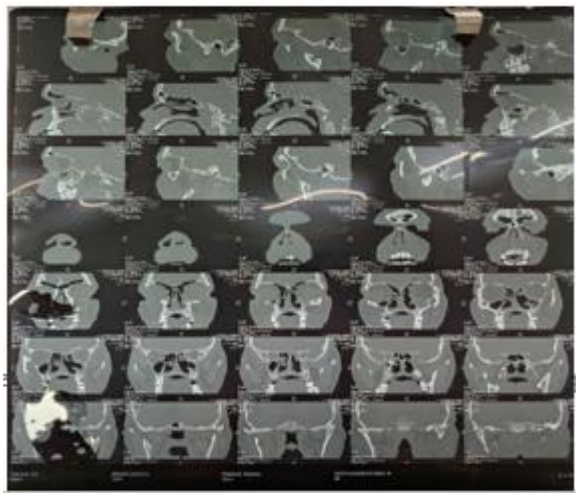

Figure 2: CT PNS film.

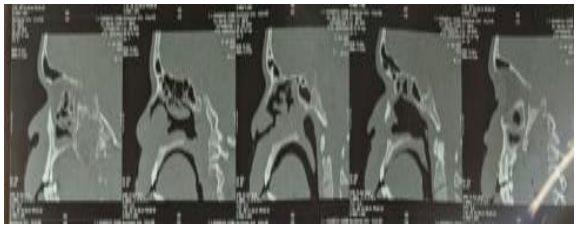

Figure 3: CT PNS sagittal view showing upper lip swelling.

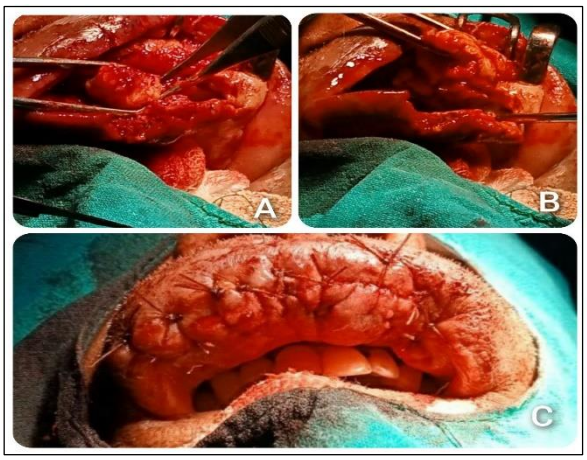

Figure 4: (A and B) Horizontal incision over upper lip and dissection all around the lesion; and (C) wound closure.

Histopathological examination (HPE) was reported as fibrocollagenous stroma interspersed by multiple non caseating epitheloid granulomas with plenty of foreign body type of giant cells (Figure 6). Periodic acid schiff (PAS) showed broad aseptate acute angled hyphal forms suggestive of mucor (Figure 7). The follow up clinical presentation of the patient was very satisfactory with complete regression of upper lip swelling and restoration of upper gingivolabial sulcus within 1 week. The orbicularis oris function was intact. It was tested by

making the lips into an O shape and asking the patient to make Pa, Ba sounds hence indicating no damage to surrounding structures during surgery (Figure 8).

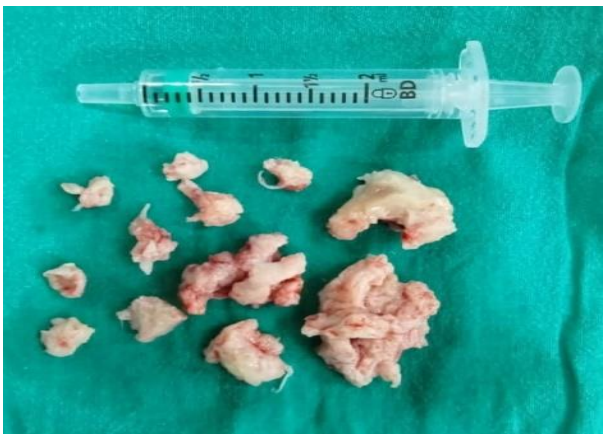

Figure 5: Excised tissue showing pale, dense, fibrous and granulomatous lesion.

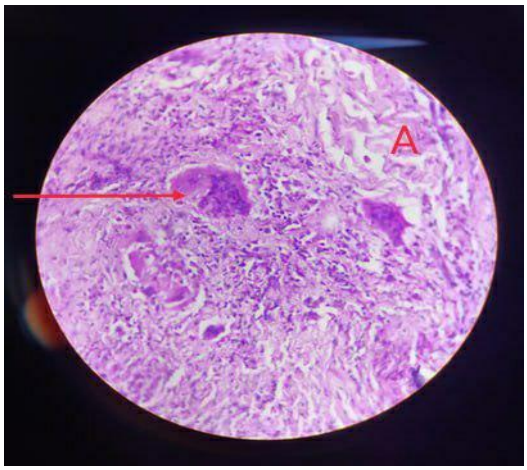

Figure 6: HPE- foreign body granuloma (red arrow showing foreign body giant cells) interspersed in fibrocollagenous tissue (marked as A) of lip.

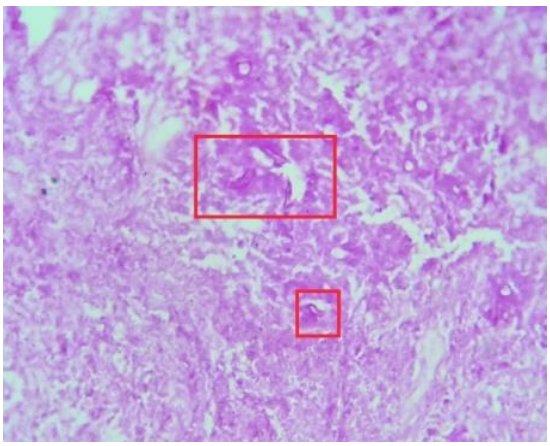

Figure 7: PAS showing 4 acute angled fungal hyphae (3 in rectangular red box and 1 in smaller square box).

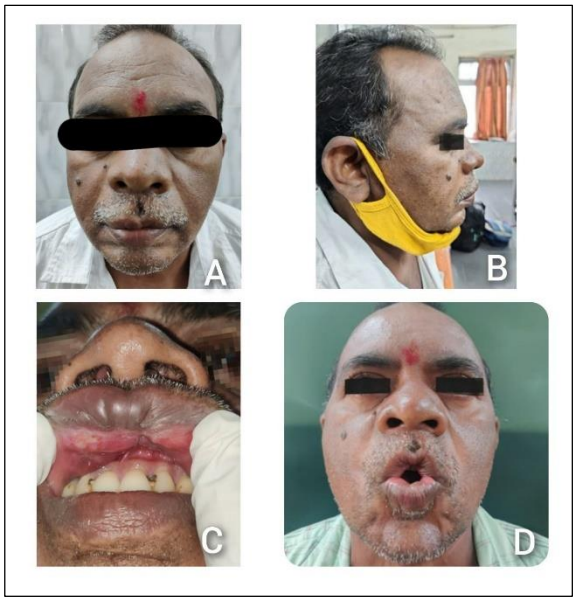

Figure 8: (A, B) Front and side profile of patient on follow up; (C) restoration of upper gingivolabial sulcus; (D) intact orbicularis oris function-making the lips into an O shape.

DISCUSSION

Mucormycosis is a rare fungal infection caused by rhizopus, rhizomucor, mucor, absidia and apophymoses.[1] There had been a steep rise of rhino-orbito-cerebral mucormycosis (ROCM) cases amid second wave of COVID 19 pandemic in India making it a notifiable disease in India. The fungi and its spores are present ubiquitously and are saprophytic in nature. In an immunocompromised host, they rapidly multiply invading the blood vessels leading to progressive avascular necrosis of facial bones and soft tissues. Dissemination of infection to orbit, paranasal sinus and brain is common, if not treated early and aggressively. Mortality, as high as 60% to 80% is associated with mucormycosis of head and neck region. Localised presentation of mucormycosis has been infrequently reported and is commonly seen in individuals with no systemic comorbidities, like diabetes mellitus.[5]

Through this research article, we intend to highlight unusual presentation of mucormycosis of upper lip with minimal involvement of paranasal sinuses or bony erosion. Such rare presentation can result in misdiagnosis as seen in this case report by the local hospital as granulomatous origin -tuberculosis leading to unnecessary usage of drugs and delay in actual treatment, which is crucial for successful outcome.

CONCLUSION

Mucormycosis is a life-threatening infection in immunocompromised patients. Neutropenia, impaired phagocyte function, hyperglycemia, acidosis and corticosteroid treatment are important risk factors. While in this research article the subject is an immunocompetent

patient without any co-morbidities. A hallmark of mucormycosis infection is rapidly progressive angioinvasion with resultant vessel thrombosis and extensive tissue necrosis but here we found no extension to surrounding structures and no bony destruction. This patient appears to be of reactive localized form of mucormycosis without any debilitating complications as seen during the epidemic of rhino-orbital-cerebral mucormycosis (ROCM) in India in 2021.

Rhinocerebral mucormycosis is the most common form of mucormycosis presenting as facial or retro-orbital pain while here, the subject had upper lip involvement with minimal paranasal sinuses symptoms. The treatment usually consists of Inj. amphotericin B along with aggressive surgical debridement.[5] But here it needed local excision of the lesion and medical therapy with tablet posaconazole.

CONCLUSION

Hence, we conclude that such localized form without bony necrosis can be seen in immunocompetent patients which need oral medical therapy and local excision of tissue rather than extensive debridement with removal of the adjacent bony framework.

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