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Case Report

Rhino-orbito-cerebral with cutaneous mucormycosis

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ABSTRACT

"Mucormycosis" term is used for a group of fungal diseases caused by the order mucorales. It typically presents as sino-nasal-orbital involvement and rarely involves skin. Management of such cases is quite challenging. Reported here is a case of a young girl, who presented with right eye swelling and reduced vision and got detected with type 1 diabetes mellitus on admission with rare occurrence of cutaneous lesions while on treatment. Direct nasal endoscopy with biopsy was undertaken. KOH and histopathology report was suggestive of mucormycosis, for which the patient was administered intravenous amphotericin B and surgical debridement was carried out. With the accurate application of a multidisciplinary approach with timely investigations, imaging studies, antifungals and debridement, successful outcome in mucormycosis is well within our reach.

Keywords: Cutaneous mucormycosis, Rhino-cerebral mucormycosis, Amphotericin, Diabetes

INTRODUCTION

Mucormycosis is an opportunistic and fulminating fungal infection caused by members of the family mucoraceae, order mucorales and class zygomycetes. Risk factors for mucormycosis are diabetes mellitus, immunosuppression, leukaemia, metabolic acidosis, organ transplants, intravenous drug abuse. The clinical presentations can be rhino-cerebral (most common), pulmonary, gastrointestinal, cutaneous and disseminated. The infection develops after inhalation of fungal sporangiospores into the nasal cavity.[1] The infection then rapidly extends into the adjacent tissues, inferiorly to invade the palate, posteriorly to invade the sphenoid sinus, laterally into the orbit and the cavernous sinus. The fungus invades the cranium through either the orbital apex or cribriform plate and ultimately can prove fatal.[1] Occasionally, cerebral vascular invasion can lead to haematogenous dissemination of the infection causing necrosis of the vessel wall and mycotic thrombi. The initial symptoms of rhino-orbito-cerebral mucormycosis (ROCM) are consistent with those of sinusitis and periorbital cellulitis and include eye and/or facial pain and facial numbness followed by vision disturbance. Cutaneous mucormycosis can be due to direct inoculation on skin or by dissemination from rhinocerebral infection. Most affected areas of skin are the arms and legs.[2] At our institute, we have treated many patients with ROCM, most of whom have been detected with or are known cases of diabetes mellitus. This is a case of a young girl, who got detected with type 1 diabetes mellitus with ROCM on admission. We presented this case with rare occurrence of cutaneous mucormycosis while on treatment needing a multidisciplinary approach for appropriate management.

CASE REPORT

An 18 years old female was referred to out-patient department of our tertiary care hospital with history of right eye swelling and right eye reduced vision since 2 days which progressively worsened until she had no light perception. She had no complaints of nasal discharge/blockage. Patient did not give any history of COVID infection or steroid intake. Patient was a newly diagnosed case of type I diabetes mellitus with

uncontrolled sugars not on any medication. Examination revealed an ill looking, debilitated girl. She had ptosis and periorbital edema with no light perception in right eye however the left eye was normal. Examination of nasal cavity showed extensive blackish-crusting with discharge in right nasal cavity with normal left side. Intra-orally there was mild discolouration noticed in the anterior region of hard palate with no palatal perforation (Figure 1). Direct nasal endoscopy of patient was done which was suggestive of crusting over the right middle turbinate and the septum (Figure 2). Specimens were sent for microscopic examination with KOH mount and histopathology with GMS staining. Both reports were suggestive of broad aseptate fungal hyphae (mucor) (Figures 3 A and B). Patient was immediately started on antifungal-amphotericin B with monitoring of blood gas, sugars, electrolytes and renal function. She was given intravenous 50 mg of liposomal amphotericin B as test dose followed by 200 mg daily with pre and post amphotericin hydration by normal saline with 15 cc KCl in each pint. A daily Amphotericin B chart was maintained for patient with daily input/output ratio, serum creatinine, urea, sodium, potassium and magnesium.

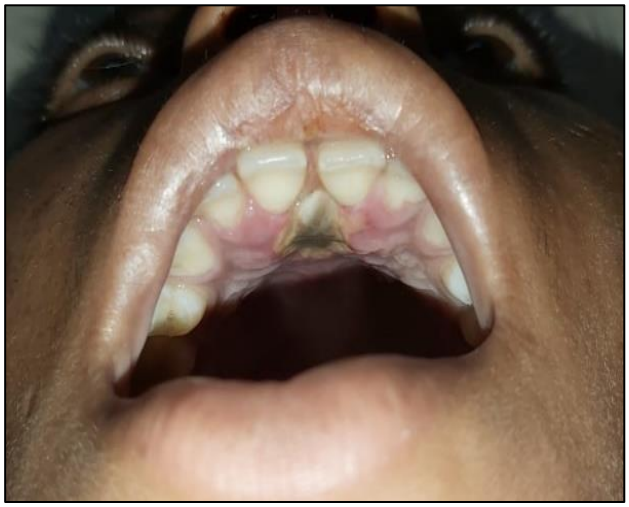

Figure 1: Discolouration over anterior hard palate.

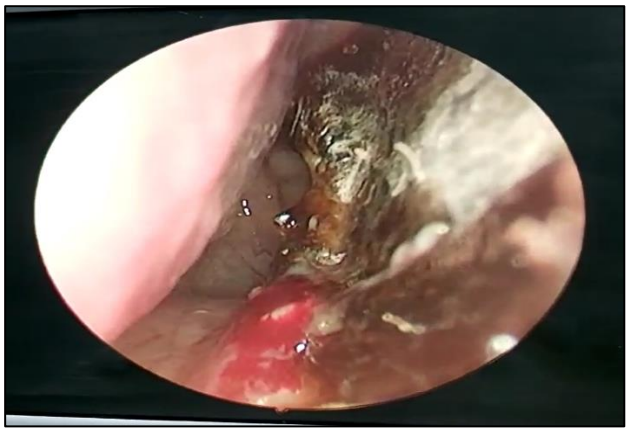

Figure 2: Right nasal cavity crusting.

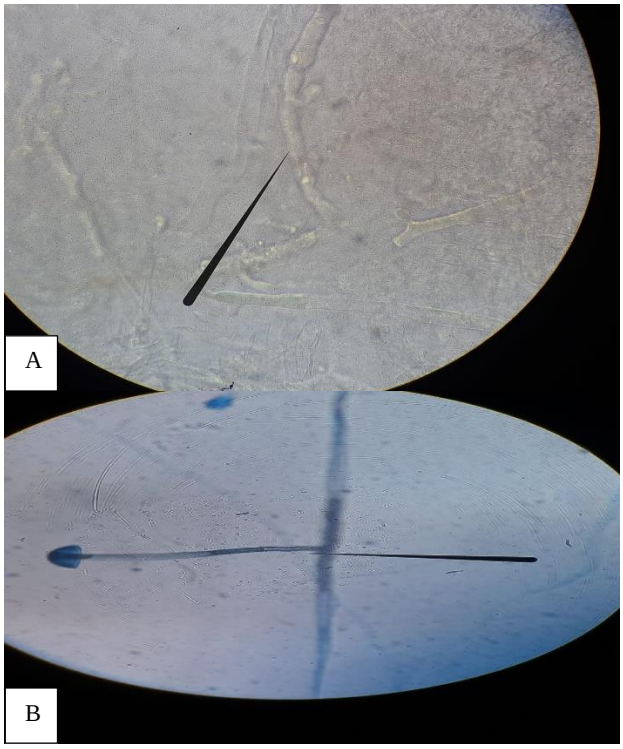

Figure 3 (A and B): Broad aseptate fungal hyphae on KOH mount. Fungal hyphae at right angle on lactophenol cotton blue staining.

Medicine opinion was taken for the poor general condition of the patient, her newly diagnosed diabetes and uncontrolled sugars (ranging from 300-350 fasting and 400 postprandial). HbA1C of the patient was >15. She was immediately started on plain insulin drip followed by a combination of plain and lente insulin. Ophthalmologist were consulted for the orbital swelling and vision loss of patient for which they gave 3 doses of transcutaneous retrobulbar amphotericin B injection (TRAMB) on alternate days, and MRI orbit was advised.

CT paranasal sinus was done which was suggestive of mildly enhancing heterogenous lesion present in right nasal cavity and paranasal along with right orbital cellulitis and partial right cavernous sinus thrombosis. MRI brain + orbit was suggestive of Ill-defined T1 isointense and T2 hyperintensity in ethmoidal air cells along the lamina papyracea and involving medial extraconal compartment extending into intraconal and encasing the infraorbital part of optic nerve with intracranial extension.

Endoscopic debridement of right sino-nasal mucormycosis was performed under general anaesthesia after obtaining a written, informed and documented consent explaining the high risk. All sinuses were opened, necrotic tissue and fungal debris was debrided along with unhealthy mucosa till healthy bleeding mucosa was seen. The affected palatal mucosa was removed, underlying bone was kept intact. Patient was advised full nasal douching in postoperative period.

According to the Sion hospital scoring system for orbital exenteration in rhino-orbito-cerebral mucormycosis, the patients score was 21, hence orbital exenteration was not indicated.[3] Subsequently there was improvement in her vision which progressed upto hand-movements.

Patient had a cutaneous lesion with pus discharge over her left forearm on the site of her previous IV line (Figure 4). USG Doppler of left upper limb was done which showed complete lumen occluding thrombus of left basilic vein, median cubital vein and cephalic vein. Debridement of the left forearm lesion was done and sample sent for KOH mount. The skin lesion of patient started aggravating and she developed multiple lesions over both forearms. Koh report of specimen was positive for mucormycosis. Infectious disease opinion was sought for the patient and she was started on anticoagulants and tablet posaconazole 300 mg daily. HRCT chest and CT abdomen showed no other organ involvement.

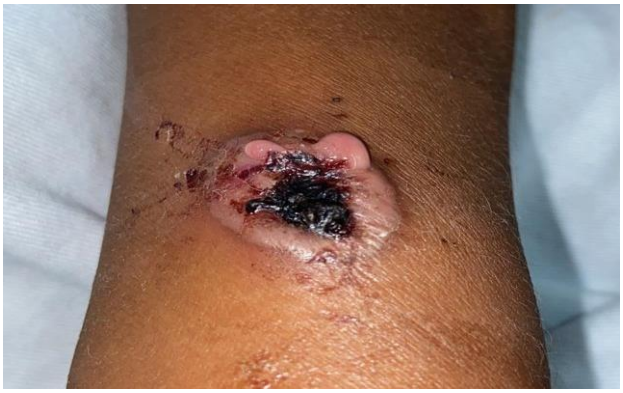

Figure 4: Cutaneous lesion over left forearm.

She received total of 2.8 g of amphotericin and 7 grams of posaconazole over a period of 1 month. After treatment the cutaneous lesions over both the upper limbs healed (Figure 5). Repeat nasal endoscopy showed healthy mucosa with wide-opened sinuses and the KOH mount was found negative of fungal hyphae. Amphotericin-B was stopped and she was discharged. Risk of recurrence was well explained to the patient and her relatives and was kept on close follow-up.

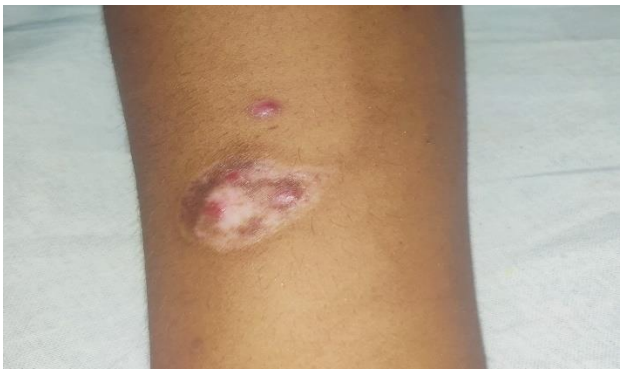

Figure 5: Cutaneous lesion post debridement and antifungals.

DISCUSSION

Mucormycosis is an invasive fungal infection first described by Paulltauf in 1885. Interestingly though mucormycosis has been noted to be an uncommon occurrence in patients with AIDS.[4] Impaired neutrophil and phagocyte response and increased available serum iron are the two underlying conditions in the majority of mucormycosis patients. Rhinocerebral mucormycosis most commonly presents in an acute setting mimicking symptoms of sinusitis or periorbital cellulitis. The only disease specific finding described in literature is blackened necrotic eschars on the nasal mucosa or palate.[4]

The diagnosis of mucormycosis can be made by direct microscopy or histopathological examination with special stains like GMS stain and PAS stains, or by culture on Sabouraud's agar. The detection of aseptate hyphae with right angled branching is pathognomonic. CT scans can be used to evaluate the progression of disease although correlation with the clinical findings may not always be accurate. MRI scans are more accurate in evaluating the extent of disease due to fungal invasion of soft tissues.[4] Our patient had involvement of all the sinuses except frontal sinus. A study conducted showed frequent involvement of the ethmoid (86%) and maxillary (80%) sinuses followed by sphenoid and frontal (17%).[5] Orbital involvement is observed in 80% as an orbital mass and/or thickening of the recti and optic nerve is seen.[5] Intracranial extension causes involvement of the frontal or the temporal lobes.

The main line of management of mucormycosis is antifungals along with surgical debridement. Reversal or control of underlying predisposing conditions is of paramount importance.[4] TRAMB can be considered as a viable option in select cases of orbital mucormycosis where exenteration or debridement are not indicated, or when there is limited orbital disease. Based on the Sion hospital scoring system for orbital exenteration, it was observed that those patients who crossed a score of 23 were eligible candidates for orbital exenteration.[3] Our patient had a score of 21 and hence orbital exenteration was avoided. Diagnosis of cutaneous mucormycosis can be difficult as initial symptoms are often non-specific and can mimic a variety of infectious skin diseases.[6] It needs extensive surgical debridement and antifungal therapy along with correction of the underlying metabolic or impaired immunological status.

Amphotericin-B is the first line systemic anti-fungal for mucormycosis. Posaconazole has been described as second line therapy in patients with failure or intolerance to amphotericin B or for those who need prolonged treatment.[2] Isavuconazole is a recently approved drug for treatment of invasive mucormycosis. HBO_2 and G-CSF are the other medical management options that have shown some promise. They act by enhancing the leukocyte killing capacity and increase oxygen delivery

to the tissues. However, their role is only additive to systemic anti-fungals.[3]

CONCLUSION

Mucormycosis is a fatal invasive fungal disease that requires a high level of clinical skills for early diagnosis and prompt treatment in order to improve survival. In spite of early diagnosis, a clear treatment algorithm is absolutely crucial to reach clinical and paraclinical improvements and to achieve survival in patients. Controlling underlying conditions is an important aspect of therapy. We could manage our patient with ROCM and cutaneous involvement only with the help of various departments. We continue to monitor the patient on OPD basis. Such a multidisciplinary approach can help to cure the patient off this debilitating disease with high mortality rate.

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REFERENCES

1. Singh V, Singh M, Joshi C. Rhinocerebral mucormycosis in a patient with type 1 diabetes presenting as toothache: a case report from Himalayan region of India. BMJ Case Rep. 2013;2013:bcr2013200811.
2. Castrejón-Pérez AD, Welsh EC, Miranda I. Cutaneous mucormycosis. An Bras Dermatol. 2012;92:304-11.
3. Shah K, Dave V, Bradoo R. Orbital Exenteration in Rhino-Orbito-Cerebral Mucormycosis: A Prospective Analytical Study with Scoring System. Indian J Otolaryngol Head Neck Surg. 2019;71:259-65.
4. Oladeji S, Amusa Y, Olabanji J. Rhinocerebral mucormycosis in a diabetic case report. J West Afr Coll Surg. 2013;3:93-102.
5. Sharma A. Presentation and outcome of rhino-orbital-cerebral mucormycosis in patients with diabetes. Postgraduate Med J. 2004;80:670-4.
6. Maleitzke T, Stahnke K, Trampuz A. A case report of cutaneous mucormycosis of the hand after minor trauma in a patient with acute myeloid leukaemia. Trauma Case Rep. 2019;23:100221.

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