DOI: 10.7860/JCDR/2023/62122.17461

Case Report

Ear, Nose and Throat Section

An Unconventional Presentation of Mucormycosis in a 10-Year-Old Child: A Case Report

ANANNYA TRIPATHY[1], LAKSHMI KRISHNAN[2], JAMES THOMAS[3], HARSH SINGH[4]

ABSTRACT

Since the Coronavirus Disease (COVID-19) pandemic, there have been several unusual presentations of mucormycosis in India, especially amongst immune-competent adults. COVID-19 infection has been found to have profound effects on the patient's immunity and some patients, though asymptomatic for COVID-19, can be infected by mucormycosis and develops dangerous complications. Skin involvement of the orbital, zygomatic and maxillary areas is a common occurrence in extensive cases of rhino-orbital mucormycosis, however, isolated involvement of the alar skin is an extremely rare occurrence in such patients. Paediatric cutaneous mucormycosis is by itself a rare entity, seen majorly in children with history of allogeneic hematopoietic stem cell transplantations, chemotherapeutic treatment, or patients with human immunodeficiency virus infections, herpes and other life-threatening viral infections. Patients receiving long-term steroid therapy are also predisposed to invasive fungal infections. This case was about a 10-year-old boy presented with a black crusted lesion over the nose to the otolaryngology outpatient department. The patient had history of contact with a COVID-19 positive individual. Examination revealed a necrotic patch over the palate and Non Contrast CT Scan of (NCCT) the paranasal sinuses showed pansinusitis. A KOH mount showed fungal elements and the patient underwent emergency debridement of nasal skin with endoscopic sinus and palatal debridement. Injectable liposomal Amphotericin-B was started. Over a period of one month, the patient showed significant clinical improvement. Though rare, sinonasal mucormycosis can present in the form of a cutaneous lesion which is an unconventional symptom. A general awareness amongst healthcare professionals, with a multidisciplinary approach, timely diagnosis and specialist intervention can improve outcomes in this sinister disease.

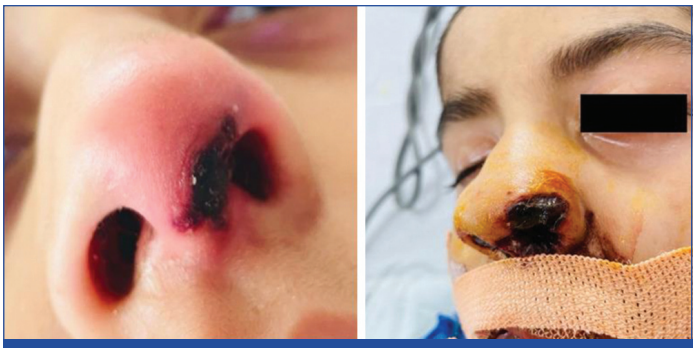
Keywords: Cutaneous, Nasal, Otorhinolaryngology, Paediatrics

CASE REPORT

A 10-year-old boy presented to the otorhinolaryngology OPD with a progressively increasing blackish discolouration of the left-side of the nose for two days. The parents also gave history of fever and sore throat for four days.The patient had no co-morbidities and was born of a non consanguineous marriage, having completed his immunisation till date. All developmental milestones had been attained normally. The patient had a history of having contact with his grandfather 15 days prior, who later tested COVID-19 positive. Informed consent was obtained from parents of the child for anonymised patient information and images to be published.

At presentation, the patient had a temperature of 102 degrees Fahrenheit and tachycardia. Extensive blackish crusting over the left ala and columella with surrounding indurated area of inflammation was present. The patient also had a blackish necrotic patch of approximate size 3×2 centimeters over the left-side of the soft palate [Table/Fig-1,2]. An elevated total leucocyte count of 27,000 cells/mL, raised ESR of 89 mL/hour and CRP of 235 mg/L were seen, with a mildly elevated serum procalcitonin level measuring 0.82 ng/mL. With the pandemic in mind and history of interaction with a COVID-19 patient, a provisional diagnosis of cutaneous and palatal mucormycosis was immediately reached.

The child was admitted to the paediatric ICU, and swabs taken from the palate and crusts from the nose were sent for immediate KOH mount. The patient was started empirically on injectable ceftriaxone and linezolid, intravenous fluids and injection paracetamol, as per weight by the paediatricians. COVID-19 RT-PCR sample was also sent, which was negative. However, the patient had tested positive for COVID-19 Ig-G antibodies. KOH mount of the nasal scrapings showed pauciseptae with flat acute angled fungal hyphae in 10%

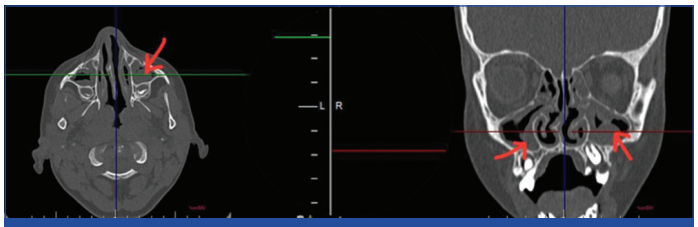


[Table/Fig-1]: Crusted lesion over nasal ala.

[Table/Fig-2]: Necrotic patch over palate.

KOH mount after 4-8 hours of incubation at 37ºC suggestive of infection with zygomycetes group fungi.

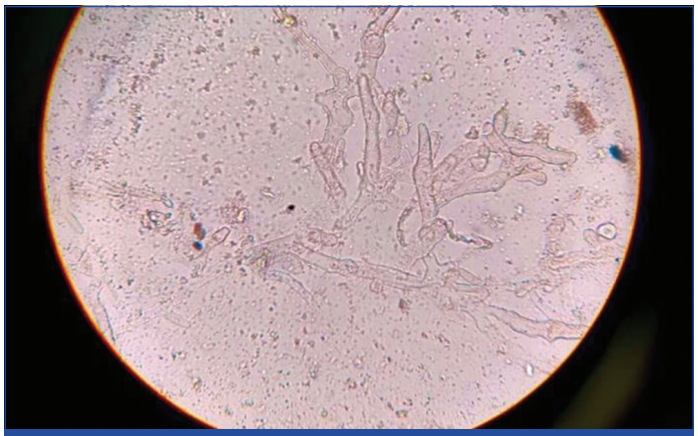
A NCCT of the paranasal sinus was done, which showed mucosal thickening in bilateral frontal, ethmoidal, maxillary and sphenoid sinuses, involving both the osteo-meatal units. However, no bony erosion or ocular involvement was noted [Table/Fig-3]. A provisional diagnosis of pansinusitis along with the differentials of fungal sinusitis was considered. According to history, radiological findings and a positive KOH mount, a final diagnosis of sinonasal mucormycosis with cutaneous involvement was made.



[Table/Fig-3]: NCCT PNS showing mucosal thickening (red arrows).

After hospitalisation, the patient developed breathlessness and required endotracheal intubation while the lesion over the nose also grew rapidly. Emergency endoscopic sinus debridement with local debridement of the ala, under general anaesthesia was taken up.

Intraoperatively, bilateral uncinectomy and maxillary antrostomy was done. Bilateral ethmoid and maxillary debridement with frontal and sphenoid sinus clearance was also done. The necrotic mucosa of the soft palate and the eschar over the nose was also debrided extensively. All debrided tissues and diseased mucosa were sent for KOH mount and histopathological examination. The intraoperative KOH mount of both the sinus debridement specimen as well as the alar crust showed fungal elements with aseptate hyphae [Table/Fig-4]. Rhizomucor species was seen in fungal culture (Sabouraud Dextrose Agar).

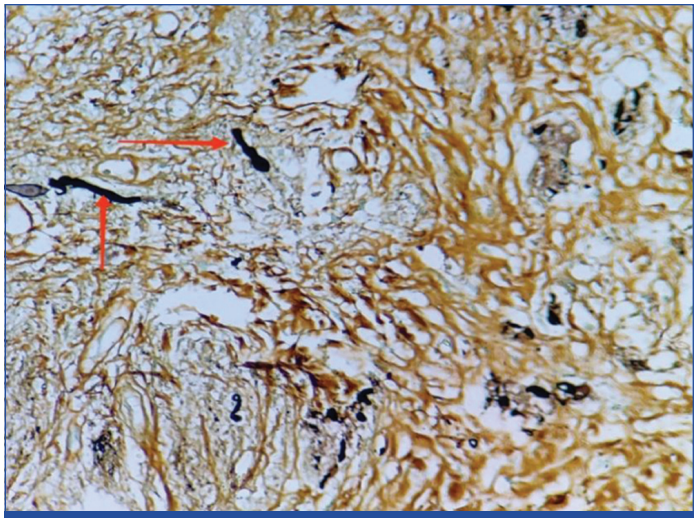


[Table/Fig-4]: KOH mount showing fungal elements (10% KOH, 400X).

Periodic acid-Schiff/Grocott Methenamine Silver (PAS/GMS) staining on histopathological examination showed epithelium with chronic inflammatory cell infiltrate, numerous broad wide angled branching and aseptate hyphae of mucor [Table/Fig-5].

Postoperatively, the patient was started on Injection liposomal Amphotericin-B as per weight. The lesion over the nose was dressed daily and topical application of lipid-based Amphotericin -B gel (0.1w/w) was done.

General condition had been improved, whereby the patient was extubated on the fifth postoperative day. However, the nasal lesion showed continued increase in size, and an intra-lesional injection of Amphotericin-B was performed on fourth postoperative day, as a salvage procedure. When the patient was stabilised and weaned off oxygen support, shifted to the paediatric ward, wherein course of injectable amphotericin had been completed. Thorough daily dressing of the wound and topical amphotericin gel application

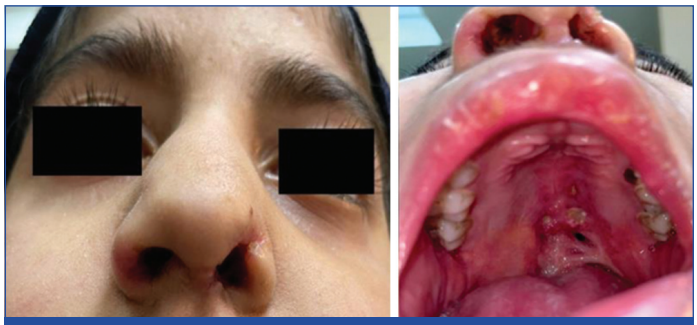


[Table/Fig-5]: GMS staining on histopathological examination revealing aseptate hyphae of mucor (red arrows) (400X).

continued. The alar lesion regressed in size, and healing of the palatal eschar occurred.

The patient was discharged on the 14th postoperative day, with instructions to carry out regular saline nasal douching and continued topical application of amphotericin gel.

On follow-up at one week, the lesion over the ala showed adequate healing, with no residual crusting or nasal symptoms. The palatal eschar was healing, and mucosal regeneration was appreciated at the site of the lesion over the palate [Table/Fig-6].



[Table/Fig-6]: Healed skin lesion and regenerating mucosa over palate.

The patient was kept on regular follow-up and within a month most of the affected tissues had returned to normal status with minimal scarring. However, the patient had a defect on his ala in the form of a concave notch, for which delayed reconstructive surgery for cosmesis had been advised.

DISCUSSION

Mucormycosis is a lethal, fulminant, angio-invasive and bone eroding disease, which was first described by Paltauf, in 1885 as mycosis mucorina. It is primarily a disease affecting immunocompromised patients [1]. Historically, it occurs in persons with uncontrolled diabetes, haematogenous malignancies, traumatic wounds with direct inoculation and patients undergoing chemotherapy and other immunosuppressive treatments [2]. Since the onset of the COVID-19 pandemic in 2019, numerous rare presentations of the disease especially in immune-competent individuals were witnessed. COVID-19 infection has been found to have profound effects on patient's immunity and some patients, though asymptomatic for COVID-19, can acquire mucormycosis and develop dangerous complications.

In a review of cases by Singh AK et al., it was found that 101 cases of mucormycosis were seen in COVID-19 patients (82 cases from India and 19 from rest of the world). It was seen predominantly in males (78.9%) and in both active cases (59.4%) as well as those who had recovered (40.6%) [3]. However, cases are severely under-reported and diagnosis is delayed in many cases. Two cases of

post COVID-19 rhino-orbito-mucormyosis in children have been reported, however these were in children diagnosed with Type-I diabetes mellitus, and did not show skin involvement [4]. In present case report, a ten-year-old boy was diagnosed with sinonasal mucormycosis with cutaneous involvement. The patient was COVID-19 negative however tested positive for COVID-19 Ig-G antibodies.

Skin involvement of the orbital and maxillary areas is common in extensive cases of rhino-orbital mucormycosis; however, isolated involvement of the alar skin has not been reported previously [1,5]. In present case report, extensive blackish crusting over the left ala and columella was present. The patient also had a blackish necrotic patch over the left-side of the soft palate. Paediatric cutaneous mucormycosis is by itself a rare entity, seen majorly in children with history of allogeneic hematopoietic stem cell transplantations, chemotherapeutic treatment, or patients with human immunodeficiency virus infections and herpes virus infections [6]. The common manifestations of sinonasal mucormycosis in children were fever, rhinorrhea, facial erythema and oedema, and pale nasal mucosa [7]. Patients receiving Long-term steroid therapy and voriconazole prophylaxis are also predisposed to invasive fungal infections [8].

In cutaneous mucormycosis, the most commonly isolated genus is *Rhizopus* spp., with the *Rhizopus oryzae, Lichenthemia corymbifera, Cunninghamella bertholletiae, Rhizomucor* spp., and *Rhizopus* microspores being the predominant strains. *Rhizopus arrhizus* is the predominant etiological agent of mucormycosis in India [2,9]. The prognosis of this disease has been historically grave, especially when neurological, ophthalmological and disseminated spread occurs [1]. In present case report, KOH mount of the nasal scrapings showed pauciseptae with flat acute angled fungal hyphae, suggestive of infection with zygomycetes group fungi.

India already showed a higher number of cases prior to the pandemic, owing to the higher incidence of undetected/uncontrolled diabetes mellitus. Following the COVID-19 pandemic however, the disease has been seen in a number of immunocompetent individuals, with a variety of uncommon presentations [3]. A high index of suspicion and a multidisciplinary approach is necessary for favourable outcomes. In present case, bilateral uncinectomy and maxillary antrostomy along with bilateral ethmoid and maxillary debridement with frontal and sphenoid sinus clearance was done. The necrotic mucosa of the soft palate and the eschar over the nose was also debrided extensively.

Reported cases of mucormycosis in paediatric age group are relatively few, and almost none in an immuno-competent child [10,11]. Considering the humongous number of COVID-19 infections which occurred during the pandemic, children presenting with such cutaneous lesions need to be carefully examined to rule out superadded mucormycosis. As with this child, who presented with only a skin lesion, without nasal symptoms, it is essential that a quick diagnosis is reached and surgical debridement is done, followed by adequate anti-fungal therapy.

CONCLUSION(S)

In most paediatric cases of mucormycosis infection, the children are immunocompromised, however healthcare professionals need to be aware of the less obvious presentations such as a cutaneous lesion alone, as seen in this particular patient. The need to facilitate swift and appropriate management to reach a quick and correct diagnosis should be understood by healthcare professionals.

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PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of Otorhinolaryngology Head and Neck Surgery, Dr. D. Y. Patil Medical College and Hospital, Pimpri, Pune, Maharashtra, India.
2. Junior Resident, Department of Otorhinolaryngology Head and Neck Surgery, Dr. D. Y. Patil Medical College and Hospital, Pimpri, Pune, Pune, Maharashtra, India.
3. Professor, Department of Otorhinolaryngology Head and Neck Surgery, Dr. D. Y. Patil Medical College and Hospital, Pimpri, Pune, Pune, Maharashtra, India.
4. Senior Resident, Department of Otorhinolaryngology Head and Neck Surgery, Dr. D. Y. Patil Medical College and Hospital, Pimpri, Pune, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Lakshmi Krishnan,
A/61, Carnation Girls Hostel, Dr. D. Y. Patil Medical College and Hospital,
Sant Tukkaram Nagar, Pimpri, Pune-411018, Maharashtra, India.
E-mail: drlakshmikrishnan92@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Dec 09, 2022
- Manual Googling: Jan 19, 2023
- iThenticate Software: Jan 24, 2023 (6%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Dec 07, 2022**
Date of Peer Review: **Dec 30, 2022**
Date of Acceptance: **Jan 27, 2023**
Date of Publishing: **Feb 01, 2023**