

Bilateral blindness in a young male of rhino-orbital-cerebral mucormycosis: A case report

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The storm of COVID-19-associated mucormycosis (CAM) has not yet settled, and it has proven itself a disfiguring and potentially life-threatening disease, complicating the course of COVID-19 infection. Mucormycosis is a rare but devastating fungal infection caused by filamentous fungi of the family Mucoraceae. We report a rare case of a 37-year-old diabetic male with bilateral rhino-orbital-cerebral mucormycosis (ROCM) where it leads to bilateral central retinal artery occlusion (CRAO) as manifestation of the disease. Bilateral CRAO secondary to ROCM is extremely rare. A strong suspicion of CAM in uncontrolled diabetics can result in early diagnosis and management.

Key words: Central retinal artery occlusion, COVID-19, mucoraceae, rhino-orbital-cerebral mucormycosis

Though reported from many parts of the globe, India particularly has had an alarming epidemic of CAM from April to June 2021. But the storm still has not settled, COVID-19 associated mucormycosis cases are still presenting with vision-threatening as well as life-threatening manifestations. Mucormycosis almost invariably affects persons with immunocompromised states, especially those with diabetes complicated by ketoacidosis, organ transplant recipients, patients on steroid or cytotoxic therapy, and those with leukemia and other disseminated cancers. The accepted clinical forms of mucormycosis are rhino-orbital-cerebral mucormycosis (ROCM), pulmonary, cutaneous, gastrointestinal, disseminated, and miscellaneous.^[1] The myriad of atypical clinical manifestations and complexity in demonstration of the causative organism poses a diagnostic challenge, which heralds a poor prognosis.

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Case Report

A 37-year-old male presented to emergency department with chief complaints of fever and cough for five days, increased urine output for one day, and shortness of breath since morning. Patient was diabetic for seven years on regular insulin, and hypertensive for three years on regular treatment. Patient was investigated and was diagnosed with diabetic ketoacidosis with pyelonephritis. Renal function tests were altered showing serum creatinine at 2.5 mg/dl and blood urea at 75 mg/dl. Ultrasound abdomen showed bilateral parenchymal disease along with pyelonephritis. Urine routine examination showed sugar 3+ and albumin 3+. Lymphopenia was noted (12% per cu mm).

On the next day of admission, the patient complained of diminution of vision in the left eye which was painless, sudden and progressive. On detailed ophthalmic examination, bedside visual acuity was 20/400 in the right eye and negative perception of light in the left eye. There was complete restriction of extraocular movements in the left eye and restriction of lateral movement in the right eye. On torch light examination the patient had bilateral incomplete ptosis, left eye lower lid edema, left ocular paresthesia, bilateral absent corneal sensations, with no conjunctival chemosis, congestion and proptosis [Figs. 1a and 1b]. Dilated fundus examination revealed normal fundus in the right eye and CRAO in the left eye.

Brain CT complemented with contrast-enhanced CEMRI of brain, PNS, and orbits showed bilateral maxillary and ethmoid sinusitis, bilateral medial and inferior turbinate hypertrophy, left premaxillary and left preseptal orbital cellulitis, acute infarct in left medial basifrontal lobe with bilateral cavernous sinus thrombosis (left > right). Mild narrowing of left internal carotid artery was also noted [Figs. 1d and 1e]. KOH report of nasal swab revealed broad aseptate hyphae. The patient was put on posaconazole, and on seeing his kidney status, amphotericin B was withheld. Patient gave history of fever 2 months back. The patient was COVID-19 RT-PCR negative. Fundus examination was repeated after 24 hours which revealed bilateral CRAO [Fig. 1c]. The patient underwent emergency left maxillectomy with left exenteration with right modified Denker approach under general anesthesia. Postoperatively, the patient was put on mechanical ventilation. Histopathological examination of sections from resected margin of optic nerve showed area of necrosis along with the presence of numerous broad aseptate, obtuse-angled branching hyphae invading the nerve bundles, thus confirming the diagnosis of mucormycosis [Fig. 1f]. On 5th postoperative day, patient developed sudden bradycardia. All resuscitation measures were started immediately but unfortunately the patient could not be revived and was declared dead.

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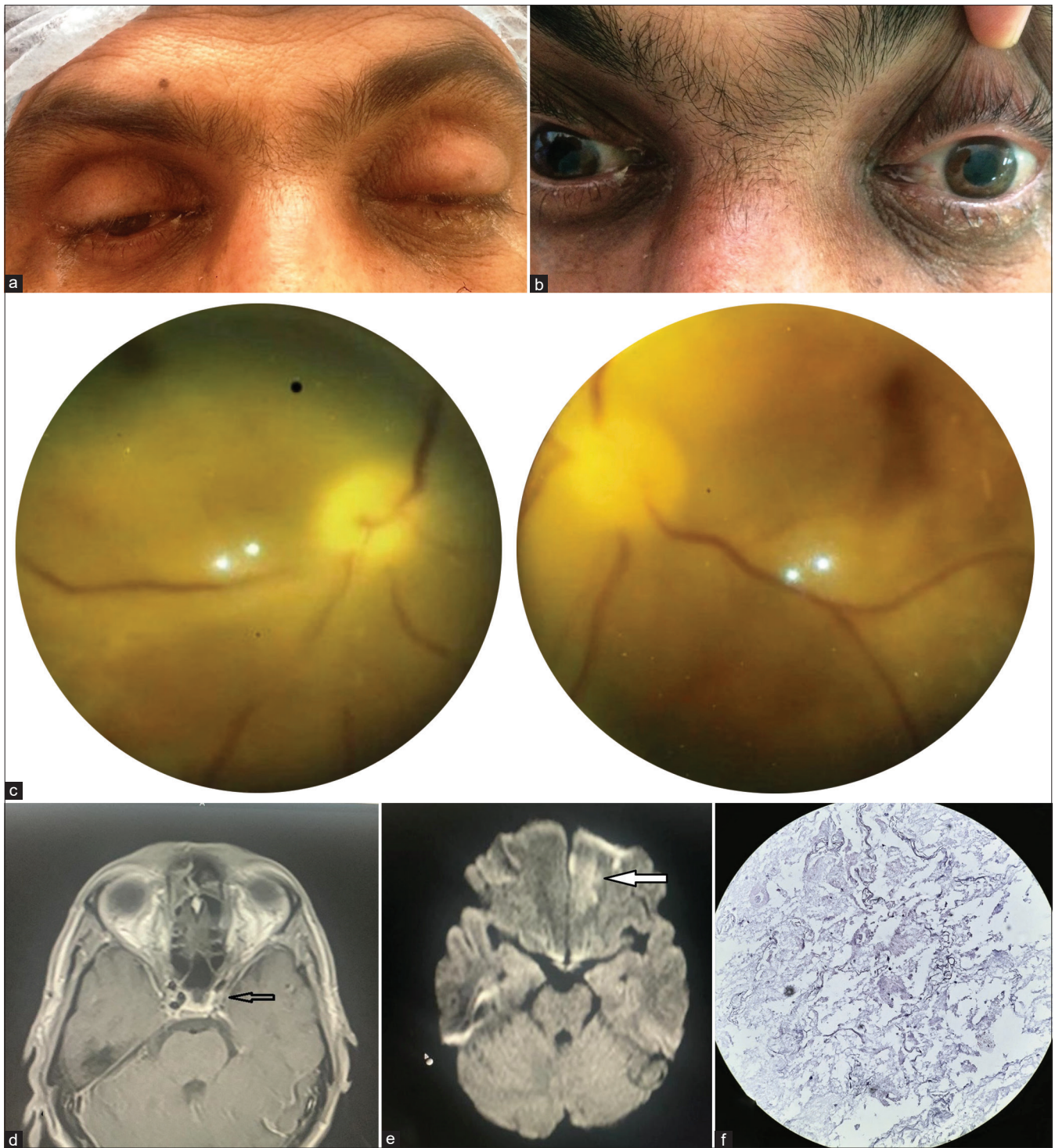


Figure 1: (a) Bilateral incomplete ptosis with fixed eyeballs. (b) Fixed and dilated pupil. (c) Bilateral CRAO. (d) MRI brain revealed bilateral opacification of both cavernous sinus (left > right) and mild narrowing of ICA (shown by arrow). (e) There was an area showing acute infarct in the left medial basifrontal region (shown by arrow). (f) Histopathological examination of sections from resected margin of optic nerve showed area of necrosis along with the presence of numerous broad aseptate, obtuse-angled branching hyphae invading the nerve bundles, thus confirming the diagnosis of mucormycosis

Discussion

Mucormycosis of orbit is a fulminant fungal infection associated with high morbidity and mortality. The common forms of mucormycosis are rhino-orbito-cerebral, cutaneous,

pulmonary, disseminated, gastrointestinal, and isolated renal. The infection has a predilection for immunosuppressed states like diabetic ketoacidosis, immunosuppressive therapy, thermal burns, chronic renal failure, cirrhosis, steroid or

cytotoxic therapy, hematological malignancies, malnutrition, solid organ transplantation, and HIV infection.^[2] It is scarcely ever reported in patients with healthy immune status.

Our patient had uncontrolled diabetes along with pyelonephritis, that may have predisposed him to ROCM. Secondly, due to compromised kidney status, standard treatment with liposomal amphotericin could not be started.

To the best of our knowledge, acute bilateral blindness in a young patient with ROCM is a rare manifestation. Malek *et al.*^[3] reported acute bilateral blindness in a young COVID-19 patient associated with ROCM. They noticed left cavernous sinus and internal carotid thrombosis along with CRAO in right eye. But in our case, a young patient had bilateral CRAO along with bilateral opacification of both cavernous sinus (left > right) and narrowing of left internal carotid artery. There was an area showing restricted diffusion on DWI and reduced ADC values on ADC mapping in the left medial basifrontal region consistent with acute infarct. Due to deterioration of systemic status, MRI could not be reported, which could have shown the status of the cavernous sinus at that moment.

CRAO is a rare manifestation of ROCM with incidence rate of 16–20%, resulting from direct angioinvasion. In the remaining 80%, it is thrombotic in nature. But bilateral CRAO is rarest in occurrence and this makes this case of a diabetic young male with compromised renal status developing acute bilateral blindness secondary to ROCM an alarming presentation.

Early diagnosis of sino-nasal mucormycosis, management of the predisposing factor, and prompt therapeutic interventions are critical for arresting intracranial spread of this fatal disease.^[4,5] A multimodality approach with intravenous antifungal, adequate sinus drainage and surgical debridement is warranted. However, compromised renal status was a limiting factor to start amphotericin in this case.

Conclusion

Successful treatment of ROCM warrants an early diagnosis of the infection, the control of underlying predisposing factors,

and aggressive and urgent surgery. Hence, ophthalmologists must have a high index of suspicion for development of mucormycosis in diabetic patients with associated co-morbidities.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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