



## ORBITAL APEX SYNDROME: FULMINANT PRESENTATION OF MUCORMYCOSIS

**Dr. Renuka Ajit  
Kukreja**

1<sup>st</sup> Year Postgraduate, M.S. Ophthalmology, Sree Balaji Medical College, Chennai

**Dr. Jigeesha Preethi  
Meda**

2<sup>nd</sup> Year Postgraduate, M.S. Ophthalmology, Sree Balaji Medical College, Chennai  
\*Corresponding Author

**ABSTRACT**

Increased incidence of fungal infection has been reported globally in the recent years. Fungal infection of the central nervous system remains one of the most difficult diseases to treat and requires multi-modality intensive therapeutic strategies. With associated co-morbidities like diabetes, aggravation of fungal infections is very common. Radiological studies are gold standard to assess disease extent and further management. Biopsy of the lesion and fungal staining help in identifying the etiological agent and final diagnosis. Rhino-orbito-cerebral mucormycosis is a major diagnostic dilemma with quick progression and a high mortality. Prompt medical management with a multi-modality approach can save the patient from orbital exenteration.

**Methodology:** We report two cases of patients who presented to us with sinusitis and total external ophthalmoplegia diagnosed with rhino-orbito-cerebral mucormycosis.

**KEYWORDS :** Mucormycosis, Uncontrolled diabetes

**Introduction**

Rhino-orbital-cerebral mucormycosis (ROCM) is an acute, often fatal, fungal infection caused by members of the class Zygomycetes and the order Mucorales. Angioinvasion with thrombosis and tissue necrosis is a key pathophysiological feature of human Mucorales infection.

The disease is characterized by fungal hyphal invasion of blood vessels resulting in thrombosis and infarction of the nasal, paranasal sinus, orbital, and cerebral tissues. Common clinical findings include rhinitis, periorbital and facial swelling, facial and mucosal necrosis, ophthalmoplegia, multiple cranial nerve palsies, facial pain, and headache. Orbital apex syndrome is an ominous complication of mucormycosis of the orbit. Once within the orbital compartment, organisms may extend posteriorly to the optic foramen, ophthalmic nerve and optic nerve are threatened by invasion, edema, inflammation and necrosis. Knowledge and identification of characteristic imaging patterns in fungal sinusitis help to refine the diagnosis, assess the disease extent, and plan management.

**Case-Report-1**

57 year old male patient was admitted with chief complaints of giddiness, facial puffiness and fever since 1 month and pain in the left eye and restricted extra-ocular movements in all directions for 5 days.

He was referred to our hospital for management as a case of left orbital apex syndrome due to cavernous sinus thrombosis.

Patient was a known case of diabetes and hypertension since 10 years.

On admission patient was unconscious and on general examination, proptosis of the left eye was found and black eschar was found in the nasal cavity.

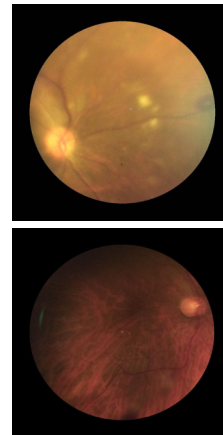
On ocular examination, left eye showed Ptosis, conjunctiva showed chemosis with a non reacting pupil and Proptosis of the left eye.

Intraocular Pressure in Right Eye was 10.2 and in the Left eye was 20.6.

On distant direct ophthalmoscopy the red glow was noted. On direct ophthalmoscopy, fundus looked uniform red. Right eye media looked clear, disc looked normal with a CD Ratio-0.3:1. Venous Dilatation and Multiple flame shaped haemorrhages were seen in all

quadrants, microaneurysms, dot and blot haemorrhages were seen mostly in the posterior pole. Hard exudates were present. Macula looked normal.

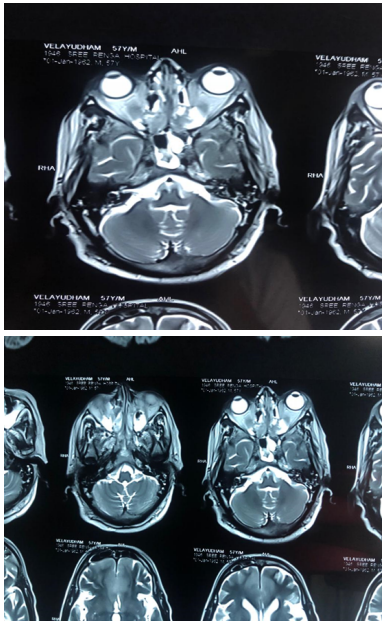
Left eye media looked clear, disc margins looked blurred and fundus shows multiple flame shaped haemorrhages, dot and blot Haemorrhages, Microaneurysms and Hard exudates in all quadrants. Cherry red spot Was present. Macula looked normal.

**MRI Brain Impression:**

- Chronic infarct with gliotic changes in periventricular left occipital and left frontal lobes.
- Acute to subacute non-hemorrhagic infarcts in bilateral basifrontal lobe and periventricular frontal lobe.
- Few lacunar infarcts in bilateral frontal lobes.
- Age related cerebral parenchymal changes.
- Thickening of mucosa of Bilateral Maxillary, ethmoidal and sphenoid sinuses.

**Left Orbit MRI Impression:**

- Proptosis of the left eye with minimal preseptal fluid.
- Left optic nerve appears hyper-intense with diffusion restriction.
- Retroconal intra-orbital space fat and sub-periosteal extraconal space along the medial orbital wall appears hyper-intense with diffusion restriction reaching upto orbital apex- Left orbital cellulitis with orbital apex syndrome
- Left cavernous sinus is enlarged with mild diffusion retraction compared with Right side- suggestive of cavernous sinus thrombosis.



**Case-Report 2**

A 68 year old male patient presented to ophthalmology Outpatient Department with complaints of Swelling in left eye, Mild pain, Redness, Inability to open eyes for 3 days.

Patient had no history of trauma/ fall/ prior swelling.

He was a known case of Diabetes Mellitus (poor control), Systemic Hypertension, CA colon on chemotherapy (Paclitaxel).

There was evidence of dental caries and maxillary sinusitis.

On General examination patient was Moderately built and nourished and systemic examination was normal. Decay was seen in left lower premolar tooth On ocular examination, Right Eye showed absent light reflex (direct and consensual) and Left Eye showed normal light reflex (only direct).

Extraocular Movements were restricted in the Right Eye and were normal in the Left Eye.

Visual acuity in the Right Eye was 6/60 and Left Eye was 6/24 and with Pinhole 6/12p.

Fundus examination was normal in both eyes.

**Investigations:**

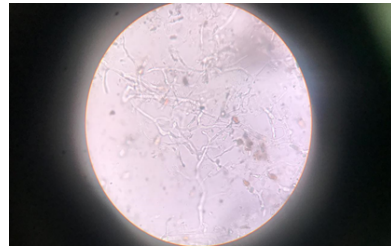
Blood work up –

Total Leucocyte count – 20,000 cells/mm<sup>3</sup>

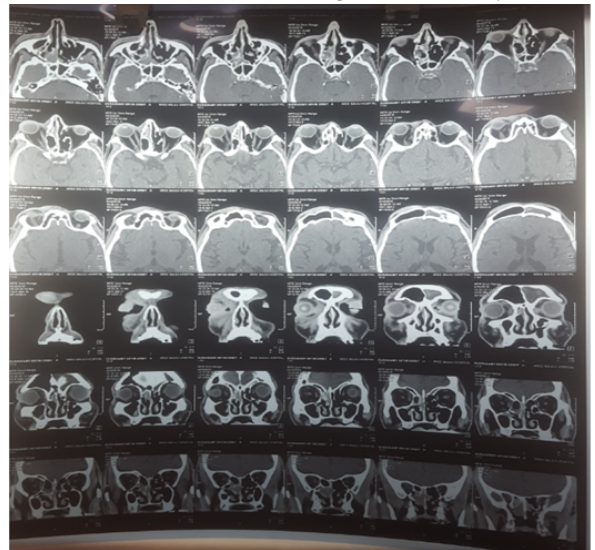
Fasting Blood Sugar – 312 mg/dl

Post Prandial Blood Sugar -300 mg/dl

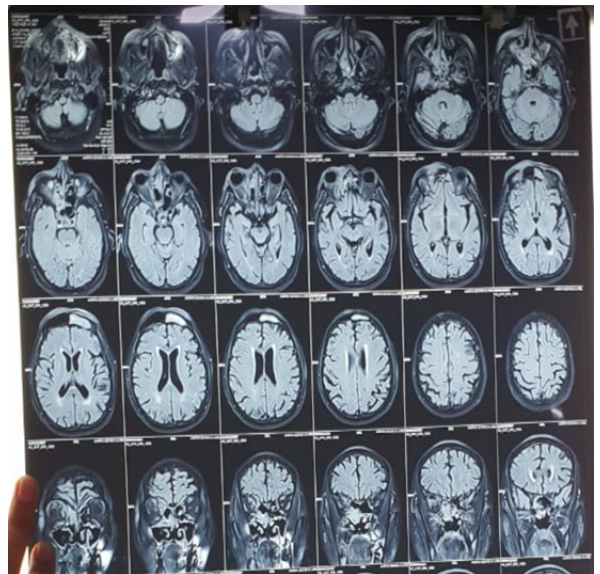
On diagnostic nasal endoscopy, black hard mass seen in ethmoid sinus region in right nasal cavity.



**CT Brain** showed orbital tissue swelling behind orbital septum.



**MRI Brain** showed mild soft tissue swelling and thickening of extra conal compartment of right orbit – suggestive of **ORBITAL CELLULITIS**



**Conclusion**

Early Diagnosis and treatment of the fungal maxillary sinusitis can limit the spread of infection and shall ultimately prevent complications. Screening of Diabetic patients with sinusitis should be done regularly and the progression of the disease should be monitored with serial Radiological investigations and patients should be treated with amphotericin B and required surgical interventions.

**REFERENCES**

1. Spellberg B, Edwards J, Ibrahim A. Novel perspectives on mucormycosis: pathophysiology, presentation, and management. Clin Microbiol Rev. 2005;18:556–569. [PMC free article] [PubMed] [Google Scholar]
2. Roden MM, Zaoutis TE, Buchanan WL, Knudsen TA, Sarkisova TA, Schaufele R.

- Epidemiology and outcome of mucormycosis: a review of 929 reported cases. *Clin Infect Dis.* 2005;41:634–653. [PubMed] [Google Scholar]
3. Gregory JL, Golden A, Haymaher W, Suresh V, Gupta A, Singh P. Presentation and outcome of rhino-orbital-cerebral mucormycosis in patients with diabetes. *Postgrad Med J.* 2004;80:670–674. [PMC free article] [PubMed] [Google Scholar]
  4. Brian M, O'Neill DDS, Alessi AS, George EB, Piro J. Disseminated rhinocerebral mucormycosis. *J Oral Maxillofac Surg.* 2006;64:326–333. Paultauf, A; mycosis mucorina. *Virchows Arch* 1885 10243, cited. [PubMed] [Google Scholar]
  5. Marty FM, Cosimi LA, Baden LR. Breakthrough zygomycosis after voriconazole treatment in recipients of hematopoietic stem-cell transplants. *N Engl J Med.* 2004;350:950–952. [PubMed] [Google Scholar]
  6. Kontoyiannis DP, Lionakis MS, Lewis RE, Chamilos G, Healy M, Perego C. Zygomycosis in a tertiary-care cancer center in the era of aspergillus-active antifungal therapy: a case-control observational study of 27 recent cases. *J Infect Dis.* 2005;191:1350–1360. [PubMed] [Google Scholar]
  7. Bitar D, Van Cauteren D, Lanternier F, Dannaoui E, Che D, Dromer F. Increasing incidence of zygomycosis (mucormycosis), France, 1997–2006. *Emerg. Infect. Dis.* 2009;15:1395–1401. [PMC free article] [PubMed] [Google Scholar]
  8. Chow V, Khan S, Balogun A, Mitchell D, Mühlischlegel FA. Invasive rhino-orbital-cerebral mucormycosis in a diabetic patient – the need for prompt treatment. *Med Mycol Case Rep.* 2015;8:5–9. [PMC free article] [PubMed] [Google Scholar]
  9. Nithyanandam S, Jacob MS, Battu RR, Thomas RK, Correa MA, D'Souza O. Rhinorbito-cerebral mucormycosis: a retrospective analysis of clinical features and treatment outcomes. *Indian J Ophthalmol.* 2003;51:231–236. [PubMed] [Google Scholar]