

Rhino-Orbital Cerebral Mucormycosis with Cavernous Sinus Thrombosis with Ophthalmoplegia – A Case Report

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Abstract: Rhino-orbital-cerebral mucormycosis is a potentially lethal, opportunistic fungal infection, rapid progression, unpredictable course and high mortality. Cavernous sinus thrombosis with ophthalmoplegia is presented. We present a case of a 47 year old female with left sided facial numbness, left eye proptosis and a chronic ulcer in the hard palate with black eschar in its floor within a period of 4 months. Later she developed severe headache, loss of vision left eye, drowsiness. The typical clinical features, imaging and treatment are highlighted in the report. Awareness of its occurrence, prompt diagnosis and appropriate management can reduce the mortality.

Keywords: ROCM, MRI, Diabetes, CT

Date of Submission: 06-07-2019

Date of acceptance: 20-07-2019

I. Introduction

The first description of mucormycosis was given by Meyer in 1815^(1,2). Mucormycosis represents a group of life threatening, rare opportunistic infections caused by fungi of the order Mucorales. It is highly invasive and relentlessly progressive. These are ubiquitous organisms, existing in the environment, soil, air, food, composite piles¹⁻⁵. These fungi cause infections primarily in immunocompromised, diabetes and on steroid treatment. Clinically mucormycosis may appear in, rhino orbital cerebral (44-49%), cutaneous (10-16%), pulmonary (10-11%), disseminated (6-11%), gastrointestinal forms (2-11%)¹. It is an invasive fungal infection initiated in the paranasal sinuses and frequently progress to involve the orbit and brain. It is a medical emergency and management comprises of treating the predisposing factors, radical surgical debridement and effective systemic antifungal therapy. One possible intracranial complication of mucormycosis is cavernous sinus thrombosis. Initial symptoms include headache, retro-orbital pain, peri-orbital edema, proptosis, diplopia. The first description of affection was made by Duncan in 1821²

II. Case report

A 47 year old female living in rural area presented to the casualty with a history of chronic non healing ulcer in the hard palate on left side and left sided facial numbness with signs of orbital involvement (extensive proptosis, periorbital swelling and loss of vision left eye) from 4 months. She was suffering from nausea, vomiting, frontal headache and left eye burning sensation and was drowsy. The lesion started with sudden onset of left sided facial numbness with darkening of skin on left side face and mouth deviation to right 4 months back. Discomfort continued to increase and patient developed a black coloured patch on the anterior part of hard palate left side which gradually increased to form an ulcer. Patient visited a local doctor where she received medical treatment for diabetes and was put on intravenous antibiotics and underwent endoscopic medial maxillectomy. Her condition deteriorated with increased ocular discomfort, sloughing of ulcer and loss of vision left eye. No history of difficulty swallowing, loss of taste. Co-morbid includes hypertension for

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which she was regularly taking tab telmesartan 40mg, Type 2 diabetes for which she was non-compliant to treatment, known case of hypothyroidism and was on thyroxine 125mg.

Examination showed a pale dehydrated patient, general condition was sick, was following verbal commands, GCS -8. Left eye proptosis and periorbital oedema was seen(Fig 1).On examination left eye pupil was found fixed and dilated with palsy of cranial nerve third, fourth and sixth. Vision was lost in left eye. Also there was loss of sensation on left side face. On local examination of nose- external nose – skin over nasal bridge, dorsum and both ala was found normal. Loss of left nasolabial angle was seen. On anterior rhinoscopy, nasal mucosa was pale, with septum midline, roof, floor anteriorly and lateral wall was normal. A defect was seen in the floor of nose just in front of the posterior nares. Defect was 1x1cm sized covered with white and black colored slough. On throat examination, mouth seen deviated to right, lips were dry and lower jaw teeth on left were found missing. Tongue and buccal mucosa were normal. A defect was seen on the hard palate 5x5 cm sized on left side, rectangle in shape, extending from the midline raphe till left side involving palatine rugae, mucosa around the ulcer appeared oedematous, floor of ulcer was covered with yellow black colored eschar, edges were smooth and inverted. (Fig2).Oropharynx and posterior pharyngeal wall was normal.

Laboratory investigations revealed white blood cells 12000 cells/cumm., haemoglobin 11gm/dl, random blood glucose levels were 260mg/dl and fasting blood sugar 140gm/dl, patient was human immunodeficiency virus and hepatitis B surface antigen negative, pH was 7.5, liver and kidney function tests were within range, bicarbonate levels, magnesium levels were normal, urine routine microscopy was positive for ketone bodies, urine was ++ for sugar, ecg was normal, neurophysician reference was also done. CT scan of paranasal sinuses showed soft tissue filling left frontal sinus, left ethmoidal air cells and bilateral sphenoid sinuses, permeative bone destruction seen in floor of left middle cranial fossa. MRI brain was done, which showed increased soft tissue component in the region of left maxillary sinus showing T2 hyperintense signal intensity. Loss of flow void in cavernous part of internal carotid artery with cavernous sinus inversion. Left proptosis with soft tissue edema, bilateral mastoiditis, mildly increased mucosal thickening of paranasal sinuses. MRI findings were consistent with cavernous sinus thrombosis.(Fig 3).Patient was carrying a biopsy report from outside which was positive for mucormycosis. Surgical debridement of necrotic tissue in ulcer was done(Fig 4).Patient was started on injection piptaz 4.5g every 8 hourly and injection Clindamycin 600mg/day in two divided dose. Injection Amphotericin B 250mg daily and suspension posaconazole were started.Patient was on thyronorm 125 microgram, telma-40, eyedrops moxi, injection emeset, tablet naxdon, injection of regular insulin and intravenous fluids were given. Patient was later shifted to tablet metformin. Patient was stable at the time of discharge and was lost to follow up.

III. Discussion

Mucormycosis represents a group of life threatening infections caused by fungi of order Mucorales. Among Mucorales, Rhizopus oryzae is the most common cause of infection. Increasing number of cases of mucormycosis due to infection with Cunninghamella species have been reported, particularly in highly immunocompromised patients. It represents 8-13% of all fungal infections found at autopsy. These fungi primarily cause infection in patients of diabetes or those with neutropenia or steroid treatment.³ It occurs as a cutaneous or subcutaneous infection after trauma, implantation of soil or vegetation. Mucormycosis is an emerging angioinvasive infection. Mucormycosis has emerged as the third most common invasive mycosis in order of importance in patients with hematological and allogenic stem cell transplantation, diabetics or defect in phagocytic function.It is believed that in most cases fungi enter the body by inhalation of aerosolised spores through the sinuses with infiltration and spread along neurovascular structures Ubiquitous in nature, found colonising mucosal surfaces of oral and nasal cavities. Seasonal variation is possible. The clinical progression of ROCM is classified into 3 stages – stage 1 – infection of nasal mucosa and paranasal sinuses; stage 2 – orbital involvement leading to superior orbital fissure syndrome and orbital apex syndrome; stage 3 – cerebral involvement to cavernous sinus thrombosis, occipital and frontal lobe infarctions.⁴

Radiological investigations are necessary for proper diagnosis. Main radiological investigation is CT⁵. Detection of aseptate hyphae with right angled branching is pathognomic of mucormycosis. Medical management alone is not effective because of poor drug delivery to the site of infection due to extensive vascular thrombosis.

Factors associated with poor survival in ROCM include delay in diagnosis and treatment, hemiparesis, bilateral sinus involvement and facial necrosis. Incases suspicious of mucormycosis, treatment must be rapidly and aggressively started, with surgical procedure for removal of areas of necrosis in nasal mucosa and utilisation of intravenous systemic antifungal agents. Even so the mortality and morbidity rates are high.Klebsiella

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rhinoscleromatis is a rare cause of an indolent facial rhinoscleroma syndrome that appears similar to mucormycosis. Tolosa Hunt Syndrome causes painful ophthalmoplegia, ptosis, headache and cavernous sinus inflammations; biopsy and clinical follow up may be needed to distinguish syndrome from mucormycosis⁶. Mucormycosis infection can progress rapidly and erode through the bony walls of sinuses into orbit, skull base and into brain. Thrombosis of cavernous sinus, carotid arteries indicate a very poor prognosis.⁷. Adjunctive therapy for mucormycosis in literature suggests use of hyperbaric oxygen therapy and iron chelating agent that improve prognosis of this disease. Hyperbaric oxygen inhibits fungal growth and promotes action of amphotericin B.

IV. Conclusion

This report has presented classical clinical features of cavernous sinus thrombosis and rhino-orbital-cerebral mucormycosis. A high index of suspicion is required for early diagnosis and treatment of mucormycosis to prevent devastating complications as fatality can exceed 80%. Early diagnosis with antifungal therapy with surgical debridement is done. In patients with intracranial involvement, fatality rates can exceed 80%.

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Fig 1- Left eye proptosis, periorbital edema



Fig 2- Defect in the hard palate covered with black colored slough

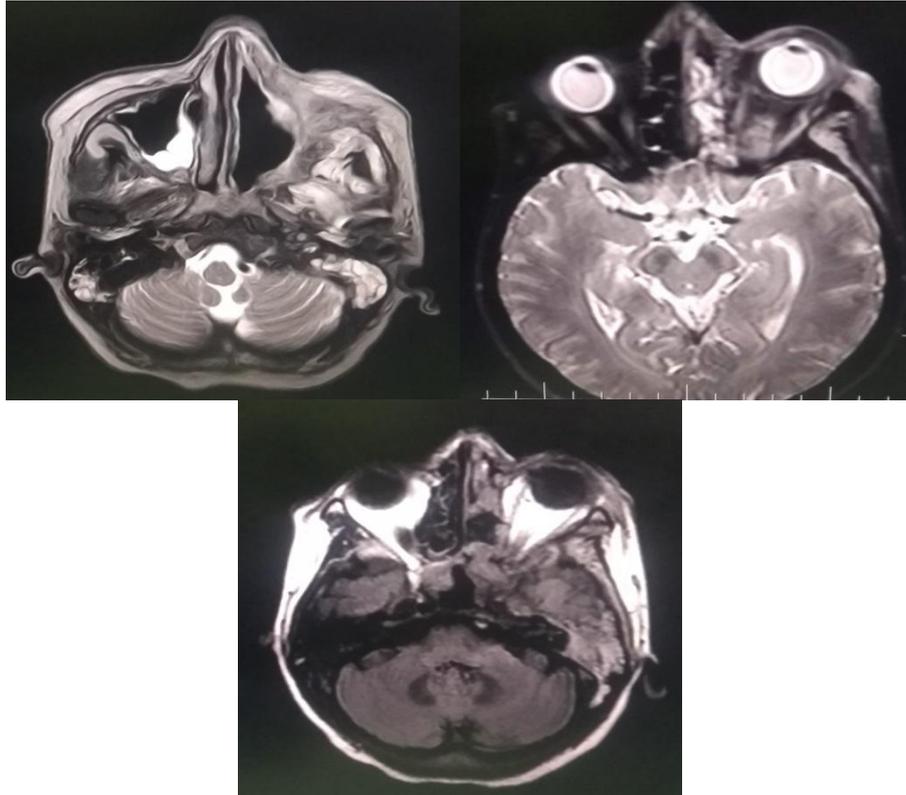


Fig 3. MRI shows increased soft tissue component in the region of left maxillary sinus showing T2 hyper intense signal intensity. Loss of flow void in cavernous part of internal carotid artery with cavernous sinus inversion. Left proptosis with soft tissue oedema



Fig 4. After surgical debridement

Dr. Vivek Kumar Pathak. "Rhino-Orbital Cerebral Mucormycosis with Cavernous Sinus Thrombosis with Ophthalmoplegia – A Case Report." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 18, no. 7, 2019, pp 50-53.