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A Journey Through Orbital Mucormycosis

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ABSTRACT

A total of 68 patients who had orbital symptoms of mucormycosis underwent MR imaging. Nearly all patients with orbital symptoms had associated rhinosinusitis. 18 patients had associated cerebral involvement. 8 patients had associated skull base osteomyelitis. Of all the imaging features, orbital cellulitis was the most common imaging finding. 6 patients had ischemic optic neuritis, and 8 had cavernous sinus thrombosis. 14 patients had associated superior ophthalmic vein thrombosis. 7 patients had orbital compartment syndrome, and 12 had orbital apex syndrome. One of the most dreaded complications of craniofacial mucormycosis is the rapid onset of blindness. This article explores the various orbital manifestations of mucormycosis, its complications, and the ideal MRI protocol to detect such complications.
Keywords: Mucormycosis, Rhinosinusitis, Orbital cellulitis.

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INTRODUCTION

During the second wave of COVID-19 in India, there was an exponential rise in the rates of infections with mucormycosis, especially craniofacial mucormycosis. [1]Before COVID, most cases of mucormycosis were due to an inherent immunocompromised state, mainly arising due to diabetes and steroid use in transplant patients. [2]Due to the rapidity of the development of complications, it is vital that imaging be performed at the earliest onset of symptoms. Imaging is crucial in mapping the disease extent, demonstrating involvement of the orbits, involvement of the optic nerve, anterior or middle cranial fossa, and skull base, which plays a major role in treatment planning. [3]Imaging may also be used as an indicator in assessing treatment response.MRI is the primary imaging modality in the assessment of orbital involvement, with better resolution of soft tissues and retro-orbital planes. CT may also help in supplementing the MR findings by better depicting bone erosions.[4,5]

MATERIALS AND METHODS

We evaluated 68 patients with biopsy/culture-proven mucormycosis who had undergone MR imaging for ophthalmic symptoms of mucormycosis. 57 patients were diabetic, and 49 patients had recovered from COVID pneumonia. All patients underwent MR imaging in a 1.5T Siemens Magnetom Amria Tim+ system with routine T1, T2, STIR, T1FS, T2FS, T1 VIBE, and post-contrast T1FS sequences. Contrast used was Gadopentetic dimeglumine (0.1mmol/kg). Senior radiologists interpreted MR images to determine the involvement of the sinuses, orbit and its contents, cerebral involvement, and skull base signal changes.

RESULTS AND DISCUSSION

All patients had sinusitis and ophthalmological symptoms. 18 patients had associated cerebral involvement in the form of cerebritis or meningitis. 8 patients had associated skull base osteomyelitis. Of all the imaging features, orbital cellulitis was the most common imaging finding. 6 patients had ischemic optic neuropathy, and 8 had cavernous sinus thrombosis. 14 patients had associated superior ophthalmic vein thrombosis. 7 patients had orbital compartment syndrome, and 12 had orbital apex syndrome.

Table 1: Pateints Manifestations

Disease Extent (A Total Of 68 Patients Were Evaluated)	No Patients With Manifestation
Rhino sinusitis with preseptal alone	6
Rhino sinusitis with orbital cellulitis	41
Rhino-orbital cerebral mucormycosis (ROCM)	18
Ischemic optic neuropathy	6

Imaging Findings

Intra-orbital extension of mucormycosis is through the lamina papyracea from the ethmoid sinus, through the nasolacrimal duct. Involvement of the post-septal soft tissue components is denoted orbital cellulitis. Imaging findings ranged from bulky oedematous extraocular muscles, enhancing optic nerves, and soft tissue components at the orbital apex to cavernous sinus thrombosis.

Extra Conal Involvement

The medial rectus thickens and undergoes lateral displacement when the infection spreads from the ethmoid sinus through the medial wall of the orbit. There is sometimes associated subperiosteal abscess collection due to the contiguous spread of the infection from the maxillary sinus through the orbital floor. There is a thickening of the inferior rectus with a mild subperiosteal abscess collection at the floor of the right orbit. There is also severe inflammation of the temporalis muscle and the infratemporal fossa.

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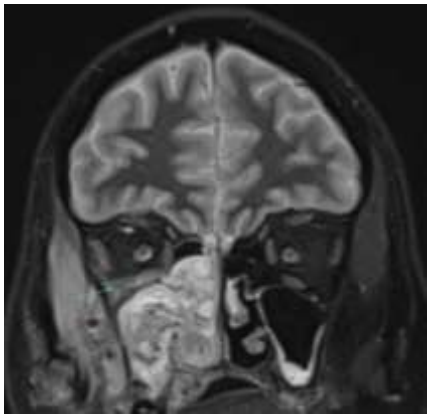

Figure 1

Intraconal Involvement

Imaging features of mucormycosis are a result of the angio-invasive nature of the fungal hyphae, which invade the blood vessels, causing necrotizing vasculitis and infarction.

Orbital cellulitis shows soft tissue involvement posterior to the septum with symptoms of proptosis, periorbital oedema, and decreased vision. Sometimes the orbital cellulitis is so severe that the necrosed tissues form an abscess or an inflammatory phlegmon.

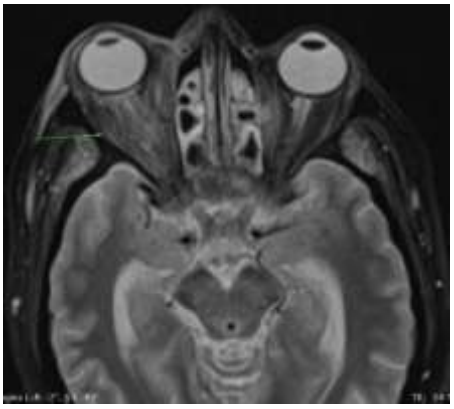

Figure 2: Right proptosis.

Soft tissue components, fat stranding, and muscle thickening were noted in the retrobulbar compartment.

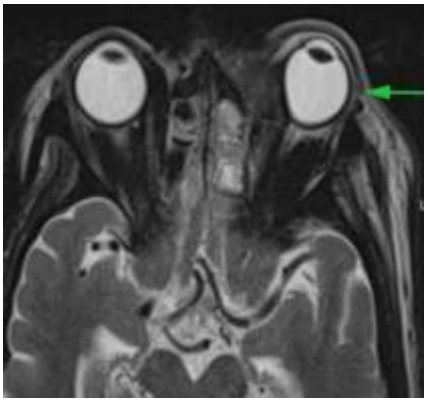

Figure 3: Left eye proptosis.

Tenting of the posterior globe – 'Guitar pick sign' seen in the orbital compartment

Orbital Compartment Syndrome

It is an emergency. Orbit is a closed space, limited by its bony walls. An increase in intra-orbital pressure can cause a pressure effect that can compromise blood supply to the optic nerve and also the globe. Symptoms can range from proptosis to decreased vision and increased intraocular pressure. Emergency orbital decompression is required in such cases as increased intra-orbital pressure, for as little as less than 2 hours can cause blindness[1]. It can manifest as posterior tenting of the globe in the shape of a "guitar pick" and stretching of the optic nerve.

C.Orbital Apex Syndrome

Orbital apex syndrome (OAS) is an uncommon complication arising from orbital mucormycosis, comprising complete ophthalmoplegia with vision loss, involving cranial nerves II, III, IV, V_1, and VI. [2] OAS occurs due to compression of crucial structures at the orbital apex and can also occur due to trauma, malignancy, and infection/ inflammation due to other causes.

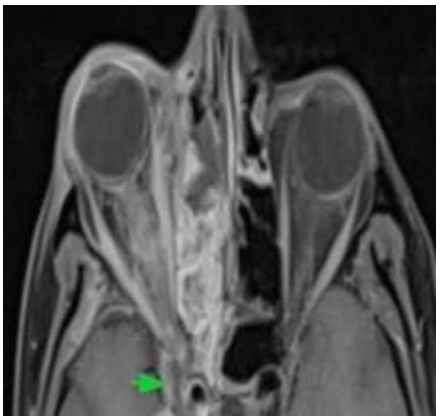

Figure 4: Right proptosis.

Involvement of the right orbital apex. Intra-cranial extension into the cavernous sinus. Another complication of the orbital apex syndrome is that there can be contiguous spread of the infection from the orbital apex into the cavernous sinus, causing cavernous sinus thrombosis.

Ischemic Optic Neuropathy

It is an infrequent complication of orbital mucormycosis. Occur due to necrotizing vasculitis caused by the angio-invasive hyphae of the fungi. Ischemic optic neuropathy is characterized by diffusion restriction along the optic nerves with low ADC, normal FLAIR, and no post-contrast enhancement of the optic nerve sheath complex. [3]

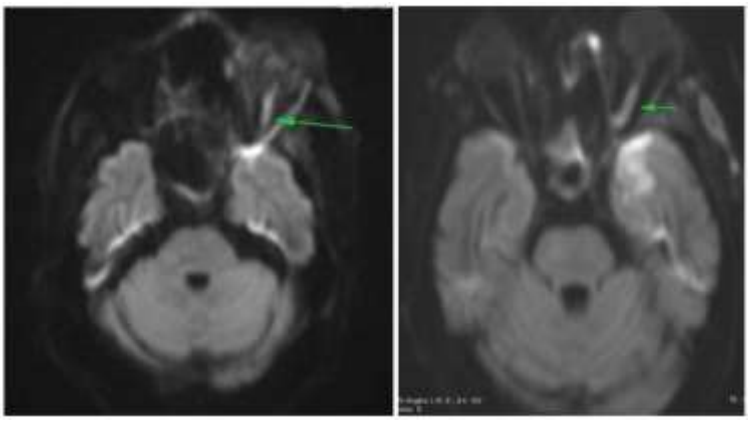

Figure 5, 6

MRI shows diffusion restriction along the optic nerves, a finding consistent with ischemic optic nerve infarction.

Cavernous Sinus Thrombosis.

Cavernous sinus thrombosis caused by rhino-orbital mucormycosis causes visual loss earlier than bacterial causes. It can occur as a result of the contiguous spread of infection from the orbit extending posteriorly. Plain MR imaging shows loss of the normal concavity of the lateral walls of the cavernous sinus, with bulging walls indicating cavernous sinus thrombosis.

C+T1W FS imaging shows a filling defect in the left cavernous sinus. It is also important to carefully examine the cavernous segment of the internal carotid artery.

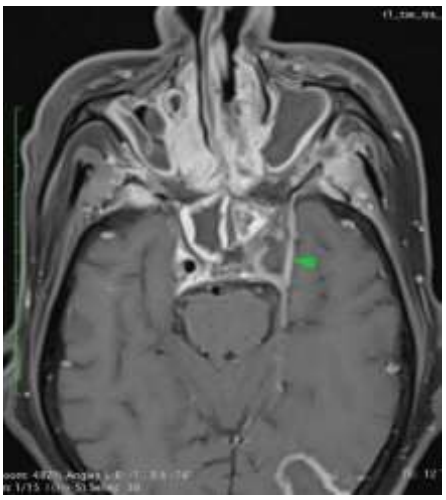

Figure 7

DILATED SUPERIOR OPHTHALMIC VEIN

The dilated superior ophthalmic vein can occur due to cavernous sinus thrombosis or compression at the orbital apex. It can also be dilated secondary to thromboses caused by orbital cellulitis from mucormycosis. Symptoms are mainly due to impaired orbital venous drainage. It can cause orbital swelling, proptosis, and limited ocular motility [4]. Patients with SOVT require urgent antibiotic therapy and sinus surgery. It may also extend intracranially, resulting in stroke, and is often life-threatening.

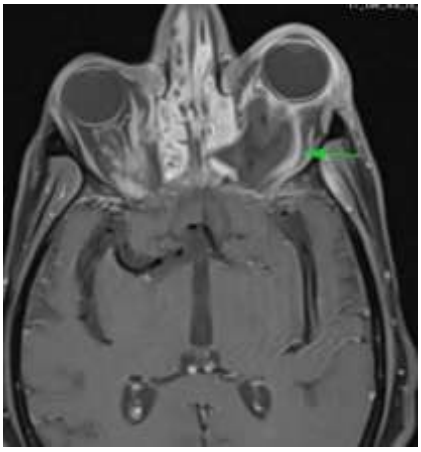

Figure 8: Dilated superior ophthalmic vein secondary to cavernous sinus thrombosis.

Endophthalmitis

Intraocular infection due to mucormycosis is rare, but it may occur due to intra-arterial spread[5]. Scleral necrosis with associated peripheral corneal melt, low IOP, and loss of vision may suggest endophthalmitis. Loss of vision may be attributed to thrombosis of retinal, choroidal, or ophthalmic vessels. Acidic pH and hyperglycaemia-induced immunocompromised state make more free iron available by disrupting the binding with transferrin. As mucor is a ferro-philic organism, this event increases the risk of infection with mucormycosis [6]. Angioinvasion leads to thrombosis & haemorrhage, resulting in tissue infarction and necrosis [7]. In the diagnosis of mucormycosis, histopathological examination is considered more sensitive than cultures.

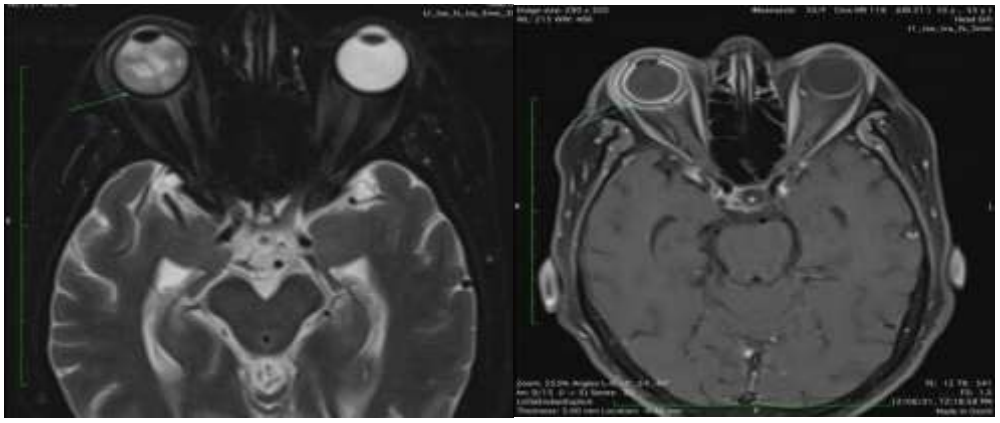

Figure 9,10

Intracranial Extension

Intracranial extension in mucormycosis commonly occurs via direct spread across the cribriform plate of the ethmoid and frontal sinuses. Extensions into the middle cranial fossa are a spread of infection from the pterygopalatine fossa.[8] In addition to direct spread, there is also perineural spread from the cavernous sinus along the trigeminal nerve, which can lead to predominant posterior fossa involvement. Early intracranial spread is seen in contrast-enhanced T1W images when there is leptomeningeal or pachymeningeal enhancement.[9] Other intracranial complications of mucormycosis include abscesses and infarcts. Cerebritis appears as ill-defined areas of altered signal intensity, usually T2 hyperintensity, in nonvascular distribution. Minimal perilesional edema and variable peripheral enhancement are present. They can progress into the abscess with a well-delineated mass with a liquified central T2 hyperintense core showing diffusion restriction. Abscesses in ROCM may not show the characteristic well-defined rim enhancement seen in bacterial abscesses because of poor immunogenic response by a compromised host. (10)

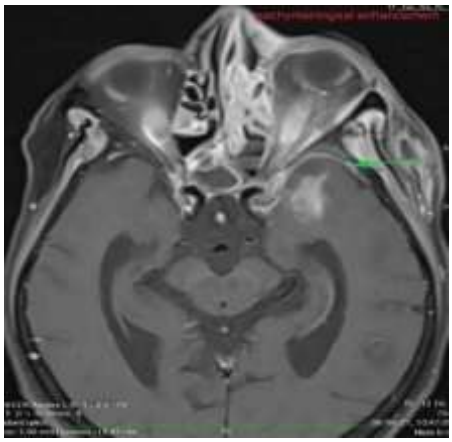

Figure 11

There is pachymeningeal enhancement along the left temporal lobe, with an ill-defined cerebritis seen involving the left temporal lobe.

Skull Base Involvement

Skull base osteomyelitis is a rare complication, usually seen in the late stages of the disease. [11,12] It occurs due to the spread of infection into the deep soft tissues through the perivascular channels even before actual bone destruction.[12] Skull base osteomyelitis causes loss of fat signal in T1W images. The marrow appears hypointense on T1W images, T2 hyperintense, with hyperintensity also seen on STIR images, with postcontrast images showing heterogeneous enhancement.[13] In the advanced stage, there is extensive heterogeneously enhancing soft tissue with infiltration into the bones. Abscess formation can occur in the adjacent soft tissues, appearing as a fluid signal intensity area with central diffusion restriction and peripheral rim enhancement.

MRI For Prognostication

The presence of imaging features of severe disease, such as involvement of the brain or orbit, florid involvement of sinuses, bilateral sinus involvement, and involvement of the palate, has been associated with poor prognosis.[11,12] While some series did not find orbital involvement to worsen the survival,[6] several other studies found that orbital involvement was a factor predicting poor survival.[3] A few recent studies [8,11,12] found that the absence of contrast enhancement in the infected soft tissues predicted poor survival. More prospective studies with a correlation of the clinical outcomes with imaging features documented using a structured reporting format are needed to confirm the role of imaging in the prognostication of mucormycosis.

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