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Original Article

Imaging features of rhinocerebral mucormycosis: A study of 43 patients

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ABSTRACT

Background: Rhinocerebral mucormycosis is a life-threatening infection caused by saprophytic fungi seen almost exclusively in immunocompromised patients.The objective of this study was to describe the imaging findings in patients with rhinocerebral mucormycosis.

Materials and methods: The case records of patients with biopsy/culture proven invasive rhinocerebral mucormycosis were reviewed. Computed Tomography (CT) and/or Magnetic Resonance Imaging (MRI) images were retrieved from the Picture Archiving and Communication System (PACS) and analyzed. Statistical analysis was performed using descriptive statistics.

Results: CT and MR imaging of 43 patients showed predominant involvement of the ethmoid (37, 86%) and maxillary (34, 79%) sinuses. Extension to the orbit (32, 76%) and face (24, 57%) preceded involvement of the deep skull base (5, 12%) and brain (13, 31%). CT showed minimally enhancing hypodense soft tissue thickening as the predominant finding in involved areas, while MRI showed T2 isointense to mildly hypointense soft tissue thickening and heterogeneous post contrast enhancement as the main finding. Bone erosion was seen less often (17, 40%), with rest (26, 60%) of the patients showing extrasinus extension across grossly intact appearing bones on imaging.

Conclusion: CT and MRI shows a spectrum of findings in rhinocerebral mucormycosis. Imaging plays a major role in assessing the extent of involvement and complications.

1. Background

Rhinocerebral mucormycosis is a life-threatening infection caused by saprophytic fungi belonging to the genera *Mucor*, *Rhizopus* and *Absidia* [1,2]. All of these belong to the order *Mucorales* and class *Zygomycetes* [3]. The disease is seen almost exclusively in immunocompromised patients since normal phagocytic activity in immunocompetent hosts provide an adequate barrier against infection [1,2]. The clinical presentation in the early stages is typically with fever, headache, facial pain, nasal discharge, nasal obstruction and crusting. The disease progresses rapidly within a period of a few hours to days leading to cranial nerve palsies and features of CNS involvement [1]. Early imaging is helpful in assessing the extent of involvement of this lethal disease which requires prompt and aggressive treatment. In order to study the imaging findings in patients who were diagnosed with rhinocerebral mucormycosis who presented to us at various stages of the disease, we performed a retrospective study of all patients with this diagnosis who presented to our hospital. We also studied the imaging findings of those patients who reported for follow up after successful therapy.

2. Materials and methods

The case records of patients with culture and histological evidence of acute invasive rhinocerebral mucormycosis were retrospectively evaluated for relevant clinical data. The Computed Tomography (CT) and/or Magnetic Resonance Imaging (MRI) images were retrieved from the Picture Archiving and Communication System (PACS) (GE Healthcare, Milwaukee, Wisconsin). CT scan was performed on a Siemens (Emotion 16 slice Siemens Medical system, Erlangen, Germany) or Philips (Brilliance 6 slice Philips Medical system, Best, the Netherlands) machine using a routine CT Paranasal sinus (PNS) protocol with 130kVP and 150–220 mA tube current. Intravenous contrast medium (low osmolar, non-ionic, 300 mg/mL iodine content) was used

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routinely at a dose of 1 ml/kg administered by pressure injector. Conventional MR Neck imaging including axial, coronal and sagittal T1 weighted (TR/TE 40/12 ms), T2- weighted images (TR/TE 4000/100 ms) and fat suppressed post contrast T1 weighted images were acquired. MRI imaging was performed using 1.5 T Siemens (Avanto: Siemens Medical system, Erlangen, Germany) or 3 T Philips (Achieva: Philips Medical system, Best, the Netherlands).

Image analysis- Retrospective review by consensus was performed by 3 staff radiologists to look for sites and extent of involvement, signal characteristics and complications.

2.1. Image interpretation

The sinuses showing opacification on CT or MRI were recorded in each case. The appearance on plain T1 and T2 images were documented. On post contrast CT and MRI, the type of contrast enhancement and involvement of any extra sinus structures including orbit, face, pterygopalatine fossa, masticator space, brain and cavernous sinus were noted. Presence of bone involvement was evaluated on the CT. Any complications like arterial thrombosis was noted on CT and MRI.

Fat stranding and soft tissue extension similar in appearance to the intrasinus soft tissue was taken as evidence of orbital, retroantral, masticator and pterygopalatine involvement. Orbital cellulitis was seen as stranding in the retrobulbar fat, without overt abscess formation. Cavernous sinus and internal carotid artery involvement was seen as thickening and non enhancement on post contrast scans with presence of abnormal surrounding soft tissue. Patients with intracranial extension were evaluated for dural enhancement, presence of extradural collections, infarcts, cerebritis and intracerebral abscess.

As patients presented to us at various stages of the disease, the patients in the study were divided into three groups based on the extent of regional involvement according to classification suggested by Rupa et al. Table 1 [4].

2.2. IRB clearance

Institutional ethics committee clearance was obtained for the conduct of this study.

2.3. Statistical analysis

Descriptive statistical methods were used for statistical analysis.

3. Results

A total of 43 patients with rhinocerebral mucormycosis having preoperative contrast CT and/or MRI imaging and histopathology confirmation were identified. All 43 patients underwent contrast-enhanced CT examination while 19 patients also had MRI. Out of the 19 patients who had MRI, only 14 underwent contrast imaging with gadolinium, primarily because of inability to use contrast in those patients with impaired renal function.

Table 1
Extent of regional involvement.

Stage	Areas involved	Number (%)
Stage 1	Nose & paranasal sinuses alone	2 (4.7)
Stage 2	Paranasal sinuses with immediate adjacent areas which are surgically resectable with minimal morbidity eg. orbit (extraconal), palate & oral cavity	16 (37.3)
Stage 3	Intracranial extension (extradural/intracerebral) or partially resectable with extension to pterygopalatine fossa, cavernous sinus, cheek and periorbital region	25 (58.1)

3.1. Demographic and clinical findings

Our study group comprised of 30 males and 13 females with ages ranging from 2 to 75 years (mean = 55 years). The majority of patients (83.7%) were aged over 40 years, with those aged 40–60 years (55%) being most affected.

Thirty nine patients (91%) had a history of uncontrolled diabetes, with 38 patients (88.4%) having type 2 diabetes mellitus and 1 patient with type 1 diabetes mellitus. Four patients (9.3%) had a predisposing haematological condition, one patient each with acute myeloid leukemia, chronic myeloid leukemia, acute lymphoblastic leukemia and post bone marrow transplant status for aplastic anemia.

The clinical symptoms reported in our series were headache (38, 88%), nasal discharge (30, 69%), facial swelling (30, 69% patients), facial pain (28, 65%), decreased vision (9, 21%), fever (6, 14%), and epistaxis (4, 9%). Eleven patients (25%) presented with cranial nerve palsy with 6 patients having multiple cranial nerve palsies at presentation.

3.2. Imaging findings

(a) Sinonasal involvement

The ethmoid sinus was the most common paranasal sinus involved in our study (37, 86%). In the majority of patients (34, 79%) multiple sinuses were involved. The combination of maxillary, ethmoid and sphenoid (21, 49%) was most frequently seen. Unilateral sinus involvement was more common (79.1%) than bilateral sinus involvement (20.9%). The sinuses involved in mucormycosis is detailed in Table 2.

(b) Signal characteristics and imaging appearances

CT

On CT three types of contrast enhancement were seen, with mild enhancement being the most common form. Other types included low density opacification with no post contrast enhancement and heterogeneously enhancing intrasinus abscess like appearance with variable enhancing and non enhancing areas. The various enhancement types are shown in Table 3. In cases with non-enhancing opacification of sinuses, presence of retroantral, facial and orbital fat stranding and hypodense soft tissue extension indicated the aggressive nature of the infection (Fig. 1). Surrounding walls of the involved sinuses were normal in 26 cases (60%) with spread of infection across uninvolved bone into the perisinus fat. Bone involvement (Fig. 2) in the form of bone rarefaction, erosions and permeative destruction was seen in 17 cases (40%), involving the sinus walls and the contiguous bony structures. Nasal cavity involvement on CT was seen in the form of non specific inflammatory turbinate hypertrophy and inflammatory fluid in the nasal cavity (17, 39%). Nasal septal involvement was seen in the form of septal perforation in 3 patients.

MRI

On MRI, the lesions were isointense or mildly hypointense on T1W images in all cases. On T2W images rhinocerebral lesions (Table 3)

Table 2
Sinuses involved in mucormycosis infection.

Sinuses involved	Number (%)
Maxillary	34(79%)
Ethmoid	37(86%)
Sphenoid	26(60%)
Frontal	1(2%)
Maxillary + ethmoid	10(23%)
Ethmoid + sphenoid	2(4%)
Maxillary + ethmoid + sphenoid	21(49%)
Pansinusitis	1(2%)

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