

# Gangrene nose: a rare presentation of Rhino-orbital-cerebral mucormycosis (ROCM)

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## Citation

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## Abstract

Rhino-orbital-cerebral mucormycosis (ROCM) is remerging as a medical and surgical emergency. The reasons are increasing numbers of patients with diabetes and immunosuppression. Here we present a case of diabetic ketoacidosis with fulminating mucormycosis and frank gangrene of the nose. The presentation was catastrophic and the patient could not be salvaged.

## CASE REPORT

A 55 year old diabetic female presented to our medical emergency with progressively worsening altered sensorium, periorbital puffiness and blackish discolorations over the nose of five days duration. She was hypotensive, with sinus tachycardia at admission. Neurologically she was arousable and had no meningeal signs or limb paresis. On local examination of nose, the tip, philtrum and left ala of nose were completely necrosed with poorly defined bluish expanding margins suggestive of wet gangrene (Figure 1).

### Figure 1

Figure 1: Clinical photograph shows gangrene of the tip, philtrum and left ala of nose.

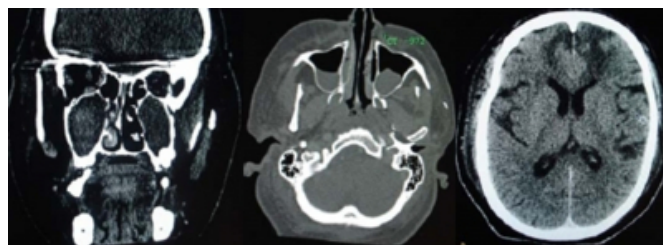


There was facial puffiness with subcutaneous crepitus over the cheeks, periorbital edema and cellulitis, bilateral ptosis with proptosis, chemosis and complete ophthalmoplegia with loss of pupillary reflexes both eyes.

On diagnostic nasal endoscopy black eschar was seen involving left inferior and middle turbinates. The investigations revealed plasma glucose of 520mg/dl, serum sodium 122mEq/l, potassium 5.4mEq/l, blood urea 110mg/dl, serum creatinine 3.4mg/dl and ketonuria. Arterial blood gas revealed severe metabolic acidosis. A non contrast CT of nose and paranasal sinuses showed gas formation in subcutaneous layer of the midface with bilateral maxillary, right ethmoidal, and left sphenoidal partial opacification suggestive of fungal sinusitis (Figure 2).

### Figure 2

Figure 2: An NCCT paranasal sinus shows bilateral maxillary, right ethmoidal, and left sphenoidal partial opacification; NCCT head reveals bifrontal hypodensities.



CT head revealed bifrontal hypodensities suggestive of cerebral infarct. Histopathological examination of nasal biopsy taken from the left lateral nasal wall revealed non septate branching mucormycotic hyphae. Patient was started on intravenous saline, inotropes, insulin infusion, liposomal

amphotericin, ceftriaxone and metronidazole. Surgical intervention was refused by the patient's relatives. Patient died on the fourth day of hospitalization.

## DISCUSSION

Rhino-orbital-cerebral mucormycosis (ROCM) is a fulminant and life threatening invasive fungal infection most commonly encountered in patients with uncontrolled diabetes mellitus. Of the seven different syndromes of mucormycosis namely the pulmonary, gastrointestinal, CNS, subcutaneous, disseminated, miscellaneous (bones, joints, heart, kidney, mediastinum) and the rhino-orbital-cerebral form, ROCM is the commonest in clinical practice<sup>1</sup>.

The term ROCM refers to the entire spectrum ranging from the sino-nasal tissue invasion (limited sino-nasal disease), progression to orbits (limited rhino-orbital disease) to finally central nervous system involvement (rhino-orbital-cerebral disease)<sup>2</sup>. ROCM is generally encountered in the settings of diabetic ketoacidosis, hematological malignancies, severe burns, renal or hepatic disease, AIDS, organ transplant recipients; however immunocompetent individuals are not immune to this dreaded disease.

In 1950 Smith and Krichner<sup>3</sup> gave the following criteria for the clinical diagnosis of mucormycosis: (i) a blood tinged nasal discharge and facial pain, both on the same side, (iii) soft peri-orbital or peri-nasal swelling, going on to discoloration, induration and progressive vascular occlusion, (iii) ptosis of the eyelid, proptosis of the eyeball and complete ophthalmoplegia, (iv) multiple unrelated cranial nerve palsies, and (v) black, necrotic turbinates, easily mistaken for dried, crusted blood. Most of the patients manifest the above symptoms and signs. Other less common features are retinal infarction, cavernous sinus, jugular and internal carotid thrombosis, terminating in seizures and death.

The typical clinical features of ROCM are as a consequence of invasion, thrombosis and necrosis of the underlying tissue. Thus demonstration of tissue invasion especially 'angioinvasion' on histopathology is mandatory for diagnosis besides the presence of broad aseptate filamentous fungi branching at right angles. The most common fungi associated with ROCM are the *Rhizopus*, *Rhizomucor* and *Absidia*, of these the *Rhizopus* accounts for 90% of the cases<sup>4</sup>. Lesser known but emerging fungi include the *Saksena*, *Apophysomyces*, and *Cunninghamella* species. The spores of these saprophytic fungi germinate favorably in an environment of low oxygen, high glucose, acidic medium

and high iron levels<sup>5</sup>.

ROCM presenting as gangrene nose is uncommon in medical literature. Bhansali et al<sup>6</sup> have published a large retrospective study of 35 patients with ROCM with underlying diabetes mellitus. The most common manifestations encountered by them were ophthalmic of which pronounced features included external ophthalmoplegia (89%), proptosis (83%), visual loss (80%), chemosis (74%), and eye lid gangrene (14%). Although in their study sinusitis was seen in 100% of the cases and nasal discharge/ulceration in 74% cases, palatal necrosis (29%) and facial necrosis (11%) were less common. However none of their patients had nasal gangrene as the primary presentation.

In a retrospective review of 34 patients of ROCM by S Nithyanandam et al<sup>7</sup> eyelid and maxillary skin necrosis was observed but none of the patients presented with gangrene nose. Similar case reports and reviews have identified eyelid gangrene, facial necrosis and palatal perforation as common features of ROCM, but there are no reports of patients presenting primarily as gangrene nose<sup>8,9</sup>.

Diagnosis of mucormycosis requires a high index of suspicion in the relevant clinical setting. The differential diagnoses in such clinical setting include septic emboli, atherosclerotic vasculopathy, vasculitis, local streptococcal and clostridial infection, and invasive aspergillosis.

An early diagnosis, prompt institution of antifungal therapy with timely surgical debridement is the key to survival. Correction of underlying disorder is of paramount importance. Survival rates in ROCM have ranged from 21-70% in the past<sup>5,10</sup> but are steadily improving with aggressive management strategies. Treatment outcome is good when the disease is anatomically confined to the sinuses. In other cases the disease carries a very high mortality with conservative treatment alone<sup>2,7</sup>.

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