

Case Report: Rhinocerebral Mucormycosis in An Immunocompetent Patient.

Majid Bani Ata¹, Mohammad Alsalem², Othman Beni Yonis³, Muawya Bani Younes¹

Abstract

Rhinocerebral mucormycosis (RCM) is a rare opportunistic infection, affecting almost exclusively patients with immunocompromised status, and represents the third most common angio-invasive fungal infection after candidiasis and aspergillosis. It is considered one of the most important medical complications in immunocompromised patients.

In spite of this, we report here a case of a RCM in an immunocompetent 48 year-old male who was referred to King Abdullah University Hospital (KAUH) in March 2013 from Jordan University of Science and Technology primary health care center (JUST primary health care center) as a case of worsening bacterial sinusitis, which was managed as an inpatient case with multiple endoscopic debridement surgery and intravenous amphotericin B with good outcome.

Keywords: Rhinocerebral; mucormycosis; immunocompetent

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Introduction

A 48 year-old non-smoker male patient, healthy and not known to have any chronic medical illnesses, was referred to Otorhinolaryngology urgent care clinic in KAUH as a case of right sided maxillary bacterial sinusitis with persistent fever and worsening symptoms despite one week treatment with antibiotics (co-amoxiclav. 1g twice daily for three days then levofloxacin 500 mg daily for four days).

Family physician's referral note stated that the patient was complaining of severe and worsening headache (8/10 on a scale from 0-10, 10 being the worst), right sided facial and temporal pain, facial fullness, hyposmia and fever (ranging from 38.0-39.1 °C orally) for about one week before presentation.

Patient's initial examination showed an ill-looking febrile patient (Temperature 38.5 °C orally) with mild right maxillary sinus tenderness and an ulcer (about 2-3 mm in diameter) at the right side of the hard palate.

1- Otolaryngology department, Faculty of Medicine, Jordan University of Science and Technology, Jordan.

2- Otorhinolaryngology-Head and Neck Surgery department, Faculty of medicine, Yarmouk University, Jordan.

3- Family medicine department, Faculty of Medicine, Jordan University of Science and Technology, Jordan.

* Correspondence should be addressed to:

Majid Bani Ata, MD.

Otolaryngology department, Faculty of Medicine,

Jordan University of Science and Technology, P O Box 3030, Irbid 22110, Jordan.

E-mail: Baniatamajid@yahoo.com

Anterior rhinoscopy showed clear nasal cavity, normal septum and no postnasal drip. Rigid nasal endoscopy was performed and showed necrotic right side middle turbinate and ulcer in the right mid-posterior part of the nasal, from both of which a biopsy was taken under local anesthesia.

Initial lab results showed an elevation in erythrocytic sedimentation rate (ESR) of 77 mm/hr, C-reactive protein (CRP) of 12 mg/l, normal WBCs of 10.3 cells/ μ l, Hemoglobin A1c (HbA1c) of 5.4%, and negative hepatitis and HIV profiles.

The patient was admitted and because the unavailability of liposomal amphotericin B in our center he was started on intravenous antifungal therapy (amphotericin B: 50 mg/day (0.5 mg/kg/day)) and intravenous antibiotic (ceftriaxone 1g twice daily) based on suspension of mixed bacterial and fungal infection.

Computed tomography (CT) scan showed opacification in the right maxillary sinus, ethmoidal sinuses, and right osteomeatal complex with evidence of bone erosion (figure 1). Chest CT was normal.

Paranasal sinuses and brain magnetic resonance imaging (MRI) showed no intracranial extension.

Histopathology report revealed the presence of clusters of fungal hyphae and spores involving bone trabeculae and the submucosa, consistent with invasive mucormycosis (figure 2).

Multiple endoscopic surgical debridements were done under general anesthesia (figure 3) and amphotericin B was continued for 35 days, during which he suffered from deterioration of renal function (serum creatinine level reached 362 μ mol/l), therefore amphotericin B was adjusted to 75mg every other day by the nephrologist. His last nasal exam under general

anesthesia and multiple tissue biopsies showed negative results and no evidence of recurrence after which he was discharged home.

Patient was followed up weekly in outpatient clinic the first month, then monthly for 6 months, then every 3 months for 6 months then every 6 months for 2 years with a total follow up duration of 3 years, during which no sign of recurrence was noted. The patient's renal function returned to normal level after 4 months of follow up with nephrology service.

Discussion

Rhinocerebral mucormycosis (RCM) is a rare disease, mainly affecting patients with known predisposing conditions such as diabetes mellitus, immunocompromised status, haemochromatosis or major trauma [3]. Mucormycosis is caused by fungi of the Mucorales order, the largest and best studied order of Zygomycete fungi. Mucorales is identified by cultural and microscopic characteristics of broad and nonseptate hyphae. In rhinocerebral and pulmonary mucormycosis, sporangiospores colonize the paranasal sinuses, nasal cavities, and lungs, whereby they invade the blood vessels and neural tracts [4, 5]

However, until 2011 A total of 212 cases were reported of immunocompetent patients affected by Mucormycosis worldwide, 81 of which were rhino-orbito-cerebral cases [6].

In immunocompetent patients, the nose and/or maxillary sinuses appear to be the predominant source of infection of the respiratory tract. Once infection has colonized nose and paranasal sinuses, if not promptly diagnosed and treated, it is likely that, in such patients, this infection may invade the base of the skull through blood vessels, disseminating to the central nervous system, giving the rhino-orbito-cerebral form or everywhere in the body,

giving the disseminated form^[7, 8]

Regarding the prognosis, a review showed that the mortality rate in the 73 patients with rhinocerebral mucormycosis and without apparent known predisposing conditions (47%) was similar to that found in the case series review of overall rhinocerebral mucormycosis (49.5%).^[9]

In general, survival rate is approximately 80% when medical and surgical interventions are both administered.^[10]

Conclusions

Identification of complication and referral of suspicious sinusitis cases is crucial in primary care setting.

Although it is rare, immunocompetent patients with no predisposing factors could get

rhinocerebral mucormycosis. However, the management stays the same as earlier recognition of rhinocerebral mucormycosis and prompt treatment with intravenous antifungal amphotericin B (preferentially liposomal amphotericin B) with endoscopic surgical debridement would improve outcome and survival rate.

Compliance with Ethical Standards:

Conflict of interest: Nothing to declare

Ethical approval: All procedures performed were in accordance with the ethical standards of the institutional committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: Informed consent was obtained from the patient in the case.

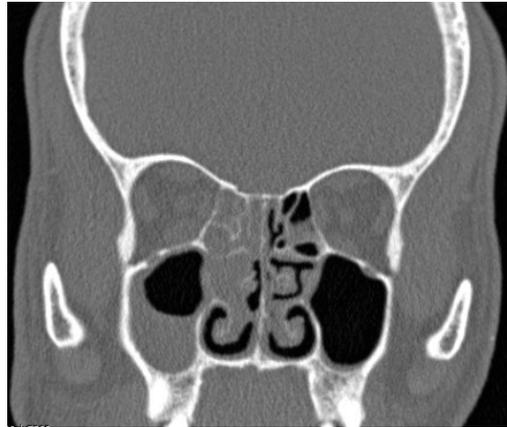


Figure 1: CT sinus (coronal view), showing right maxillary sinus, ethmoidal sinuses and osteomeatal complex involvement.

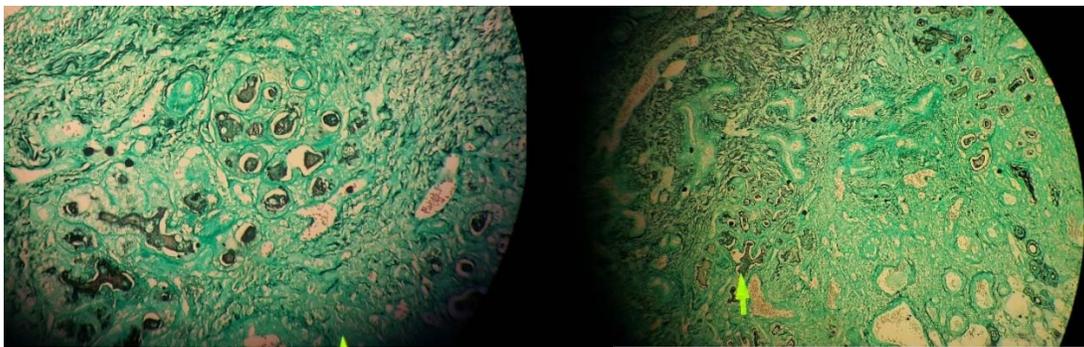


Figure 2: Histopathology slides showing mucormycosis in GMS stain.



Figure 3: intraoperative picture showing a remnant of the right middle turbinate and necrosis in the right side of the nasal septum.

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تقرير حالة: التهاب فطري في الجيوب الأنفية من نوع *Mucormycosis* في مريض طبيعي المناعة

ماجد بني عطا¹ ، محمد السالم² ، عثمان بني يونس³ ، معاوية بني يونس¹

1. قسم جراحة الأنف والأذن والحنجرة ، كلية الطب ، جامعة العلوم والتكنولوجيا الأردنية، الأردن.
2. قسم جراحة الأنف والأذن والحنجرة وجراحة الرأس والعنق ، كلية الطب ، جامعة اليرموك ، الأردن .
3. قسم طب الأسرة ، كلية الطب ، جامعة العلوم والتكنولوجيا الأردنية، الأردن.

الملخص

التهاب الفطري في الجيوب الأنفية من نوع Mucormycosis هو مرض نادر، ويؤثر بشكل شبه حصري على المرضى الذين يعانون من حالات مؤهبة معروفة مثل داء السكري أو وضعفية نقص المناعة أو وجود داء ترسب الأصبغة الدموية أو في حالة الإصابات البالغة. حتى عام 2011 تم الإبلاغ عن 212 مريضاً من ذوي المناعة الطبيعية المتأثرين بالتهاب فطري في الجيوب الأنفية من نوع Mucormycosis في جميع أنحاء العالم. في مراكز الرعاية الأولية من الصعب تشخيص التهاب فطري في الجيوب الأنفية من نوع Mucormycosis لأنه غالباً ما يكون في صورة مشابهة لالتهاب الجيوب الأنفية البكتيري .

هنا نبلغ عن حالة من (RCM) عند رجل يبلغ من العمر 48 عامًا ، وقد تمت إحالته إلى مستشفى الملك عبد الله الجامعي في مارس 2013 من مركز الرعاية الصحية الأولية التابع لجامعة العلوم والتكنولوجيا الأردنية كحالة متفاقمة من التهاب الجيوب الأنفية البكتيري. مستشفى الملك عبد الله الجامعي -مركز تحويلي من الدرجة الثالثة- ومركز الرعاية الصحية الأولية التابع لجامعة العلوم والتكنولوجيا الأردنية، هي مراكز معتمدة من اللجنة المشتركة الدولية (JCIA) في شمال الأردن.

الكلمات الدالة: التهاب فطري في الجيوب الأنفية من نوع Mucormycosis ، طبيعي المناعة.