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Case Report

Rare presentation of Nasopharyngeal Mucormycosis – Isolated headache as a Clinical feature to consider

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Abstract
We report a patient who was investigated for occipital head ache with no rhino-orbital symptoms developing a rapidly progressing mucormycosis, with the only initial sign being isolated nasopharyngeal wall necrosis. High degree of suspicion and prompt treatment was key to the successful treatment of this patient.

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Introduction

Rhino-orbito-cerebral mucormycosis is a rare fungal infection with high morbidity and mortality. We wish to highlight a rare presentation of initial occipital headache and isolated nasopharyngeal necrosis which was diagnosed as rapidly progressive invasive mucormycosis.

Case Report

A 42 year lady with long standing poorly controlled insulin dependant diabetes mellitus, with history of severe migraine and bronchial asthma, presented to the neurology department with isolated severe occipital headache for 3 days. She did not have any nasal symptoms and later complained of mild odynophagia. Initial investigations for Cerebrovascular accidents, meningitis and AV Malformations were negative. Despite, the patient had a persistently rising CRP value from 33 to 124mg/dl. LP studies and CT angiographies revealed nothing except insignificant fullness of bilateral fossae of Rossenmuller. She had a negative RT-PCR test for SARS COV-2.

Due to the suspicious CT findings a rigid nasal endoscopic evaluation revealed a necrotic patch over the posterior pharyngeal wall. Rest of nasal cavity was unremarkable. Urgent MRI revealed no specific sinus pathology or black turbinate sign except for mild enhancing posterior pharyngeal wall (Figure1).

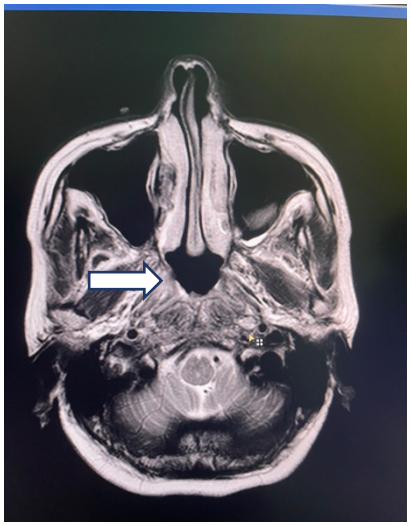

Figure1: MRI showing mild enhancement of posterior pharyngeal wall (Arrow)with normal sinus mucosae

High degree of suspicion led to examination under anaesthesia within 4 hours of initial assessment. The patient had developed complete necrosis of posterior pharyngeal wall, nasal septum, all turbinates, bilateral infratemporal fossae and pterygopalatine fossae. Prompt debridement followed by treatment with liposomal and conventional Amphotericin B were undertaken in the ICU setting due to initial reaction to drugs. With serial debridement patient showed signs of improvement. Mycological evaluation of the tissue biopsies from the nasopharynx revealed broad ribbon-like aseptate fungi on KOH wet mount suggestive of Zygomycetes with invasion. Extensive involvement of right infratemporal fossa was noted with no remnant of maxillary artery noted. Debridement of posterior pharyngeal wall was difficult due to the tough tissues attached to it. Patient also developed Sepsis due to long term Central venous access.

After 48 days of treatment with Liposomal Amphotericin(18 days) and Conventional Amphotericin B(30 days), she underwent second MRI to confirm only inflammation in bilateral infratemporal fossae with no evidence of residual disease. 2 consecutive negative reports for mucormycosis were obtained and on the advice of the Mycologist at MRI, Colombo, the patient was declared disease free. Serial weekly nasal endoscopies were done for the next 1 month.

Discussion

Mucormycosis is commonly caused by the broad class of Zygomycetes species and can rapidly progress to be a deadly disease with high mortality reported between 20 to 80% with a mortality rate of 46% in Rhinocerebral mucormycosis[1,2]. It is commonly seen in immunocompromised patients with uncontrolled diabetes[2]. COVID 19 infection with the use of steroids have been a recent risk factor[3], though our patient was covid negative.

MRI imaging is preferred to CT scans[4] when radiologically diagnosing patients due to the enhancement of soft tissue.Unfortunately,MRI and CT both only showed evidence of soft tissue inflammationaround the posterior wall of the nasopharynx. No soft tissue or bony involvement of the Atlas, Axis,or any ligaments. No enhancement of dura was noted. This left us with a conundrum on explaining the cause of isolated occipital headache.

High index of suspicion is needed to successfully diagnose this condition and early wide debridement of all necrotic tissues is key to the success of treatment[2]. AmphotericinB-soaked nasal packing was used[5]while liposomal Amphotericin was administered intravenously[2]. Conversion of Liposomal to conventional amphotericin Bwas made due to the lack of availability of the former. Regular nasal endoscopies and cleaning were undertaken atleast every 2 to 4 days.

Online literature search did not show any isolated nasopharyngeal mucormycosis, though its extensive aggressive nature has been well described.

Empirical treatment and high degree of suspicion of Mucormycosis of this patient at the onset of symptoms were a key factor in achieving a good outcome. Similarly, early surgical intervention and use of local Amphotericin packs may have been a contributary cause[5]. All management decisions were taken in line with theGlobal guidelinein diagnosis and management of mucormycosis-2019[2] and with a multi-disciplinaryapproach with team of Otorhinolaryngologists, Radiologists, Neurologists, Medical intensivistsat Teaching Hospital Anuradhapura and Consultant mycologist at Medical Research Institute, Colombo.

Conclusion

One needs to have a high degree of suspicion,especially with atypical symptoms such as isolated occipital headache in diagnosing mucormycosis. Empirical treatment and early surgical interventiongives the patient the best chance of survival.Immediate starting of intravenous Liposomal Amphotericin, regular debridement and multi-disciplinary approach is key to achieving a better outcome in patients with multiple comorbidities and risk factors with Mucormycosis.

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References:

1. Roden MM, Zaoutis TE, Buchanan WL, Knudsen TA, Sarkisova TA, Schaufele RL, et al. Epidemiology and outcome of zygomycosis: a review of 929 reported cases. Clin Infect Dis [Internet]. 2005;41(5):634-53.https://doi.org/10.1086/432579. PMid:16080086
2. Cornely OA, Alastruey-Izquierdo A, Arenz D, Chen SCA, Dannaoui E, Hochhegger B, et al. Global guideline for the diagnosis and management of mucormycosis: an initiative of the European Confederation of Medical Mycology in cooperation with the Mycoses Study Group Education and Research Consortium. Lancet Infect Dis [Internet]. 2019;19(12):e405-21. https://doi.org/10.1016/S1473-3099(19)30312-3
3. Raut A, Huy NT. Rising incidence of mucormycosis in patients with COVID-19: another challenge for India amidst the second wave? Lancet Respir Med [Internet]. 2021;9(8):e77. https://doi.org/10.1016/S2213-2600(21)00265-4
4. Groppo ER, El-Sayed IH, Aiken AH, Glastonbury CM. Computed tomography and magnetic resonance imaging characteristics of acute invasive fungal sinusitis. Arch Otolaryngol Head Neck Surg[Internet].2011;137(10):1005-10.https://doi.org/10.1001/archoto.2011.170.PMid:22006778
5. Seiff SR, Choo PH, Carter SR. Role of local amphotericin B therapy for sino-orbital fungal infections.OphthalPlastReconstrSurg[Internet].1999;15(1):28-31. https://doi.org/10.1097/00002341-199901000-00007.PMid:9949426