International Journal of Otorhinolaryngology and Head and Neck Surgery
Gavendra S et al. Int J Otorhinolaryngol Head Neck Surg. 2024 Jun;10(3):331-334
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20241332

Case Report

Fungal carotiditis with aneurysm - a rare complication of mucormycosis: presentation and management

Sakshi Gavendra, Gopishankar Subramaniasamy*, Anagha Joshi, Renuka Bradoo

Department of ENT and Head and Neck Surgery, Lokmanya Tilak Municipal Medical College and Government Hospital, Mumbai, Maharashtra, India

Received: 05 March 2024
Revised: 06 May 2024
Accepted: 08 May 2024

***Correspondence:**
Dr. Gopishankar Subramaniasamy,
E-mail: sgshere@gmail.com

ABSTRACT

During the COVID-19 pandemic, cases of mucormycosis (ROCM) had increased drastically, especially in the second wave. Most of these cases involved the paranasal sinuses, orbit and occasionally brain. This case report presents a rare case of mucormycosis complicated with a mycotic aneurysm of the petrous part of the internal carotid artery (ICA). A 43-year-old man presented with left-sided facial weakness for which high resolution computed tomography scan of the Temporal bones was done. A scrutiny of the computed tomography temporal bone showed evidence of osteomyelitis extending to the petrous apex around the foramen Lacerum. Computed tomography and magnetic resonance imaging of the paranasal sinus done later confirmed the diagnosis of mucormycosis. The patient underwent endoscopic surgical debridement of the nose and sinuses, with post-operative amphotericin B. After 3 weeks, the patient presented with bouts of profuse nasal bleed. Computed tomography angiogram showed an aneurysm in the petrous segment of the ICA suggestive of mycotic carotiditis complicated from mucormycosis spread along the skull base. It was successfully managed neurosurgically by a bypass shunting procedure. Mucormycosis is a disease that typically has an aggressive and startling fatality rate. ICA aneurysm is an uncommon occurrence in mucormycosis. Subjects with ROCM should be examined for brain and vascular involvement. Computed tomography angiogram should be performed as soon as possible if the skull base is involved in the ICA area. Morbidity and mortality can often be reduced by early diagnosis and interventions by a multidisciplinary team.

Keywords: Carotid aneurysm, Fungal carotiditis, Rhino-oculo-cerebral mucormycosis

INTRODUCTION

Though the incidence of rhino-oculo-cerebral mucormycosis (ROCM) is high in immunocompromised patients, due to the widespread use of steroids, chemotherapy, and immunosuppressive agents, the incidence of aneurysm of internal carotid artery (ICA) by mucormycosis is very rare.[1-3]

Invasive fungal disease of the skull base or cavernous sinus results in fatal vascular complications through inflammation or invasion of the adjacent ICA, leading to intra-arterial thrombosis, aneurysm formation, or rupture. Invasive fungal carotiditis can have devastating consequences, including cerebral infarction, subarachnoid haemorrhage and death.[4]

There are several case reports of fungal ICA aneurysms caused by invasive aspergillosis in literature. However, ICA aneurysm caused by invasive mucormycosis is a very rare phenomenon and only one case of petrous ICA aneurysm has been reported so far.[5-7] Here, we report a case of fungal ICA aneurysm due to mucormycosis of the skull base that was treated successfully by neurosurgical intervention.

CASE REPORT

A 43-year-old diabetic male presented to the ENT Outpatient department with complaints of left-sided ear discharge and left-sided facial weakness since 15 days. Patient did not have any nasal or oral complaints and no history of recent COVID infection. The patient had a history of stroke (cerebrovascular accident) 2 years ago and was on tablet Aspirin. The patient was a known diabetic and hypertensive on medication for two years.

Clinical examination revealed features suggestive of lower motor neuron facial palsy (Grade V House-Brackman classification). There was no crusting or pale mucosa on the anterior rhinoscopy examination. Ophthalmic and oral cavity examinations were unremarkable.

Ear findings were confirmed under the microscope which suggested a bulging inflamed tympanic membrane for which myringotomy was done. Pure tone audiometry was suggestive of left-sided moderate to severe conductive hearing loss. High-resolution computed tomography (HRCT) temporal bone showed focal erosions of the horizontal part of the facial nerve canal.

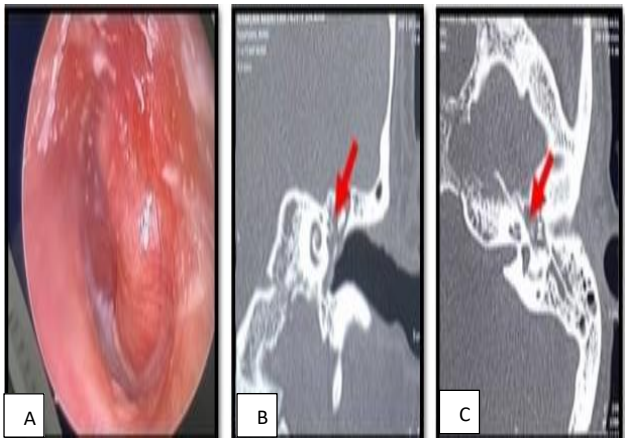

Figure 1 (A-C): Left-sided ear finding showing inflamed bulging tympanic membrane. HRCT temporal bone CT scan in coronal and axial plane showing erosion in facial canal in the tympanic segment.

There was evidence of abnormal soft tissue in the pterygopalatine fossa and infratemporal fossa mostly posterior periantral region with focal erosions of the maxillary sinus wall at the level of pterygomaxillary fissure on CT imaging of the temporal bone. There was thinning and rarefaction of the walls of sphenoid sinus with multiple erosions of the body, left lateral wall of sphenoid bone and clivus, suggestive of mucormycosis.

CT and MRI scan of the paranasal sinuses was then done, which showed contrast enhancement in the turbinates, perisinus area, pterygopalatine fossa and infratemporal fossa in the T1 weighted images, as in Figure 2 (A). There was loss of contrast uptake in the left nasopharynx and fossa of Rosenmuller reaching till foramen lacerum area and till near the ICA as shown in Figure 2 (B and C).

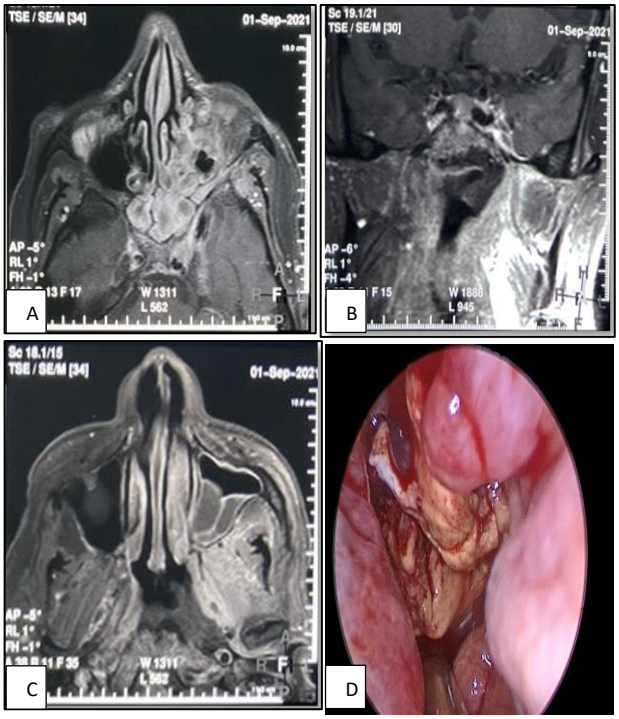

Figure 2 (A-D): MRI of paranasal sinus T1 post contrast axial section showing contrast enhancement in the turbinates, perisinus area, pterygopalatine fossa and infratemporal fossa, loss of contrast uptake in the left nasopharynx and fossa of Rosenmuller reaching till foramen lacerum area and till near the ICA and nasal endoscopy showing necrotic sloughed tissue over the sphenoethmoidal recess area, posterior to inferior turbinate and nasopharynx.

Diagnostic nasal endoscopy showed healthy nasal mucosa with normal middle meatus and turbinates. There was evidence of necrotic sloughed tissue over the sphenoethmoidal recess area, posterior to inferior turbinate and nasopharynx as in Figure 2 (D). A sample was sent for KOH mount from the nasal cavity and nasopharynx which tested positive for Mucormycosis.

Amphotericin B injection in the dose of 1-2 mg/kg/day, antibiotics, anti-diabetic treatment and facial physiotherapy were started. Endoscopic surgical debridement of the nose and paranasal sinuses was done, and the maxillary, ethmoid and sphenoid sinuses were widely opened. Left-sided sinus mucosa was healthy and the sinuses were opened wide with debridement of the slough in the nasopharynx. Minimal slough was left behind as it was seen extending deeper into the fossa of Rosenmuller. Intra-operative samples from the sinuses and nasopharynx were sent in a petri dish for KOH mount and histopathological examination. The fungus was tested negative for samples from paranasal sinus and was positive from the nasopharynx.

Post-operatively, the patient was continued on injection amphotericin B and medical treatment. Liposomal amphotericin B was given in the dose of 250 mg/day along with sugar level control, electrolyte correction and serum creatinine monitoring. Nasal endoscopy revealed no unhealthy tissue and nasal tissue samples were taken and proven negative for fungus before stopping anti-fungal treatment. The patient received a total dose of 6.5 g of liposomal amphotericin B.

After 1 month, the patient presented with complaints of nasal bleed on and off, for which nasal endoscopy was done, but it did not reveal any active bleeding focus, hence managed conservatively. The second episode was a massive bout of bleed that was so severe as to drop the haemoglobin levels from 10 gms% to 6 gms%. The patient was immediately admitted, for blood transfusion and management of the haemodynamic status.

CT Angiography was done which revealed an irregular lobulated fusiform aneurysm involving the petrous segment of left ICA measuring 2.4×1.2×1.6 cm as shown in Figure 3. A tiny 6×6 mm saccular outpouching was seen arising from the anterior aspect of this aneurysm protruding into the sphenoid sinus. Inferiorly, it was protruding into the soft tissue in the posterior wall of the nasopharynx. Bony erosions of the left half of body and greater wing of sphenoid, including the carotid canal, vidian canal, foramen ovale and clivus was seen.

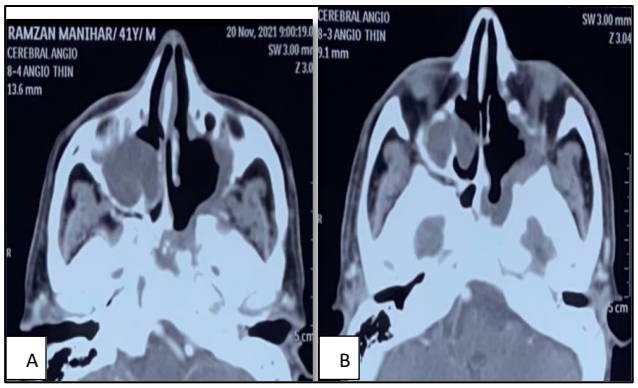

Figure 3 (A and B): CT angiogram showing aneurysm in the petrous segment of ICA.

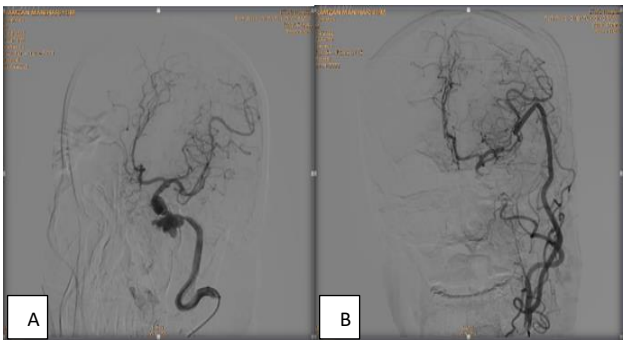

Figure 4 (A and B): Digital subtraction angiography-pre operative and post operative.

The patient was planned for neurosurgical intervention. Left-sided mini fronto-temporal craniectomy with superficial temporal artery (STA)-middle cerebral artery (MCA) end to side bypass (side to side) was done.

There were no further episodes of nasal bleed and nasal endoscopy showed healthy nasal mucosa with minimal slough in the nasopharynx. The patient was continued on posaconazole 300 mg daily for 3 months and regular follow-up.

DISCUSSION

Mucormycosis is an acute to subacute infection caused by invasive fungi from the order Mucorales, commonly found in soil and decaying plants. The primary genera responsible for human infection include *Rhizopus*, *Lichthemia*, *Apophysomyces*, *Mucor*, *Rhizomucor*, and *Cunninghamella*. This fungal infection manifests in various forms, including rhino-orbital-cerebral syndrome (most frequently observed in India), pulmonary, gastrointestinal, cutaneous, and disseminated mycosis.[8] Mucormycosis predominantly affects individuals with uncontrolled diabetes mellitus, hematological malignancies, hemopoietic or solid organ transplant recipients, and those with iron overload conditions.[8] Initial symptoms are diverse and may include pain, nasal congestion, nasal discharge, nasal bleed, edema, visual disturbances, and headaches. Survival rates of around 70% are reported with a combination of Injection Amphotericin B and prompt, aggressive surgical intervention. However, due to delays in diagnosis and treatment, most cases of mucormycosis carry a poor prognosis.[9]

Mucormycosis infiltrates the intracranial space through various pathways: (i) direct erosion of bone structures like the cribriform palate, frontal and sphenoid sinus bone walls, or orbital apex; (ii) via cranial nerve pathways; or (iii) intravascularly, following the blood circulation of the ophthalmic vessels, ICA, and basilar artery. This invasion leads to necrosis of the vessel walls, predisposing them to intraluminal thrombus formation and brain infarcts.[10-12] In cases of hematogenous dissemination, fungal aneurysms typically develop at distal sites of the anterior cerebral artery (ACA), MCA, or posterior cerebral artery (PCA) rather than the ICA, which is characteristic of mycotic aneurysms resulting from infectious endocarditis. Conversely, invasion from the paranasal sinus directly affects the intracavernous and supraclinoid portions of the ICA.[2]

Due to angioinvasion, the fungus involves intracranial vessels leading to ischemic stroke, aneurysm, carotico-cavernous fistula, sub-arachnoid hemorrhage (SAH) and intracerebral hemorrhage (ICH).[4] The main reason for lethality of mucormycosis is its high affinity for blood vessels, particularly for the elastic membrane of the arteries. The spores invade the vascular structures and are multiplied in the elastic lamina of the arteries. Hyphae

erodes the endothelium of the vessel walls and then causes necrosis, thrombi and infarcts.[13] Invasive fungal carotiditis are commonly due to aspergillosis followed by Mucorales.[4] The carotiditis complicates as an occlusion by thrombosis or stenosis or in a rare occurrence as an aneurysm or rupture. In the meta-analysis by Little et al the mortality rates in patients with mucormycosis associated carotiditis were 46% at 6 weeks to 65% at 2 years.[4]

Invasive fungal carotiditis represents a rare yet distinct manifestation of cranial invasive fungal disease. Due to its nonspecific presentation, technical challenges in microbiological identification, and limited literature available on this syndrome, diagnosis often experiences delays. There is scant knowledge about the optimal therapeutic approach to this disease; however, collaboration among otolaryngology, vascular medicine, and infectious disease teams is imperative. Early consideration of cranial invasive fungal disease, a proactive approach to biopsy and surgical debridement, determination of appropriate antifungal therapy, and regular vascular imaging and endovascular intervention should be considered to mitigate the high mortality associated with this unique condition.[4]

CONCLUSION

In conclusion, mucormycosis is a disease that typically manifests aggressively with an alarming mortality rate. ICA aneurysm is a very rare occurrence in mucormycosis. Imaging must be carefully interpreted to assess the extent of the disease, and surgical debridement should be correlated with the imaging findings. Patients with ROCM should be screened for brain as well as vascular involvement. In cases of skull base involvement around the ICA, CT angiography should be performed at the earliest opportunity. Morbidity and mortality can often be reduced by early diagnosis and interventions by a multidisciplinary team.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Al-Tawfiq JA, Alhumaid S, Alshukairi AN, Mohamad-Hani T, Mazin B, Al Mutair A, et al. COVID-19 and mucormycosis superinfection: the perfect storm. Infection. 2021;49(5):833-53.
2. Hot A, Mazighi M, Lecuit M, Poirée S, Viard JP, Loulergue P, et al. Fungal internal carotid artery aneurysms: successful embolization of an Aspergillus-associated case and review. Clin Infec Dis. 2007;45:e156-61.
3. Yamaguchi J, Kawabata T, Motomura A, Hatano N, Seki Y. Fungal Internal Carotid Artery Aneurysm Treated by Trapping and High-Flow Bypass: A Case Report and Literature Review. Neurol Med Chir (Tokyo). 2016;56(2):89-94.
4. Little JS, Cheng MP, Hsu L, Corrales CE, Marty FM. Invasive Fungal Carotiditis: A Rare Manifestation of Cranial Invasive Fungal Disease: Case Series and Systematic Review of the Literature. Open Forum Infectious Dis. 2019;6(10):ofz392.
5. Alvernia JE, Patel RN, Cai DZ, Dang N, Anderson DW, Melgar M. A successful combined endovascular and surgical treatment of a cranial base mucormycosis with an associated internal carotid artery pseudoaneurysm. Neurosurgery. 2009;65:733-40.
6. Jao SY, Weng HH, Wong HF, Wang WH, Tsai YH. Successful endovascular treatment of intractable epistaxis due to ruptured internal carotid artery pseudoaneurysm secondary to invasive fungal sinusitis. Head Neck. 2011;33(3):437-40.
7. Kim YC, Lee H, Ryu HH, Beom SH, Yang Y, Kim S, et al. Aspergillus-associated cerebral aneurysm successfully treated by endovascular and surgical intervention with voriconazole in lupus nephritis patient. J Korean Med Sci. 2012;27(3):317-20.
8. Prakash H, Chakrabarti A. Global epidemiology of mucormycosis. J Fungi (Basel). 2019;5(1):26.
9. Kumar NSS, Padala RK, Tirupati S, Anil KT. Rhinocerebral Mucormycosis with Top of Basilar Artery Syndrome. J Stroke Cerebrovasc Dis. 2016;25(2):378-82.
10. Thajeb P, Thajeb T, Dai D. Fatal strokes in patients with rhino-orbito-cerebral mucormycosis and associated vasculopathy. Scand J Infect Dis. 2004;36(9):643-8.
11. Turunc T, Demiroglu YZ, Aliskan H, Sule C, Hande A. Eleven case of mucormycosis with atypical clinical manifestations in diabetic patients. Diabetes Res Clin Pr. 2008;82(2):203-8.
12. Ma J, Jia R, Li J, Liu Y, Li Y, Lin P, et al. Retrospective clinical study of eighty-one cases of intracranial mucormycosis. J Glob Infect Dis. 2015;7(4):143-50.
13. Suganya R, Malathi N, Karthikeyan V, Janagaraj VD. Mucormycosis: A Brief Review, J Pure Appl Microbiol. 2019;13(1):161-5.

Cite this article as: Gavendra S, Subramaniasamy G, Joshi A, Bradoo R. Fungal carotiditis with aneurysm - a rare complication of mucormycosis: presentation and management. Int J Otorhinolaryngol Head Neck Surg 2024;10:331-4.