Case Report

Atypical Presentation of Fungal Skull Base Osteomyelitis: Middle Ear as the Primary Focus

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ABSTRACT

Introduction

Skull base osteomyelitis (SBO) is a complex condition seen in immunocompromised patients with severe mortality and morbidity. Fungal SBO is very rare and mostly sinogenic in origin. Otogenic (middle ear) fungal SBO is exceptionally rare. This case report details a case of atypical fungal SBO due to mucormycosis originating in the middle ear with its unusual clinical course.

Case Report

A 60 year old male with uncontrolled diabetes mellitus presented with left ear pain and multiple cranial nerve palsies. Initial imaging was suggestive of pansinusitis with SBO. Histopathology confirmed mucormycosis exclusively in the middle ear granulation tissue, not the sinuses, pinpointing an otogenic primary source. The patient received antifungal therapy with management of Diabetes Mellitus and other co-existing medical issues.

Discussion

This case highlights an extreme rare case of fungal skull base osteomyelitis from middle ear origin. It underscores the importance of histopathological examination given the initial misleading radiological findings. The patient showed a favourable response with medical management of Diabetes Mellitus and antifungal therapy despite his initial grim neurological presentation with multiple cerebral infarcts defying his grave prognosis.

Conclusion

This case highlights that though fungal SBO aggressive symptoms and signs of fungal SBO can lead to severe complications. Inspite of initial grim outlook, with vigilant management of underlying comorbid conditions and antifungal therapy there was remarkable clinical improvement.

Keywords

Fungal Skull Base Osteomyelitis; Mucormycosis; Antifungal Therapy

Skull base osteomyelitis (SBO) is a life threatening disease causing severe morbidity and even mortality in affected individuals. It affects immunocompromised individuals particularly with diabetes mellitus. Pseudomonas Aeruginosa is the most common organism involved in the disease.[1] However, cases of fungal skull base osteomyelitis are encountered infrequently in clinical practice. Fungal skull base osteomyelitis reported in literature have been mostly due to Aspergillus or Scedosporium species usually arising from contiguous external ear infection.[2,3,4] Fungal skull base osteomyelitis as a result of sinus infection in 55% cases was reported in a retrospective study.[5] Fungal skull base osteomyelitis is a rare entity. Interestingly fungal SBO secondary to otitis media is exceedingly rare. This case report highlights a presentation of fungal SBO due to mucor in a 60 year old patient with uncontrolled diabetes mellitus who presented with multiple cranial palsies, and initial work up was suggestive of pansinusitis with SBO, however it was only with histopathological examination which confirmed mucor solely in the middle ear granulation tissue. This case of fungal SBO secondary to

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otitis media is a unique and interesting case in the context of fungal skull base osteomyelitis.

Case History

A 60 year old farmer, who was a known case of Type II Diabetes Mellitus on irregular treatment presented to ENT OPD with left ear pain which was dull aching and with aggravation of pain at night since 5 days, left sided facial weakness , double vision, difficulty in chewing and swallowing since 3 days. Upon further inquiry he revealed a history of left ear discharge since childhood which was intermittent, profuse, mucoid, non foul-smelling and non blood stained and the last episode of ear discharge was 3 months back. Clinical examination revealed minimal purulent discharge in the left ear which was sent for gram staining and culture sensitivity. The tympanic membrane revealed a moderate sized perforation with protruding granulation tissue. Further clinical examination revealed a left side grade 4 lower motor neuron type of facial palsy (Figure 1), left lateral rectus palsy, deviation of tongue to left side on protrusion suggesting left hypoglossal nerve palsy, absent gag reflex, reduced movement of soft palate on the left side.

Fig. 1. Clinical picture of patient with left facial palsy

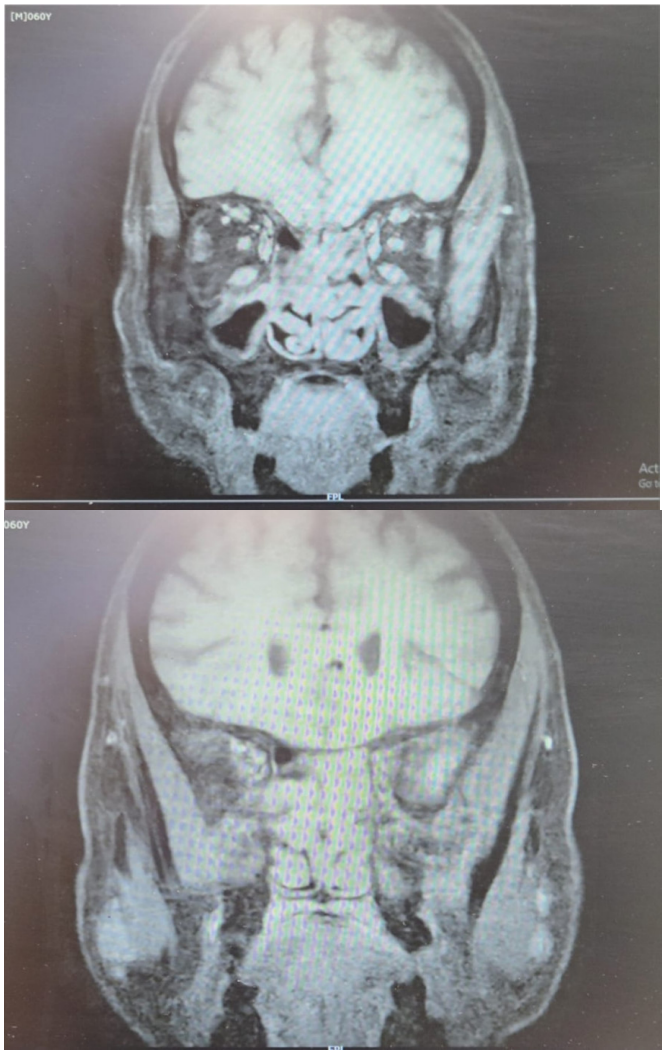

Fig. 2a, 2b. MRI showing features of pan-sinusitis

Diagnostic nasal endoscopy revealed normal mucosa without any discharge from the middle meatus. Videolaryngoscopy revealed normal vocal cord movements. A clinical diagnosis of skull base osteomyelitis with multiple cranial nerve palsies and uncontrolled diabetes mellitus was made. The patient was admitted in the intensive care unit as initial blood and urine work revealed very high blood sugar levels, dyselectrolytemia and a state of ketoacidosis. In the ICU the patient's condition deteriorated further and had to be put on ventilatory support. Concomitant medical issues were vigorously managed and Injection Meropenem was

initiated as a part of management of SBO. MRI of brain and skull base revealed pansinusitis (Figure 2a, 2b) with features of skull base osteomyelitis with thrombosis of left internal carotid artery and also infarcts involving areas of left anterior cerebral artery/middle cerebral artery and middle cerebral artery/posterior cerebral artery territories (Figure 3).

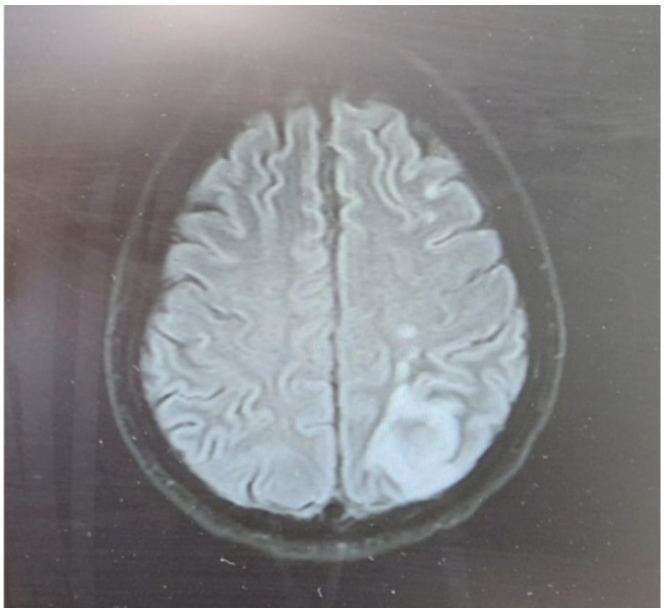

Fig. 3. MRI brain showing infarcts in left middle cerebral artery/posterior cerebral artery territory

A high resolution CT scan of the temporal bone revealed soft tissue opacification in the middle ear and partially in the mastoid. The air cell septations in the mastoid were maintained. The initial gram stain and culture from the left ear discharge revealed no growth or organism. Considering the imaging findings and the masked presentation of sinusitis, a fungal etiology was suspected, leading to the planning of a Functional Endoscopic Sinus Surgery (FESS). Concurrently, a plan was made to obtain and send tissue from the middle ear for histopathological examination during the same surgical setting. Intraoperatively, a pus sample from the right sphenoid sinus was collected and sent for Gram stain, culture, Ziehl-Neelsen (ZN) stain, and potassium hydroxide (KOH) mount. Sinus samples yielded no microbial growth/organisms. Further tissue samples from bilateral maxillary, anterior and posterior ethmoid sinus were sent for HPE, which showed features of chronic non-specific inflammation. However, HPE of the granulation tissue obtained from the left ear revealed necro-inflammatory changes with broad, aseptate hyphae consistent with mucormycosis (Figure 4a). Periodic acid–Schiff (PAS) staining highlighted magenta-coloured fungal colonies characteristic of *Mucor* species (Figure 4b).

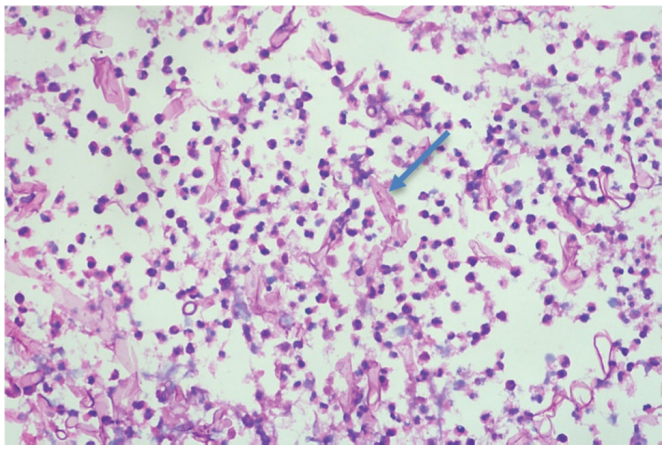

Fig. 4a. Fungal hyphae, obtuse angled, broad and aseptate PAS Stain 4x

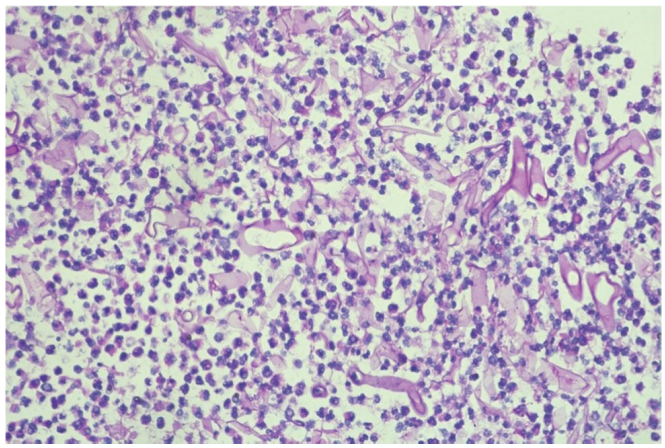

Fig. 4b. Fungal colonies PAS stain 4x

The patient's clinical status improved, enabling extubation, a positive outcome achieved through stringent glucose and electrolyte management, and notably, occurring despite the initial grim MRI findings. Following the histopathological report confirming mucormycosis from the middle ear granulation tissue , the patient was initiated

on Liposomal Amphotericin B, which was administered for 14 days. Subsequently, oral Isavuconazole was commenced. Injection Meropenem was discontinued. The patient's general condition improved, and ear pain significantly reduced after the initiation of this treatment. The patient was discharged on oral Isavuconazole and other medical treatments for blood sugar and other coexisting medical issues. At the time of discharge, the granulation tissue from the middle ear was no longer present, and a moderate-sized perforation of the tympanic membrane was observed. There was slight improvement in tongue deviation , but the remainder of the cranial nerve examination findings remained consistent with the previous assessment. Surprisingly, on follow-up after three weeks of discharge, there was no tongue deviation(Figure 5), soft palate movements were normal, and the gag reflex was present. The patient came walking despite such extensive infarcts seen on initial imaging. He reported no ear pain or discharge and was carrying out day-to-day activities. The rest of the cranial nerve examination was almost the same. Repeated imaging could not be performed due to financial constraints, necessitating reliance on clinical signs and symptoms to assess treatment outcomes.

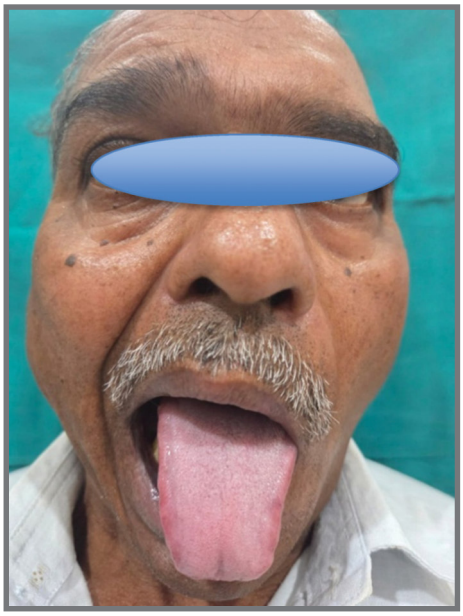

Fig. 5. Improvement in hypoglossal nerve function on follow up

Discussion

Skull Base Osteomyelitis is a complex and troublesome condition requiring rigorous and long term management including management of underlying conditions. Pseudomonas aeruginosa is the most common organism in 90-98% of cases of SBO.[6] Fungi like candida ciferri, Aspergillus species, candida parapsilosis, mucormycosis, Blastomycosis may also lead to SBO.[2,7,8] However, Aspergillus is the most common cause of fungal SBO in both immunocompromised and immunocompetent individuals.[9] These infections can lead to SBO either through contiguous spread of sinus or ear infection or hematogenous route. The pathophysiology of fungal SBO is less well understood and the complications of fungal SBO are more frequent than bacterial SBO[10,11] While SBO itself is a complex disease especially a fungal one adds a layer of diagnostic and therapeutic challenge. The diagnosis of fungal SBO is considered usually following failure of response to antibiotics.[2] The diagnosis requires microbiological confirmation. In our case despite the MRI showing extensive pan-sinusitis with SBO the pus samples from the sinus revealed no organism and histopathological examination of sinus mucosa only revealed chronic non-specific inflammation. In stark contrast the granulation tissue from the middle ear showed features of necro-inflammatory tissue with broad based hyphae suggestive of mucormycosis. This discrepancy strongly suggests the middle ear was the primary site of fungal infection. Blyth et al[5] in a retrospective study from 1990 to 2007 reported that out of 11 cases of fungal SBO only 2 had chronic otitis media, however the majority 54.5% were secondary to chronic sinusitis. This highlights an unusual primary site of infection.

For bacterial or fungal SBO the recommended treatment is 3-6 months of culture directed antibiotic or antifungal therapy.[12,13] Liposomal Amphotericin B with its lower nephrotoxicity is recommended[12] Aggressive surgical debridement is recommended by few clinicians for fungal SBO[4] in contrast to those with bacterial SBO.[14] In our case beyond functional endoscopic surgery no other aggressive debridement was done. The decision was influenced by the patient's remarkable and sustained clinical improvement with 2 weeks of liposomal

amphotericin B and continued oral Isavuconazole therapy as evidenced by reduction in ear pain, improvement of glossopharyngeal and hypoglossal nerve function. These improving clinical signs coupled with financial constraints preventing repeat imaging led to reliance on clinical assessment for observing treatment outcome and justified a more conservative approach.

Conclusion

Fungal skull base osteomyelitis can originate from a middle ear infection, and is exceptionally rare compared to the more common sinogenic route. Therefore, this case serves as a critical reminder that while rare, fungal infections of the middle ear can lead to severe and widespread complications like skull base osteomyelitis, and should be considered in the differential diagnosis, especially in patients with predisposing factors.

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