

# Malignant otitis externa with skull base osteomyelitis

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## ABSTRACT

Malignant otitis externa associated with skull base osteomyelitis is a condition seen classically in the elderly, diabetic patient. This disease is difficult to manage, often requiring long-term antibiotic therapy. Here we present such a case, seen in a 74- year-old lady. Initially, she was treated for a number of years in the outpatient department with intermittent ear complaints, but eventually required a hospital admission that lasted for 6 months due to a severe malignant otitis externa complicated by skull base osteomyelitis. We will discuss the clinical features, diagnostic criteria, imaging and management of this life-threatening clinical entity.

## INTRODUCTION

Malignant otitis externa associated with skull base osteomyelitis is a rare but life threatening condition. This disease is difficult to diagnose and manage, although advances have been seen with antibiotics and hyperbaric oxygen. Despite this, the disease has a significant mortality and morbidity rate. Prolonged intravenous antibiotics, regular aural toilet and strict glycaemic control are essential for successful treatment. [1](#)) Here we describe such a case involving an elderly, diabetic lady requiring a 6 month hospital stay.

## CASE REPORT

A 74-year-old lady presented to the Ears, Nose and Throat (ENT) clinic with episodic discharge, predominantly from the left ear. Multiple examinations revealed a central pinpoint perforation associated with occasional discharge. This was treated with Tri-adcortyl ointment or Otomize spray. There was no evidence of polyp formation or Cholesteatoma. Her right ear showed a healed perforation.

Subsequent severe headaches and right ear pain warranted hospital admission. Intravenous (IV) Piperacillin with Tazobactam and topical Ciprofloxacin drops were commenced. A Computed Tomography (CT) scan of the right temporal bone showed opacification of the right middle ear and mastoid cavity with focal erosion of the posterior canal wall. There was no bony involvement. Three days later, our patient was discharged on six weeks of oral ciprofloxacin and topical Aurecourt. Ear swabs reported no growth.

Three weeks later the patient was admitted with a grade II right sided facial nerve palsy. A cortical mastoidectomy and atticotomy were performed. Histology showed granulation tissue, consistent with chronic infection. The facial nerve palsy remained unchanged following

surgery. A further two weeks of IV Piperacillin with Tazobactam were commenced, followed by four weeks of oral Ciprofloxacin.

A further two weeks later, our patient was readmitted. A repeat CT showed a 5cm soft tissue mass along the skull base, inseparable from the walls of the nasopharynx associated with bony destruction. Deep post-nasal biopsies were taken by skull base surgeons, histologically these showed granulation tissue. She was discharged on a further course of oral Ciprofloxacin.

Our patient represented with ear pain and discharge. IV Meropenem and Vancomycin were commenced for two weeks. A repeat CT scan showed no improvement in the skull base mass. IV antibiotics were continued along with IV Voriconazole .

A magnetic resonance image (MRI) scan showed significant disease progression. Following review by the skull base consultants, malignant otitis externa and skull base osteomyelitis was felt still felt to be the most likely diagnosis and long term IV antibiotics were continued with input from microbiology. Our patient has now completed a further two months of IV antibiotics (Meropenem and Vancomycin) and a repeat MRI scan showed a small improvement in the soft tissue surrounding the skull base.

Clinically our patient has improved, with resolution of her pain, headaches and facial nerve palsy. She is afebrile and her inflammatory markers have normalised. The inflammatory polyp in the right ear resolved with silver nitrate cautery.

Our patient has been discharged on oral Co-amoxiclav.

## DISCUSSION

Malignant otitis externa progressing to skull base osteomyelitis is an aggressive infection of the temporal bone and skull base, associated with possible involvement of the facial nerve, carotid artery, jugular vein and mastoid. It was termed malignant, due to the high mortality rate and poor response to treatment. (2)

The most common causative pathogen is *Pseudomonas Aeruginosa*, but cases of *Staphylococcus*, *Candida* and *Aspergillus* have also been reported. The only pathogen identified in this case was *Aspergillus*, possibly due to prolonged antibiotic usage. (3)

The pathogenesis of this condition is unclear, however, a number of factors contribute; microangiopathy, hypoperfusion and diminished host resistance due to diabetes. (4) External otitis progresses from the external auditory canal to the temporal bone and the skull base via the fissure of Santorini and the osteocartilaginous junction.

Mortality and morbidity has improved with antibiotics. Mortality remains around 33% and increases to 80% with cranial nerve involvement. Predictors of an adverse outcome include skull base osteomyelitis, intracranial extension and multiple cranial nerve involvement.

Affected patients classically are elderly diabetics. If suspected in children, Human Immunodeficiency Virus (HIV), malnutrition and immunodeficiency must be considered.

Levenson's criteria can be used for diagnosis. Criteria include: refractory otitis externa, severe nocturnal otalgia and purulent otorrhea associated with Pseudomonas infection and granulation tissue in an immunocompromised or diabetic patient. Facial nerve palsy, swallowing problems and hoarseness may occur if cranial nerves are involved. (3) Inflammatory changes and granulations are noted in the external auditory canal. Pain is often disproportional to changes seen at otoscopy.

A CT scan is the investigation of choice for both diagnosis and monitoring as it delineates subtle changes in bone density and establishes the extent of soft tissue swelling. It is less useful in identifying disease resolution due to the duration of bone re-mineralization. MRI scanning shows excellent soft tissue detail and serial scans have been used for follow up. Radioisotope scans (technetium 99 / gallium 67) are useful for monitoring treatment response. They are not used for initial assessment due to poor anatomical localisation. Serial monitoring of CRP and ESR can help evaluate response to antibiotics. (3,4)

Treatment of malignant otitis externa and skull base osteomyelitis is a long process. Meticulous aural toilet, antibiotics and strict glucose control in patients with diabetes, are absolutely vital for success.

Antibiotic choice depends upon hospital policy and microbiology advice. Classically, first line treatment is oral Ciprofloxacin. If disease progression occurs then intravenous antibiotics are used. A number of regimes have been documented in the literature. The length of antibiotics required is dictated by the patient's clinical picture and inflammatory markers.

Concomitant use of hyperbaric oxygen and antibiotics can be considered for cases of intracranial spread, when the disease is recurrent or refractory to antibiotics. A Cochrane review concluded not enough data was available to provide recommendations. (1,5)

Surgical resection of diseased bone is not recommended due to disease spread through fascial and vascular planes. Biopsies can be obtained and any abscess drained. In the presence of facial nerve palsy, decompression is not indicated.

Complications of malignant otitis externa and osteomyelitis include; meningitis, abscess, sagittal, dural and cavernous sinus thrombosis.

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