

## Case Report

## Malignant External Otitis: Report on a Rare Case

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### Abstract

Necrotizing or malignant external otitis is an inflammatory condition of the external ear that is usually caused by infection by *Pseudomonas*. It is more prevalent among elderly, diabetic or immunosuppressed people. It generally occurs through trauma and/or iatrogenic maneuvers in the external auditory canal, thus providing an entrance for infection. This entity is therefore more common unilaterally, and bilateral occurrence is rare. Facial paralysis is common on these cases; along with occasional involvement of other cranial nerves. The present case report describes a patient with bilateral malignant external otitis who presented destruction of the infratemporal bone and auditory canals on both sides. This case was refractory to optimized clinical treatment, and a surgical approach was needed to improve the symptoms.

**Keywords:** Bilateral malignant external otitis; Infratemporal destruction, surgical approach for malignant external otitis; Infection due to *Pseudomonas*

### Introduction

Necrotizing External Otitis (NEO) is an infrequent infectious disorder but it is a serious condition with potential risk of mortality. It begins in the auditory canal and progresses with osteomyelitis of the temporal bone and base of the cranium. It can lead to paralysis of the cranial nerves, thrombosis of the sigmoid sinus, meningitis and death. The bacterium most often found is *Pseudomonas aeruginosa*, which is an obligate aerobic Gram-negative species that is not usually found in cultures from normal auditory canals. The initial clinical condition is uncharacteristic, resembling acute diffuse external otitis, which leads to delay in the diagnosis. NEO usually occurs subsequent to skin trauma that gives rise to small lesions in the auditory canal. Severe otalgia is developed as the most frequent symptom (in 90% of the cases), followed by otorrhea (50 to 80%), edema, occipital headache and temporal headache. NEO characteristically affects only one side, but because the pathophysiological basis consists of systemic immunosuppression, bilateralism is plausible. However, turning this clinical suspicion into a diagnosis is expensive. In this regard, NEO should generally be a differential diagnosis in cases of severe refractory otalgia in patients presenting comorbidities. As NEO evolves, chondritis occurs first, followed by osteitis of the auditory canal without involvement of the middle ear. Propagation of osteitis to the base of the cranium occurs, with lesions in the cranial nerves and osteomyelitis of the base of the cranium. The diagnosis is complemented through computed tomography on the temporal bone, with contrast, and this is the imaging examination that is most important for evaluating the extent of the condition. Nonetheless, computed tomography does not aid in evaluating its evolution, since the alterations are persistent. Scintigraphy using technetium-99 has high sensitivity (97.8%) and can depict osteoblastic activity. However, scintigraphy using gallium-67 has the capacity to differentiate acute infection and is useful for follow-up. Here, we report on a case of malignant external otitis with involvement and destruction of bone tissue bilaterally.

### Case Report

The patient was a retired 81-year-old white man who had been born in and was living in São José do Rio Preto. He was examined by an ear,

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nose and throat specialist because of a complaint of severe bilateral otalgia that he had had for four months. This examination revealed that the both external auditory canals presented edema, with a significantly diminished lumen, otorrhea and debris. The tympanic membrane was inflamed, but there was no perforation and no evident alteration in the middle ear. Granulomas were present in both auditory canals in the posteroinferior region.

He presented left-side facial paralysis that was classified as House-Brackmann grade IV. Audiometry showed severe deep sensory-neural loss on the left side and anacusis on the right side.

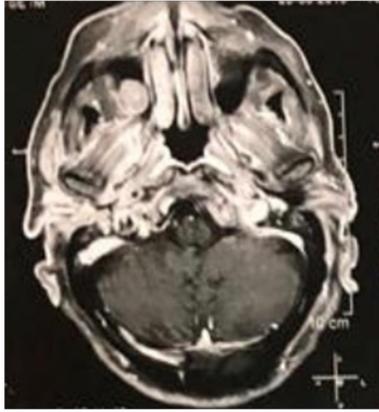
He had histories of diabetes mellitus and high blood pressure. He had been seen recently by other medical professionals within these fields, and presence of tumoral lesions was among the diagnostic hypotheses raised. However, after examinations had been conducted, it was concluded that this was a case of bilateral malignant external otitis and treatment for this condition was started.

From a pre-treatment swab, growth of *Pseudomonas aeruginosa* was found bilaterally. Computed tomography showed erosion of the tympanic bone, thickening of the soft tissues and bilateral erosion of the external auditory canal (Figures 1-3). Scintigraphy using technetium-99 was then indicated. This showed the presence of intense osteoblastic activity in the region of the right and left mastoid, which was concordant with presence of osteomyelitis bilaterally. In this manner, a final diagnosis of bilateral malignant external otitis could be reached.

The treatment that was proposed after the diagnosis had been made consisted of hospitalization for intravenous antibiotic therapy and an attempt to control glycemia levels. Anti-*Pseudomonas* antibiotics were chosen, consisting of ciprofloxacin (400 mg twice a day) in association with cefepime (1 g twice a day). After four weeks of this treatment, which did not yield complete clinical success, therapy in a hyperbaric chamber was attempted as an adjuvant to the drug treatment. After this combined therapy, the patient was released from hospital with a course of oral ciprofloxacin.

Subsequently, the patient again sought medical assistance from an ear, nose and throat specialist because he continued to have a complaint of bilateral stabbing otalgia. He was continually using analgesics, including morphine, and his left-side peripheral facial paralysis persisted.

On this occasion, we proposed that the patient should undergo a surgical approach, given that clinical treatment had been unsuccessful. The technique used comprised radical open mastoidectomy on both



**Figure 1:** Axial cutting computed tomography show ederosion of the etympani cbone, thickning of the soft tissues.



**Figure 2:** Enlargement axial cutting computed tomography show ederosion of the etympani cbone also erosion of the external auditory canal.



**Figure 3:** Coronal cutting computed tomography showed the involvement of soft tissues and the degree of steomy elitis of the base of cranyum.

sides, with the surgical aim of debridement of the devitalized tissues and the bone tissue that had been affected by osteomyelitis. The patient evolved with improvement of the symptoms after the operation.

## Discussion

The first case report on this condition was published in 1836, [1] and the term 'malignant external otitis' was coined in 1968, given its high mortality. Malignant external otitis is an infection that originates in the external auditory canal and progresses from initial cellulitis to chondritis, osteitis and, finally, osteomyelitis [2]. Because the cranial

bone is involved, there is progressive replacement of compact bone with granulation tissue [2,3]. It is important to bear in mind that the diagnosis of malignant external otitis should be considered in all cases of diabetic or immunocompromised patients presenting external otitis [4,5]. Diabetes continues to be the most important associated condition, and there are several mechanisms explaining its role, including defects of chemotaxis and phagocytosis of lymphocytes, monocytes and macrophages, thereby increasing the adherence of microorganisms beyond the situation of diabetic microangiopathy [5]. Hence, the possibility of attempting treatment in a hyperbaric chamber is valid, given that this enables increased phagocytic activity among bacteria, boosts vascularization and has synergism with antibiotics [6]. The systemic comorbidities presented by our patient were what made it possible for bilateral disease to develop.

In most cases, the causative agent is *Pseudomonas aeruginosa*, and this is present in 96-98% of cases of NEO [4]. The infection generally leads to severe otalgia that is resistant to analgesics. This irradiates to the frontotemporal and parietal regions and it worsens at night [7]. Furthermore, there may be complaints of aural fullness, fetid otorrhea, purulence and conductive hypoacusis. The material collected from the external auditory canal should routinely be cultured and the patient's sensitivity to anti-*Pseudomonas* antibiotics should be investigated [4]. In addition, if abnormal tissue is present in the external auditory canal, this should be biopsied to rule out malignancy. Physical examination will usually reveal situations of edema of the auditory canal that obliterate the lumen and increase sensitivity [5,8]. The tympanic membrane may appear to be normal. Erythrocyte sedimentation rate (VHS) is a nonspecific test that can be used to monitor the response to antibiotic therapy. Non-diabetic patients should be evaluated regarding control over diabetes.

Presence of refractory otalgia, paralysis of the facial nerve, jugular foramen syndrome and high uptake of technetium in scintigraphy are signs of poor prognosis [2]. Radiological examinations are necessary in order to assess the extent and severity of the disease. Computed Tomography (CT) is used initially, to evaluate the involvement of soft tissues and the degree of osteomyelitis of the base of the cranium [8]. However, its usefulness for monitoring the response to treatment is limited, because the initial alterations due to demineralization that are detected at the start of the disease persist despite resolution. Magnetic Resonance Imaging (MRI) is of only limited use for detecting bone alterations, but it is better than CT for identifying alterations to soft tissues. Again, the alterations that are identified initially do not become resolved with resolution of the disease and, therefore, MRI is also not useful for monitoring the response to antibiotic therapy. Nuclear scintigraphy has been the basis for diagnosing and following up patients with NEO. Early bone involvement can be detected by using technetium, while the continuing response to therapy is best monitored using a gallium scan, since this provides a sensitive indication of active infection. The treatment for NEO requires initial hospitalization for administration of intravenous antibiotic, using ciprofloxacin. After this, a prolonged period of oral drug administration, lasting up to six months, is needed [4]. However, there are reports of drug resistance in therapies for osteomyelitis, affecting around 20% of the patients [9]. One important clinical sign that demonstrates that the therapy is ineffective is the continuing presence of otalgia [10]. Most patients achieve improvement through clinical treatment. Before efficient anti-*Pseudomonas* antibiotics became available, this disease was only dealt with surgically, and the mortality rate associated with this was almost 50% [6,9]. With today's efficient antibiotics, surgery has become reserved for a few cases that are more aggressive and persistent, and it is used in approximately 6.5% of the cases [3]. In these surgical procedures, the aim is to clean the site, with debridement of devitalized tissue and infected bone. In most cases, a favorable outcome regarding eradication of infection is achieved. Thereafter, patients need to be routinely

followed up and every effort needs to be made to identify and treat any evidence of residual infection, with the aim of progressively diminishing the mortality rate [3,4].

## Conclusion

Necrotizing external otitis is a severe and potentially lethal disease. It generally affects only one side. Bilateral cases are extremely rare, and these have even more aggressive and unpredictable evolution. Thus, it is important to have a high degree of suspicion in order to ensure correct management, as in the case described here.

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