Case Report

Extensive Skull Base Osteomyelitis Secondary to Malignant Otitis Externa

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INTRODUCTION

Extensive skull base osteomyelitis is a severe complication of malignant otitis externa (MOE). It is most commonly diagnosed in patients with diabetes and in immunocompromised patients with bacterial and/or fungal infection. The primary infection is often bacterial with fungal superinfection. Pseudomonas aeruginosa is the causative pathogen in 50%-90% of cases, although other agents may be isolated [1].

CASE PRESENTATION

A 76-year-old man was referred to our Ear Nose and Throat (ENT) department in 2015 with a 4-month history of worsening right-sided MOE with the involvement of the ipsilateral otomastoid structures and ipsilateral temporal bone. Over the following 3 years, despite specific extended antibiotic therapy, the skull base osteomyelitis entirely involved the skull base, up to the contralateral petrous portion of the temporal bone, and it affected the cervical vertebral processes. This report describes an exceptional extent of unilateral malignant otitis externa with a severe involvement of the skull base on the contralateral side and the cervical spine.

KEYWORDS: Malignant external otitis, osteomyelitis, skull base

Skull base osteomyelitis is a severe complication of malignant otitis externa that affects the marrow of the temporal, sphenoid, and occipital bones. Skull base osteomyelitis is usually diagnosed based on clinical, microbiological, and radiological findings. Here, we present the imaging findings of a 76-year-old man who initially presented with right-sided malignant otitis externa, with the involvement of the otomastoid structures and ipsilateral temporal bone. Over the following 3 years, despite specific extended antibiotic therapy, the skull base osteomyelitis entirely involved the skull base, up to the contralateral petrous portion of the temporal bone, and it affected the cervical vertebral processes. This report describes an exceptional extent of unilateral malignant otitis externa with a severe involvement of the skull base on the contralateral side and the cervical spine.

Computed tomography (CT) revealed occlusion of the right external auditory canal, chronic osteomyelitis of the right temporal bone extending to the right mastoid and tympanic cavity, erosion of the ossicular chain, partial erosion of the inner ear, and preservation of a small portion of the cochlea (Figure 1a). The osteomyelitis extended to the right stylomastoid foramen, jugular foramen, and hypoglossal canal. The patient underwent surgery to obtain biopsies of the cortex of the mastoid bone and tympanic cavity for histopathologic and microbiologic examination. The results confirmed a *Corynebacterium Amycolatum* infection, extensive inflammation, and an absence of fungal organisms, and they were negative for BK. Due to poor response to antibiotic therapy, we decided to shift to fluoroquinolones. In particular, a therapy with moxifloxacin (Moxifloxacinca Teva, Teva Italia s.r.l., Milano, Italy) 400 mg per day, orally, was administered for 2 years. In an attempt to improve response to oral antibiotic treatment, we proposed to the patient a mastoidectomy, but he refused. During a follow-up period of over 3 years, the MOE had been persistent and, inexorably, progressive. The patient presented several complications including liquorrhea, caused by a retro-auricular fistula in December 2016, epistaxis and right otorrhagia in May 2017, and in October 2018, he expired. The osteomyelitis had involved the entire skull base up to the contralateral petrous portion of the temporal bone and affected the spinal processes of the cervical vertebrae. The final CT scan of the brain obtained before the patient’s death revealed an area of marked bone lysis in the right petrous portion of the temporal bone and the right mastoid, with an aerial component caused by drainage of the infective-inflammatory material through pre- and post-ear fistulas of soft tissue (Figure 1b). The osteomyelitis crossed the midline to involve the contralateral petrous temporal bone and included the bony structures of the right occipital condyle, the lateral masses of C1 and C2, and the posterior arch of the atlas (Figure 2). The fluid-filled space at the cervicomedullary junction was decreased, and the occipito-parietal skull was involved.

Informed consent was obtained from the patient during data collection and also from the patient’s wife, after his death.

**DISCUSSION**

Malignant otitis externa is the osteomyelitis of the temporal bone that usually occurs in elderly diabetic or otherwise immunocompromised patients; occasionally, the infection can spread and cause a skull base osteomyelitis [7]. In particular, microangiopathy and impaired blood circulation in patients with diabetes may play a main role in the pathogenesis of the disease [8]. Other causes can favor vascular impairment, such as the radiotherapy or the unhealthy diet [9].

The disease is associated with serious complications with a cranial nerve involvement and high mortality and morbidity rates. MOE is also reported in immunocompromised or diabetic children; however, the incidence is not as common as in elderly diabetic patients [8].

Clinical features of MOE include severe otalgia, purulent otorrhea, aural fullness, and hearing loss [10]. Temporomandibular joint pain, hemifacial pain, headache, and trismus are other common features that can develop from the anterior extension of the disease. Facial nerve palsy can be the presenting feature in some cases [11]. Levenson’s criteria can be used for diagnosis. These criteria include refractory otitis externa, severe nocturnal otalgia, purulent otorrhea, the presence of granulation tissue in the external auditory canal, growth of *Pseudomonas* in the culture from ear discharge, and presence of diabetes or immunocompromised state [10].

*Pseudomonas aeruginosa* is the most commonly isolated microbiologic agent for this disease [7]. Other bacteria such as *Staphylococcus aureus*, *S. epidermidis*, *Proteus mirabilis*, *Klebsiella oxytoca*, and *P. cepacia* have been isolated in MOE [3]. Moreover, a mycotic super infection can complicate the disease. *Aspergillus fumigatus* is the most common fungal organism causing MOE. Bacterial culturing provides the basis for antibiotic selection. If culture results are negative, ciproflo-
The pathogen isolated from the mastoid biopsy was *Corynebacterium Amycolatum*. The spectrum of human infections with *Corynebacterium* is broad ranging from community-acquired infections such as conjunctivitis, pharyngitis, genitourinary tract infections, prostatitis, skin and soft-tissue infections, and breast abscess to nosocomial-acquired infections such as cerebrospinal fluid infections, pneumonia, intraabdominal blood stream infections, endocarditis, post-surgical infections, urinary tract infections, and peritoneal dialysis-related peritonitis [11]. *Corynebacterium* is a cause of otitis and osteomyelitis, and the infection is associated with prior otologic procedures and treatments [12]. The association of previous radiotherapy with the surgical procedure may have caused the development of this rare infection.

The patient was treated exclusively with antibiotic therapy. He underwent surgery only to obtain biopsies of the mastoid cortex and tympanic cavity for histopathologic and microbiologic examination. The introduction over the years of newer, more effective, and less toxic antibiotics has reduced the role of surgery. Moreover, according to us, an extensive surgical procedure in this particular patient could have caused spreading of the infection and increased morbidity and complications.

In our case, clinically there was a marked progression of unilateral MOE with a severe involvement of the skull base, the contralateral side, and the cervical spine inferiorly. Bilateral skull base osteomyelitis is very rare, and there are no reports of bilateral involvement of the skull base from extension of unilateral disease. However, cases of bilateral skull base osteomyelitis concomitant with or following bilateral MOE have been described [13]. In some cases of bilateral skull base osteomyelitis, there is no evidence of an obvious active source of infection, making it impossible to identify MOE as the source. Furthermore, the spread of the infection to the cervical spine in patients with skull base osteomyelitis is extremely rare, with only two such cases being described in the literature [13].

**CONCLUSION**

Unilateral MOE complicated by extensive skull base osteomyelitis in a non-diabetic and immunocompetent patient is uncommon. An extensive skull base osteomyelitis extending up to the contralateral petrous temporal bone and cervical vertebra is exceedingly rare. This case presented multiple concurrent factors: prior radiotherapy treatment, an extensive otitis externa, a rare bacterial infection; perhaps, the sum of these conditions could have caused the extraordinary extension of the infection to the skull base. We recommend early detection of such cases in non-diabetic and immunocompetent patients.

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**REFERENCES**


