



Case report

Multiple cranial nerve palsies in malignant external otitis: A rare presentation of a rare condition



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ABSTRACT

Malignant external otitis (MEO) is a rare inflammatory and infectious condition, typically caused by *Pseudomonas aeruginosa*, that mainly affects diabetic or immunocompromised elderly patients and is associated with severe morbidity and mortality. It begins in the external auditory canal and rapidly progresses through the skull base, leading to osteomyelitis and may result in cranial neuropathy, especially of the facial nerve. Here we describe a rare neurological presentation of MEO in a 65-year old diabetic man, who presented with an 8-month progressing left otitis externa and evolved with ipsilateral proptosis, ophthalmoplegia, blindness, facial palsy, hearing loss and contralateral involvement of the temporal bone with hearing impairment. He was initially treated with oral ciprofloxacin and after one week was transferred to our tertiary hospital, where antibiotic therapy was switched to meropenem and vancomycin due to the severity of the case and to the hospital's microbiological profile. The patient underwent left canal wall-up mastoidectomy with insertion of ear ventilation tube bilaterally, with good recovery of right ear hearing capacity, but with no improvements of neurological deficits nor left hearing function. All microbiological tests performed were negative, and this was interpreted as a possible consequence of the early use of antibiotics. Unfortunately, the patient was infected by Sars-CoV-2 during hospitalization and passed away after ten days of COVID-19 intensive care unit internment.

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Introduction

Malignant external otitis (MEO) is a potentially life-threatening infection of the soft tissues of the external ear, quickly spreading to involve the periosteum and bone of the skull base. The condition typically begins in the outer ear, manifesting with refractory otalgia that irradiates to the frontotemporal and parietal regions, usually worsening in intensity at night, accompanied by smelly otorrhea with purulent aspect [1]. Its extension to the skull base

adds severity and results in an increased risk of cranial nerve involvement, with facial nerve palsy being the usual finding [2]. The most common etiologic agent is *Pseudomonas aeruginosa*, a microorganism that is unlikely to cause infections in healthy tissues, but able to initiate an infectious process through small traumatic injuries caused by iatrogenic manipulation in the external ear [1]. This type of infection is mostly described in elderly, diabetic and immunocompromised patients and, considering the high lethality rate, should always be suspected when there is an external otitis in a patient with a known risk factor and in those who do not respond well to the treatment of an apparent uncomplicated external otitis [3].

Even though MEO is a rare disease, it should be recognized by primary care and emergency physicians, who are frequently the first professionals sought by these patients. Thus, with the early diagnosis of MEO, treatment can be promptly instituted, resulting in significant reduction in the morbidity and mortality of this disease [4].

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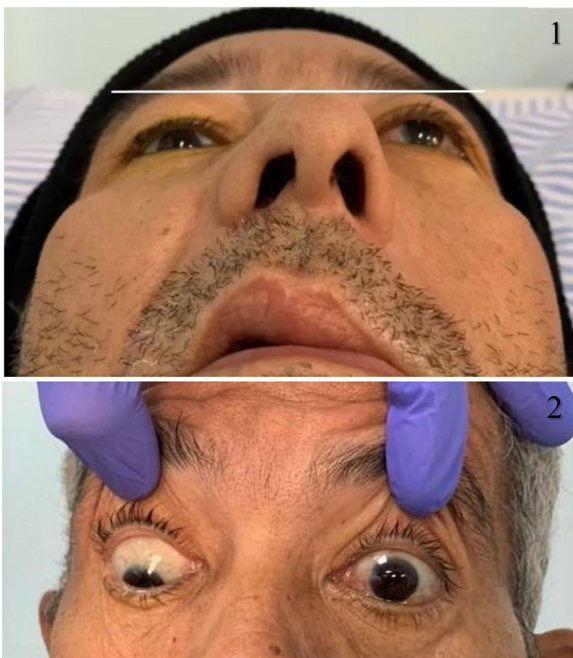
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Case report

A 65-year-old man with poorly controlled diabetes and hypertension reported progressive otalgia and otorrhea in his left ear for 8 months. He was only treated with symptomatic medications in the emergency services during this time. Five months later, he noticed loss of visual acuity in the left eye and hearing loss in the left ear, progressing soon after with left peripheral facial palsy. During the next two months, he evolved with left eye proptosis (Figs. 1 and 2), complete visual loss and ophthalmoplegia (Figs. 1 and 2), in addition to hearing loss in the right ear. He was initially evaluated by a primary care otorhinolaryngologist, who suspected the diagnosis of MEO and initiated ciprofloxacin 500 mg twice a day. After 7 days he was referred to our tertiary hospital for additional specialized diagnostic and therapeutic evaluations. On physical examination, there was a clear left eye proptosis and blindness, with absent direct photomotor reflex and preserved consensual reflex, impairment of left oculomotor, trochlear and abducent nerves, determining complete ophthalmoplegia, left peripheral facial palsy, complete hearing loss on the left and partial hearing on the right.

Considering the severity of the case, the high risk of infectious spread to the brain parenchyma and meninges and the microbiological profile of the hospital, it was opted to switch the antibiotic therapy to meropenem 2 g intravenous infusion every 8 h and vancomycin 1 g intravenous infusion twice a day.

Initial computed tomography (CT) of the head showed erosion of the anterior wall of the left external ear conduct and of the left mandible condyle and dehiscence of the external auditory canal, without other bone changes. (Fig. 3). Besides, there was a bilateral veiling of the mastoid air cells, more pronounced on the left. The magnetic resonance image (MRI) of the brain revealed a tissue occupying the left superior orbital fissure intraconal fat, with invasion of the left orbit, determining nerve compression (Figs. 4 and 5). There was involvement of the left optic, oculomotor, trochlear and abducent nerves, enhancement of the extrinsic ocular muscles and proptosis. Superior ophthalmic veins were



Figs. 1 and 2. Patient image showing proptosis with facial asymmetry and ophthalmoplegia (patient asked to look down), respectively.

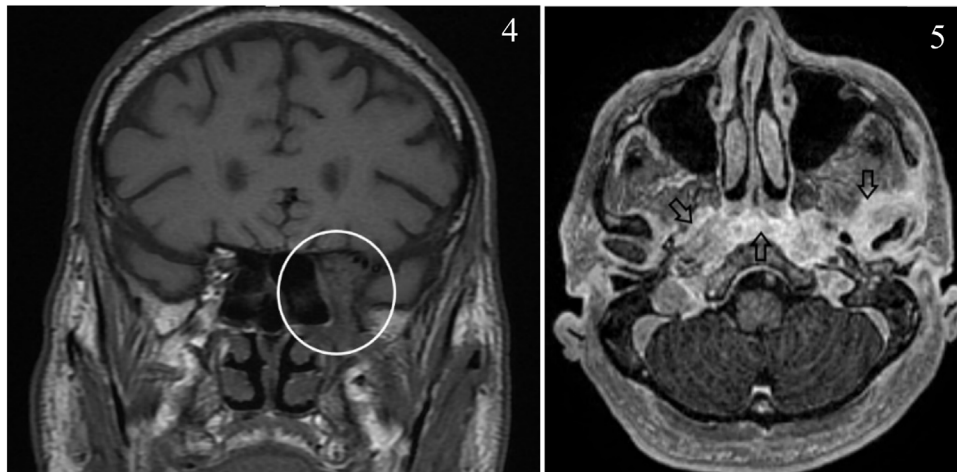


Fig. 3. CT image showing erosion of the anterior wall of the external ear conduct (arrow head) and erosion of the left mandible condyle (white arrow).

preserved bilaterally and there was no evidence of cavernous sinus thrombosis. There was also heterogeneity of the whole sphenoid bone with hyperintensity on T2/FLAIR images and contrast enhancement (Figs. 4 and 5), tissue thickening and enhancement around the left temporo-mandibular articulation, besides contrast enhancement on the trajectory of the left facial nerve.

Ten days after initiation of antibiotics (7 days of oral ciprofloxacin and 3 days of meropenem and vancomycin), the patient underwent left canal wall-up mastoidectomy, with removal of tissue samples for microbiological evaluation, including *Mycobacterium tuberculosis* and fungus. During the surgical procedure, the mastoid cavity was cleaned, an integral ossicular chain was observed, as well as the tympanic membrane and disease-free middle ear. It was also opted for the insertion of ear ventilation tube bilaterally. The external ear canal skin fragments revealed mixed inflammatory infiltrate content with necrosis areas and multinucleated giant cells; the mastoid bone material showed chronic inflammatory infiltrate content. All microbiological tests performed were negative, and this was interpreted as a possible consequence of the early use of antibiotics.

The patient had no surgical complications, presented good recover of the right ear hearing capacity after 2 days, but showed no improvements of the neurological deficits nor the left ear hearing function, probably because of the chronicity of the case. Due to clinical stability and normal laboratory parameters, after 7 days of the surgical procedure the antibiotic therapy was switched to ciprofloxacin 400 mg intravenous infusion twice a day with a further objective of oral domiciliar ciprofloxacin, in order to totalize 6 week treatment period. Unfortunately, we have been facing dramatic months of COVID-19 pandemic, causing great apprehension specially over the hospitalized patients. Although the patient in case was not in the COVID-19 hospital section, on the 14th day of hospitalization, he initiated fever, dry cough and evolved after 2 days with dyspnea, motivating oral intubation and transfer to COVID-19 intensive care unit. The reverse transcription polymerase chain reaction (RT-PCR) confirmed Sars-CoV-2 infection. As an elderly man, with poorly controlled diabetes and in current malignant external otitis treatment, the patient had a poor outcome on this new coronavirus infection and sadly passed away after 10 days. This reflects the actual reality of the world and the difficulty of managing hospitalized patients during this COVID-19 pandemic.



Figs. 4 and 5. respectively. MRI T1 image showing infiltrative soft tissue occupying the left superior orbital fissure determining the nerves compression (white circle). MRI T1 post-contrast image showing hyperintensity (arrows) in the skull base, worse on the left side.

Discussion

First described in 1968 by James R. Chandler [5], MOE is a life-threatening infection of the external ear that usually begins in the external auditory conduct and spreads through the skull base, determining a severe osteomyelitis with compromise of surrounding tissues. It typically affects the elderly diabetic patients and in up to 98 % of the cases is caused by *Pseudomonas aeruginosa* [1], an aerobic gram-negative bacteria. More virulent strains produce collagenases and elastases, being responsible for vasculitis and cranial nerve involvement, respectively. Facial nerve is the most frequently involved [3], followed by glossopharyngeal, vague and accessory nerves [2]. Involvement of the abducent nerve is rare, and even rarer is the affection of the superior orbital fissure and optic foramen, which has only been described in 2013, by Baig et al. [6]. In this scenario, there is damage to the optic, oculomotor, trochlear and abducent nerves, and the ophthalmic branch of the trigeminal nerve, which may result in ophthalmoplegia, proptosis and vision loss. In our case report, a chronic non-treated otitis externa was followed after months by hearing loss, left eye proptosis with blindness and complete ophthalmoplegia, and left peripheral facial palsy. The clinical evaluation by specialized otolaryngologist and neurologist, followed by MRI, corroborated the diagnosis of MEO with multiple cranial neuropathy, manifested by involvement of the optic, oculomotor, trochlear, abducent and facial nerves. Besides, the patient was elderly and diabetic, which are important risk factors associated with this condition. As mentioned above, orbital commitment with multiple cranial neuropathy is a rare phenotype of MEO which was only described more recently and whose early recognition and treatment are decisive in the patient's prognosis.

Conclusion

MOE is a rare, but life-threatening condition, that can evolve with neurological complications. Facial palsy is a well established one, but, to our knowledge, multiple cranial nerves involvement due to orbital fissure invasion is a rare phenotype. Greater recognition of this disease is required by the general practitioner as well as greater caution in the treatment of ear infections, especially in elderly and patients with comorbidities. Guidance on treatment, reassessment and day-to-day care, such as avoiding trauma to the external auditory canal or local humidity, makes a big difference in the treatment of the disease.

Authorship contributions

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Acquisition of data: R.Q. Silveira, V.T. Carvalho, H.N. Cavalcanti, F.C.E. Rodrigues

Analysis and/or interpretation of data: R.Q. Silveira, V.T. Carvalho, C.B. Braune, E.P.C. Ramírez

Category 2

Drafting the manuscript: R.Q. Silveira, V.T. Carvalho, H.N. Cavalcanti, F.C.E. Rodrigues

Revising the manuscript critically for important intellectual content: C.B. Braune, E.P.C. Ramírez

Category 3

Approval of the version of the manuscript to be published (the names of all authors must be listed): R.Q. Silveira, V.T. Carvalho, H. N. Cavalcanti, F.C.E. Rodrigues, C.B. Braune, E.P.C. Ramírez.

Informed consent

A written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT authorship contribution statement

Rodrigo Queiroz Silveira: Conceptualization, Data curation, Investigation, Visualization, Writing - original draft. **Viviane Tavares Carvalho:** Data curation, Investigation, Writing - original draft. **Haline Novais Cavalcanti:** Data curation, Investigation, Writing - original draft. **Fabiana Carraro Eduardo Rodrigues:** Data curation, Investigation, Writing - original draft. **Caroline Bittar Braune:** Conceptualization, Formal analysis, Methodology, Supervision, Visualization, Writing - review & editing. **Edna Patrícia Charry Ramírez:** Conceptualization, Formal analysis, Investigation, Methodology, Supervision, Visualization, Writing - review & editing.

Declaration of Competing Interest

The authors report no declarations of interest

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