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Unusual Case of Necrotizing External Otitis

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Abstract: ***A 66-year-old caucasian, diabetic male came to our emergency room with an medical treatment refractory acute external otitis. He was then admitted for a week, with resolution of the earache and the discharge; 3 months after, he recurred again to our emergency room with headache, left earache, left hypoacusis and complaining of progressive imbalance, without otorrhea. He undergone ear MRI which revealed osteomyelitis located on left petrous apex. He was again admitted with a rigorous glycemic control, antibiotherapy and hyperbaric oxygenotherapy; After 16 weeks of parenteral antibiotic treatment, the patient was discharged, and treatment was continued with oral ciprofloxacin for 4 months, leading to a radiological complete response.***

Keywords: necrotizing external otitis, malignat otitis externa, skull base osteomyelitis, hyperbaric oxygenotherapy

1. Introduction

Skull base osteomyelitis (also known as necrotizing otitis externa or malignant otitis externa) is a serious, life-threatening condition seen most commonly in older adults with diabetes (85-90%), or immunocompromised male patientsand has a high mortality rate, up to 20% [1]. Usually, it is a complication of otitis externa when repeated occurrences fail to resolve with topical treatments and aural toilet. [2]

The disease was first described as a standalone entity by Meltzer in 1959 and formally defined by Chandler in 1968; [3, 4] it is an invasive infectious disease involving the external auditory canal which extends to the skull base. The spread of the disease is thought to begin along the fissures of Santorini, through which can involve the stylomastoid foramen and the jugular foramen. In this region, it can cause severe morbidity thanks to the impairment of the cranial nerves passing through.

Most patients present with unrelenting otalgia that is disproportional to the clinical signs. There can be persistent purulent otorrhea, with an intact tympanic membrane and usually intact hearing. Otological examination may reveal edema of the external auditory canal (EAC) and the presence of granulation tissue or polyp of the EAC floor near the junction of the osseous and cartilaginous portions. [5] Patients may also present with cranial nerve deficits, which can be attributed to necrosis, neurotoxins and compression. [6]

Pseudomonas aeruginosa is involved in a high percentage of all cases of Skull Base Osteomyelitis (50–90%), while other bacterial agents, such as staphylococcal species, *Klebsiella* and *Proteus mirabilis* play a minor role. In case of fungal pathogens, *Aspergillus fumigatus* is frequently found. [7]

The optimal treatment is not clear; usually a course of intravenous treatment is followed by a course of oral antibiotics with a highly variable duration. [8]

The prognosis of this disease has improved since antibiotics were introduced, with 5-year survival rate ranging between 44% and 75%, depending on age, comorbidities, therapeutics and the extent of the lesion;

Adjuvant hyperbaric oxygen therapy has a possible role on treatment of advanced Skull Base Osteomyelitis. Cranial nerve palsies can recover completely under optimized treatment including adjuvant HBO therapy.[9]

2. Case Presentation

A 66-year-old caucasian male with medical history of arterial hypertension and type 2 diabetes under treatment with oral antidiabetics was admitted to the emergence department of otolaryngology, complaining of left otalgia and discharge without fever for 2 weeks. An external otitis was diagnosed and he was then medicated with topic drops polimixin b + neomycin + dexamethasone. At the same time, it was conducted a microbiologic exam which posteriorly came positive to *Pseudomonas aerugiona.*

On the following week, the patient returned to our emergence department with the same complaints and it was decided to associate a 8-day cycle of oral ciprofloxacin. After completing the oral cycle and 2 weeks of topic antibiotherapy, the patient persisted with aural discharge and otalgia and was admitted to our ward to complete an endovenous antibiotic treatment with ciprofloxacine. After 8 days of treatment, the patient had no complains of pain, discharge or other. The CT scan did not reveal structural irregularities or signs of inflammatory disease (Fig 1). As a result, the patient was discharged.

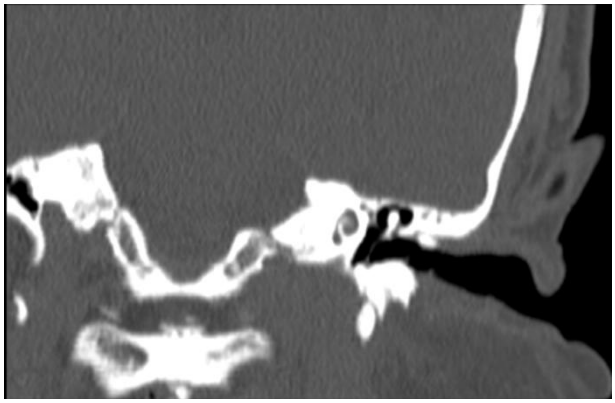

Figure 1: CT scan: No morphological or soft tissues density variations of the external auditory channels, and there is also no anomalous abstraction of the contrast product.

One month after, the patient was admitted in the vascular surgery ward for ischemic stroke and undergone a left carotid endarterectomy with patch. The patient was hospitalized for 30 days due to an infection of the surgical wound.

On the following month, the patient gradually started complaining of headache and left earache associated with gradual left deafness and unsteadiness without purulent discharge or fever. He was evaluated in a following otolaryngologic consultation where a cranial MRI was requested, which revealed a left cranial base/petrosal apex osteomyelitis (Fig. 2 and 3).

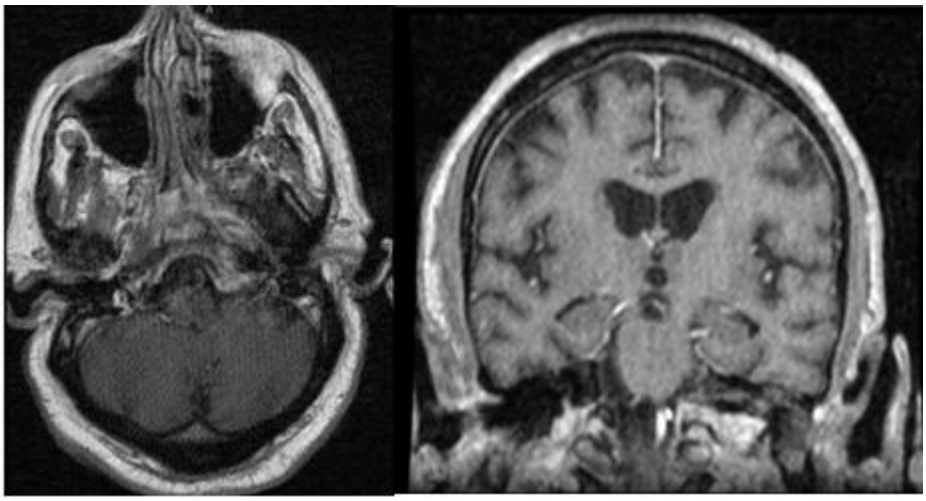

Figure 2 and 3: MRI: Involvement of the left petrous apex region / skull base and posterior tissues of the nasopharynx by tissue area with intense contrast product capture that represents inflammatory / infectious process in the context of probable osteomyelitis.

The patient was again admitted in our department with a diagnosis of necrotizing external otitis and treated with endovenous ceftazidime 2g 8-8h, aural toilet and topic antibiotherapy with ofloxacin 12-12h (even without otologic infection signs) and hyperbaric oxigenotherapy. An insulin regimen was used to achieve a rigorous glycemic control.

An audiometry was done, which revealed a profound left neurossensorial deafness (fig 4). A brainstem evoked response audiometry was also made which revealed an electrophysiologic threshold of 80-90dB on the left side (fig 5). The videonistagmography revealed left vestibular arreflex.

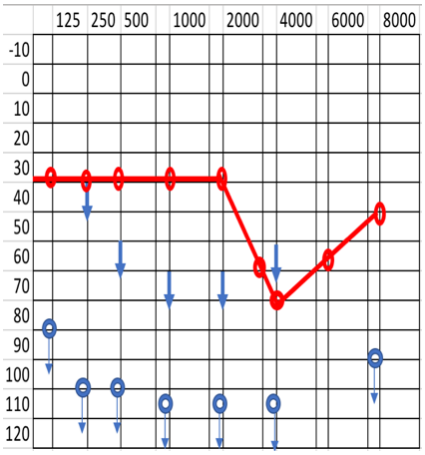

Figure 4: Audiometry: Profound left neurosensorial deafness.

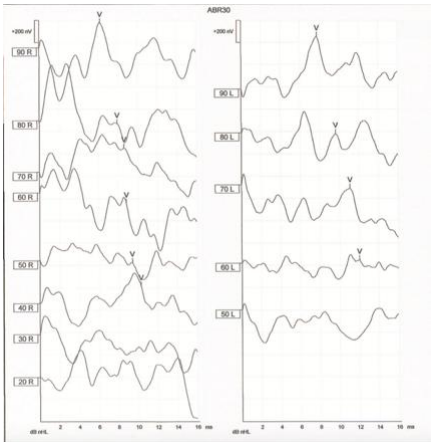

Figure 5: Auditory evoked potentials threshold of 40dB on the right side and 60dB on the left side.

After 4 weeks of treatments, a cranial Ga-67 scintigraphy was conducted which revealed local osteomyelitis (Fig 6) and an avid captation on the left clivus (Fig 7). A biopsy, realized under general anesthesia, only revealed inflammatory tissue. The microbiological exam did not reveal any pathological microorganism.

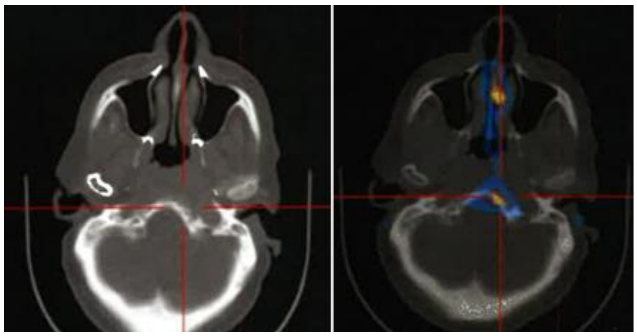

Figure 6 and 7: Cranial Ga-67 scintigraphy and CT: A slightly increased avidity focus for Ga-67 in the left clivus that may be related to active osteomyelitis at this site.

On the 8th week of treatment, the patient had more imaging exams performed.

This time a PET-CT with F-18-FDG was completed and revealed glycolytic hypermetabolism on the inferior part of left sphenoid sinus and active osteomyelitis on left clivus (SUV Max on late study= 7) (Fig. 8).

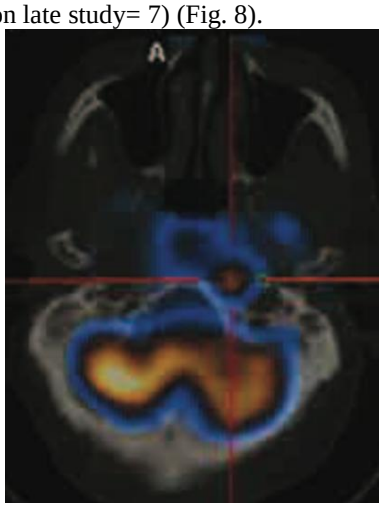

Figure 8: PET-CT with F-18-FDG: glycolytic hypermetabolism on the inferior part of left sphenoid sinus and active osteomyelitis on left clivus (SUV Max on late study= 7);

The PET-CT with F-18-FDG was repeated on the 12th week of treatment, presenting glycolytic hypermetabolism on the inferior part of left sphenoid sinus, but with a reduction of metabolic activity comparing with the previous exam with SUV max on late study= 5,7. (Fig 9)

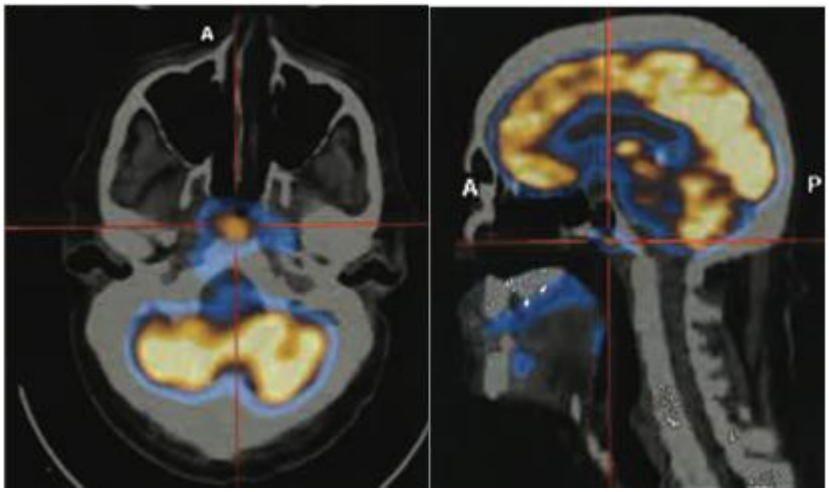

Figure 9 and 10: PET-CT with F-18-FDG: glycolytic hypermetabolism on the inferior part of left sphenoid sinus, but with a reduction of metabolic activity comparing with the previous exam with SUV max on late study= 5,7;

We repeated the imaging evaluation on a regular schedule of 4 weeks and at the 16th week we reevaluated again with PET-CT with F-18-FDG, revealing a step reduction of the metabolic activity on the left clivus (SUV Max on late study= 3,6) (Fig 10 e 11)

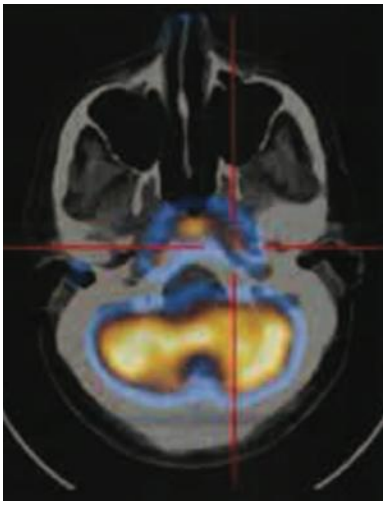

Figure 11: PET-CT with F-18-FDG: step reduction of the metabolic activity on the left clivus (SUV Max on late study= 3, 6)

During his stay in the hospital, the patient completed in our hospital, 60 sessions of hyperbaric oxigenotherapy at 2 atmospheres for 1, 5 hour each session. The patient was discharged after 16 weeks, with evidence of a good response to the treatment and clinically asymptomatic.

Outcome and Follow-Up

On the follow-up, the patient completed another 12 weeks of ciprofloxacin treatment (500mg 12-12h), after that another PET-CT with F-18-FDG which did not reveal metabolically active disease avid to FDG, suggesting complete response to the treatment (Fig 12 and 13).

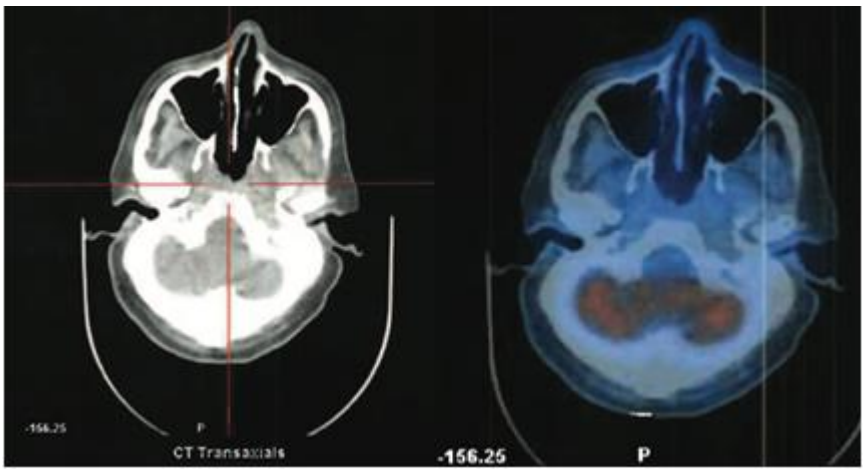

Figure 12 and 13: PET-CT with F-18-FDG: no signs of metabolically active disease avid to FDG

The patient has been through a 24-month follow-up and was advised to keep a rigorous glycemic control and starting a vestibular rehabilitation training. He remained with neurossensorial deafness and softer unbalance complaints.

3. Discussion

We described the case of skull base osteomyelitis with a progressive VIII cranial nerve injury in a patient with multiple confounding factors such as a previous diagnose of external otitis apparently successfully treated and a carotid patch implant in the context of a stroke with subsequent wound infection.

Skull base osteomyelitis as external otitis complication (previously known as malignant external otitis) is a rare condition. It is more common in humid and warm climates as the external otitis is. This affects the external auditory canal and the temporal bone, and it can affect several cranial nerves. The facial nerve is by far the most commonly affected, usually in the stylomastoid foramen. Other cranial nerves can be affected as IX, X and XI at the jugular foramen, XII at the hypoglossal canal, or even V and VI at the petrous apex. Though uncommon, the VIII have been reported as possibly affected on a skull base osteomyelitis [10] Extremely unusual is the VIII nerve affection without any other cranial nerve palsy.

It is important to understand that the infectious spread of the skull base osteomyelitis does not occur in a standard pattern, and the understanding and recognition of anatomical structures is vital to understand the pathology. The most common direction of Skull Base Osteomyelitis is expansion through the temporal bone with destruction of the temporomandibular joint or erosion of clivus (80%). [11]

The skull base osteomyelitis appears as a progression of the ear disease along the fissures of Santorini on the external ear canal.

The presented case is atypical on its presentation because it was diagnosed 3 months after the patient was admitted for endovenousantibiotherapy for an external otitis resistant to oral and topic treatment. At that time, the patient was considered cured because no signs of otitis were apparent and, at the same time, the patient was without symptoms and the CT control exam was normal. We believe that the infection was latent on the temporal bone and was after reactivated. The osteomyelitis location reinforces this hypothesis of external otitis complication as it was on the left temporal bone without any other known infection of

other possible origin. The patch complication hypothesis was dismissed after considering the pathologic history and anatomy. [11, 12]

As established, there is a clear difficulty on the diagnosis of necrotizing external otitis due to nonspecific appearance of the process on the traditional imaging tools like MRI or CT. As lesions can consist of soft tissue abnormalities and bony erosions, as well as the dynamic process of inflammation, an optimal imaging modality has to be found. CT and MRI are used for anatomical imaging, whereas nuclear techniques are used for the functional process. Hybrid techniques such as PET-CT that combine anatomical and functional biomarkers are important on detecting disease extent and evaluating the treatment response.

There is no consensus on the treatment of skull base osteomyelitis. Its advocated by most that aural toilet, meticulous glycemic control, systemic and ototopic antimicrobial therapy have a central role in the treatment. [13, 14] At the same time, there is a discussion on the role of hyperbaric oxygen therapy. The choice of antibiotic, and the duration of the treatment, are also not consensual. After infectiology and microbiology counselling, we decided to use ceftazidime (2g 12-12h). Ceftazidime has a good bactericidal activity against *pseudomonas aeruginosa*, [15] which is the most common causative agent of necrotizing external otitis and was the agent isolated for the first time on our patient.

At the same time, we started a hyperbaric oxygen treatment in our hospital. 60 sessions were undertaken, with pressures of 2,5 atmospheres of 100% oxygen for 1,5 hours.

At this time, there is still insufficient evidence to make clear recommendations about the use of hyperbaric oxigenotherapy on necrotizing external otitis but there are some studies that suggest the benefit of using this as adjunct treatment. [9, 16]

The patient was only discharged after 16 weeks of endovenous ceftazidime and 60 hyperbaric sessions. We decided to end the protocol after a clear evidence of a good response to the treatment using the CT-PET with F-18 FDG. After that, and together with infectious disease department, we decided to institute a cycle of oral ciprofloxacin that the patient ended after there was no disease evidence (12 weeks later).

The patient is still being followed after 24 months and has remained with neurossensorial deafness and softer unbalance complaints but without other major complaints.

4. Conclusion

Skull base osteomyelitis as an external otitis complication remains a serious invasive infection. The diagnosis is challenging, which sometimes delays the treatment. The presentation of the disease is not always linear, making it important to be aware of high-risk groups, such as elderly individuals with diabetes, patients on chemotherapy or in immunocompromised states.

Pseudomonas aeruginosa is the bacteria most commonly isolated. As a result, the treatment is completed with antibiotics with sensitivity against it, such as ceftazidime or ciprofloxacin. Hyperbaric oxygenotherapy is argued to increase the efficacy of the treatment so it can be used together with antibiotics. However, none of these treatments should be carried on without a rigorous glycemic control and aural toilet.

The follow up of these patients must be rigorous with imaging control until there is evidence of cure.

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