Malignant otitis externa - a case report

Nguyen Thi Phuong Thao[1]*, Dao Trung Dung[2], Ho Chi Thanh[1] and Nguyen Van Huu[1]

[1]108 Military Central Hospital, [2]Bach Mai Hospital

Summary

Malignant or necrotizing otitis externa (MOE) is a severe infectious condition of the external ear canal. It is rare but has a high mortality rate due to complications such as osteomyelitis of the skull base and intracranial involvement. The reported clinical case involves a 72-year-old male patient with a long history of type 2 diabetes, who presented with severe ear pain, stenosis of the external ear canal, and no response to medical treatment one month prior to hospitalization. The CT scan showed bone destruction of the skull base and part of the external ear canal. Despite being treated aggressively with radical mastoid surgery and broad-spectrum antibiotics, the patient's condition worsened, leading to complications including cranial nerve paralysis, meningitis, and ultimately death. This report aims to analyze the clinical and paraclinical characteristics and progression of the disease to draw lessons for the diagnosis and treatment of this dangerous condition.

Keywords: Malignant otitis externa, the external auditory canal.

I. BACKGROUND

Malignant otitis externa (MOE) is a severe infection that originates in the external auditory canal and subsequently spreads to nearby structures, particularly leading to skull base osteomyelitis, and causes dangerous complications such as meningitis, brain abscess, and septic thrombophlebitis of the venous sinuses. Its prevalence ranges from 0.221 to 1.19 cases per 100.000. The disease is primarily bacterial, especially involving *Pseudomonas aeruginosa*, though in rare cases it may be caused by fungi[1].

MOE mainly affects elderly individuals with compromised immune systems, such as uncontrolled diabetes, HIV/AIDS, or those on immunosuppressive medications (post-organ transplant, etc)[1]. The condition was first recognized in 1838 when Toulmouche reported a case of temporal bone osteomyelitis[2]. In 1968, Chandler JR introduced the term "malignant otitis externa" to highlight the disease's dangerous nature, with a mortality rate reaching approximately 50%[3]. Later, the term "necrotizing otitis externa" was suggested to avoid confusion with true malignant diseases. Both terms are still used interchangeably today.

Common symptoms of MOE include ear pain, otorrhea, edema, granulation tissue observed in the exteranal auditory canal and a sensation of fullness in the ear, often leading to misdiagnosis as acute otitis externa, especially in the early stages. Despite advances in treatment, MOE is still a potential devastating condition that continues to pose a diagnostic and therapeutic challenge. This report describes a case of MOE, analyzing the clinical, paraclinical features, and disease course to draw lessons for diagnosis and treatment.

II. CASE PRESENTATION

T.D.T - A 72-year-old male presented to the hospital with a 20-year history of diabetes treated with insulin injections. He had suffered a stroke one year prior, resulting in residual right hemiparesis and left peripheral facial paralysis. The patient also had a history of endoscopic surgery for fungal maxillary sinusitis one month before admission, which had

Received: *15 November 2024,* ***Accepted***: *28 December 2024*

***Corresponding author**: drthao108@gmail.com - 108 Military Central Hospital*

resolved (No nasal discharge and a clean middle nasal meatus on endoscopy). The main reasons for presentation was his left-sided severe headache, accompanied by ear pain and foul-smelling ear discharge on the same side for one month. He had been prescribed with oral and topical antibiotics at a local clinic, but the symptoms persisted.

At the time of admission, the patient was alert, afebrile, without signs of meningitis, and had left peripheral facial paralysis grade IV due to previous history of stroke (according to House-Brackman Classification), with no clear signs of propression as reported by the patient's family. He experienced severe pain on the right side of his head, right side of his face, and the entire right ear, with a VAS score of 7-8. On examination, the right auricle showed edema and moist inflammation, the left external ear canal was narrowed, preventing visualization of the tympanic membrane while the skin over the mastoid was normal, mild trismus and dysphagia. The tympanic membrane on the contralateral ear appeared to be intact and thickened. Blood tests revealed an increased white blood cell count of 14.7G/L, neutrophils at 75%, CRP at 114mg/L, glucose level at 24.2mmol/L, and HbA1c at 13.5%.

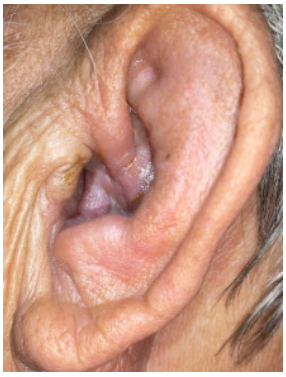

Figure 1. Imflammation of the left auricular

Initially diagnosed was inflammation and stenosis canal of the external auditory canal in a patient with a history of stroke. The patient was treated aggressively with intravenous levofloxacin 750mg per day and daily ear toilet. Intravenous and oral paracetamol- tramadol and subcutaneous 20UI insulin 3 times/day were prescribed additionally. After treatment with insulin, glucose in blood in within the normal limits. However, after two weeks, the patient continued to experience severe pain, and the canal stenosis remained unchanged and the tympanic membrane could not been observed. A temporal bone CT scan showed a fully stenose ear canal, fluid and inflamed tissue in the mastoid and middle ear, a thickended left side nasopharynx mucosa, extensive bone erosion at the tegmen and the temporomandibular joint. MRI showed irregular thickening of the nasopharyngeal on the left side.

The patient underwent radical mastoidectomy in the third week of admission, revealing ulcerative inflammatory lesions and fungal involvement confirmed by Grocott staining of middle ear tissues.

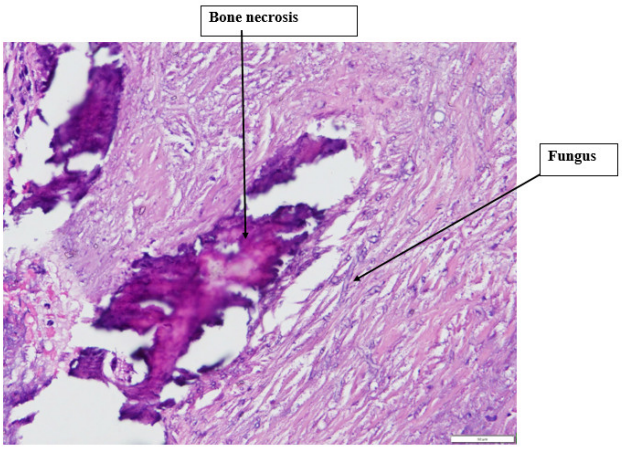

Figure 2. Histopathology of middle ear tissue

Despite treatment with intravenous ceftazidime 6g/day, pain management, and continued blood glucose control, the patient's condition deteriorated after 10 days with increasing dysphagia and multiple cranial nerve palsies. CRP increased at the level of 290mg/L. The CT scan showed difuse brain edema corresponding to the blood supply regions of the anterior and middle cerebral artery on both sides. The patient then developed pneumonia, bacterial meningoencephalitis, recurrent stroke, and ultimately died at the 30th day of hospitalization.

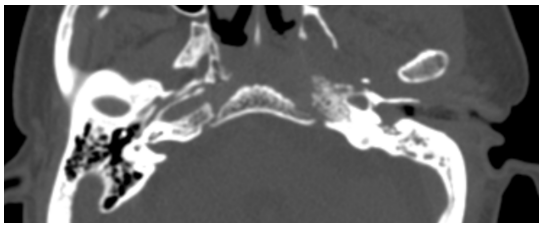

Figure 3. Preoperative temporal bone CT scan

III. DISCUSSION

Malignant otitis externa is a rare disease with a high mortality rate that requires timely diagnosis and treatment. A systematic review by Gonzalez and colleagues indicates that common symptoms include severe ear pain unresponsive to standard pain relief, ear discharge, hearing loss, temporomandibular joint pain, and headaches. However, due to the symptoms resembling those of other common external ear infections, the diagnose is ofter delayed, especially when neurological symptoms become pronounced. Consequently, Cohen and Friedman proposed definitive diagnostic criteria, emphasizing key indicators such as disproportionate pain relative to the clinical findings, edematous external ear canal, discharge, granulation tissue, numerous micro-abscesses observed during surgery, and bone scans with Tc-99 MDP showing strong uptake in lesions, which do not respond to local treatment lasting over a week. Additional criteria may include diabetes, cranial nerve palsies, bone erosion on imaging results, cachexia, and old age[4]. Symptoms of cranial nerve palsies, particularly facial nerve involvement, are complications of the disease and are considered poor prognostic factors[5].

Our case aligned with the epidemiological characteristics described in the literature, demonstrating that in elderly patients with uncontrolled diabetes who present with acute external otitis symptoms such as ear pain and discharge, malignant otitis externa should be considered.

In this patient, due to presenting at a late stage, the characteristic sign granulation tissue found at the junction of the cartilaginous and bony ear canal—was replaced by a stenosed ear canal. In addition, the main symptoms such as severe headache, ear discharge, and swelling of the external ear canal had not improved with local treatment and had been present for about a month before hospitalization. Because of the diffuse nature of the pain along with the coexisting sinus pathology, the patient was initially diagnosed with headache due to sinusitis and was referred for endoscopic sinus surgery. However, the symptoms did not improve afterward.

Among the other symptoms of the disease, notable is the condition of cranial nerve paralysis. In this patient, due to the sequelae of a previous stroke, there were signs of peripheral paralysis of the VII cranial nerve on the same side as the lesion, as well as pre-existing cranial nerve damage. As a result, this group of neurological symptoms was not grouped with the newly appearing ear symptoms. This comorbid factor made the diagnosis more difficult and delayed. Signs of small abscesses were found during the radical mastoidectomy. Here, due to the severity and fairly typical nature of the disease, the patient had not undergone a bone scan using Tc-99m MDP.

According to the consensus of the British ENT Association in 2020, a patient is diagnosed with malignant otitis externa (MOE) if all of the following criteria are met: Ear pain with ear discharge, granulation tissue or external ear canal inflammation, histopathological results excluding malignant pathology, CT scan showing bone destruction of the external auditory canal, soft tissue involvement of the external ear canal, or MRI showing edema of the temporal bone marrow. Clinically, this patient met all the diagnostic criteria and diagnose wass confirm[6].

In MOE, imaging diagnosis is a useful tool for both diagnosis and prognosis. Peleg and colleagues classified MOE into two forms: “severe” and “non-severe” by scoring the imaging signs on CT scans[7]. However, the drawback of this scale is that it does not evaluate the progression of the disease. Later, Tengku, based on the results of temporal CT scans and bone scans, divided MOE into five stages[8]. Stage I involves localized inflammation in the soft tissue of the external ear canal, stage II includes inflammatory bone lesions confined to the mastoid bone, stage III shows invasive inflammatory lesions penetrating inward, infiltrating the petrous bone and the temporomandibular joint, with or without accompanying tissue in the oropharyngeal region.

In stage IV, lesions extends to the nasopharyngeal area, with or without abscesses. Finally, in stage V, the inflammatory process spreads to the contralateral skull base. According to Tengku's grading scale, this patient presented to us in stage IV, where the CT scan showed destruction of the petrous bone, the body of the sphenoid bone, the left lateral skull base, and infiltrative inflammation in the nasopharyngeal area on the same side. This is a severe stage with a high mortality rate.

The causative agents of MOE can be bacteria or fungi. Among them, Pseudomonas aeruginosa is the primary pathogen, accounting for over 95% of cases. Identifying the causative microorganisms plays an important role in conducting antibiotic susceptibility tests, which helps to adjust the appropriate antibiotics. However, in practice, approximately 13% of bacterial cultures may be negative[9]. In this patient, the negative result could be due to the prolonged use of antibiotics and local ear drops prior to the culture, which may have prevented the identification of the causative bacteria. The patient was also completely ruled out for suspected cancerous lesions when the postoperative pathology results indicated acute inflammatory ulcers and the biopsy of the ipsilateral mucosa showed no malignant cells. A biopsy sample from the middle ear was then stained with Grocott, resulting in a positive finding for Aspergillus.

Treating MOE is always a challenge for clinicians, even in developed countries. In principle, the treatment regimen needs to be comprehensive, involving systemic and local interventions, blood glucose strictly controlled, supportive care, and functional rehabilitation[10]. Surgery is also indicated in cases where medical treatment is ineffective, primarily to remove necrotic tissue, drain abscesses, and collect infected bone for pathology test. In patient T.D.T, we recommended surgery due to the worsening local ear inflammation despite intensive medical treatment. Additionally, surgery would allow for the removal of the pathological tissue and help exclude malignant conditions. After undergoing a radical surgery, there were significant improvements in clinical symptoms, such as reduced headache and less ear discharge. The antibiotic regimen used for the patient in the early stages, given the negative microbial culture results, was based on general recommendations: Ceftazidime 6g/day in combination with intravenous levofloxacin. Due to the patient's severe condition, exhaustion, and limited experience in treating fungal diseases, we did not administer antifungal medications. Additionally, the patient's blood glucose levels were closely monitored and adjusted in collaboration with the endocrinology department, along with continuous supportive care using nutritional solutions. After an initial period of relative improvement, the patient quickly deteriorated, especially when experiencing gastroesophageal reflux episodes leading to aspiration pneumonia, and showing signs of meningitis such as fever, neck stiffness, and a positive Rivalta reaction on cerebrospinal fluid analysis. The patient's condition then worsened rapidly, resulting in death when lung function was lost, along with diffuse brain edema due to extensive bilateral cerebral infarction and occlusion of the feeding vessels of the anterior circulation and both middle cerebral arteries.

It can be said that this patient belongs to the severe disease group with a poor prognosis right from the first day of hospitalization, based on the criteria in the literature. Specifically, according to Tengku's imaging diagnostic criteria, at the time of diagnosis, the patient was in stage IV-V with multiple lesions at the ipsilateral skull base and beginning to affect the contralateral side. At the time of admission, signs of VII cranial nerve paralysis and other cranial nerves such as IX and X causing aspiration and difficulty swallowing were also indicators of a severe prognosis[8]. Additionally, according to Stevens, the presence of fungi in the affected tissue tends to lead to rapid deterioration of the disease, increasing the mortality rate from 0% in the group without fungi to 46% in the group where fungi were found. In this patient, the fungus identified in the tissue was Aspergillus[11]. The fact

that antifungal antibiotics had not yet been used could also be a factor contributing to the rapid progression of the disease.

IV. CONCLUSION

Malignant otitis externa is a severe and dangerous condition of the external auditory canal. The diagnosis should be considered when severe ear pain and headaches are disproportionate to clinical findings and unresponsive to treatment, particularly in high-risk patients (elderly, immunocompromised). Imaging in cluding CT-scan, MRI and Tc-99m are crucial for diagnosis and staging as. Prognostic factors include comorbidities such as diabetes and liver/kidney dysfunction, and neurological or endocrine complications should be carefully monitored. Comprehensive, multidisciplinary care is essential for favorable outcomes in this challenging disease.

REFERENCES

1. González JLT, Suárez LLR, de León JEH (2021) *Malignant otitis externa: An updated review*. American Journal of Otolaryngology 42(2): 102894.
2. Kumar S, Singh U (2015) *Malignant otitis externa: A review*. J Infect Dis Ther 3(204): 2332-0877.1000204.
3. Chandler JR (1968) *Malignant external otitis*. The Laryngoscope 78(8): 1257-1294.
4. Cohen D, Friedman P (1987) *The diagnostic criteria of malignant external otitis*. The Journal of Laryngology 101(3): 216-221.
5. Marina S, Goutham M, Rajeshwary A, Vadisha B, Devika T (2019) *A retrospective review of 14 cases of malignant otitis externa*. Journal of otology 14(2): 63-66.
6. Hodgson SH, Khan MM, Patrick-Smith M et al (2023) *UK consensus definitions for necrotising otitis externa: A Delphi study*. BMJ open 13(2): 061349.
7. Peleg U, Perez R, Raveh D, Berelowitz D, Cohen D (2007) *Stratification for malignant external otitis*. Otolaryngology Head Neck Surgery 137(2): 301-305.
8. Kamalden TMIT, Misron K (2021) *A 10-year review of malignant otitis externa: Anew insight*. European Archives of Oto-Rhino-Laryngology: 1-8.
9. Takata J, Hopkins M, Alexander V et al (2023) *Systematic review of the diagnosis and management of necrotising otitis externa: Highlighting the need for high-quality research*. Clinical Otolaryngology 48(3): 381-394.
10. Al Aaraj MS, Kelley C (2023) *Necrotizing (Malignant) Otitis Externa*. StatPearls [Internet]. StatPearls Publishing.
11. Stevens SM, Lambert PR, Baker AB, Meyer TA (2015) *Malignant otitis externa: A novel stratification protocol for predicting treatment outcomes*. Otology & Neurotology 36(9): 1492-1498.