

Squamous Cell Carcinoma Of The Ear Masquerading As Malignant Otitis Externa

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While both acute and chronic otitis externa represent conditions that are encountered commonly by primary care physicians, malignant otitis externa can be a diagnostic and therapeutic challenge to generalists and subspecialists alike. Malignant otitis externa is a devastating infection of the external auditory canal, which begins as a pseudomonas dermatitis but can progress to an invasive vasculitis and osteomyelitis of the base of the skull. The disease is associated with numerous complications and requires extensive treatment. Whereas the severity of the external otitis infection can range from acute to chronic to malignant, the possibility of other disease, including carcinoma, must be considered when evaluating unresolving external otitis. Squamous cell carcinoma has many of the same symptoms as malignant otitis externa, and the persistence of symptoms after antibiotic treatment should provoke further investigation to rule out the possibility of an underlying neoplastic lesion. The following case presentation illustrates the need for careful evaluation and follow-up in patients who have apparent malignant otitis externa.

Case Report

A 70-year-old woman with a history of chronic left otitis externa was admitted to the hospital with worsening pain of the left ear, persistent drainage of the left ear, and the new onset of left-sided facial weakness, headaches, and disequilibrium. The patient had been followed by an otolaryngologist for 7 years for chronic otitis externa, and she continued to receive treatment with topical corticosteroid solutions. One week before her hospitalization she had a left earache with a purulent discharge from the ear canal and was treated with corticosteroid ear drops for otitis externa. On the day prior to hospitalization, she

developed increased epiphora of her left eye, and on the day of admission, she awakened with a left-sided facial droop.

On initial physical examination, the patient had a purulent discharge from the left external auditory canal. The canal itself contained material that appeared to be granulation tissue. The tympanic membrane could not be visualized because of the presence of this material and the concurrent purulent fluid. The left mastoid process was tender to palpation, but no periauricular edema or mass was noted. With the Weber test, the sound was heard best in the left ear. The right ear was normal in appearance. The left eye had severe ptosis with an ectropion and exposure keratitis. On neurological examination, she had palsy of the left cranial nerve with weakness of the muscoli frontalis, orbicularis oculi, and orbicularis oris. Decreased taste sensation was also noted.

Admission laboratory data showed her white cell count was $7.7 \times 10^9/L$ (7700 mm^{-3}) with a normal differential. Hemoglobin, hematocrit, platelet count, and chemistry profiles were all within normal limits, as was a urinalysis. The erythrocyte sedimentation rate (Westergren) was 15 mm/hr.

The patient was admitted to the family medicine inpatient service, and an otolaryngologist was consulted. A computed tomographic scintiscan of the head was obtained, as advised, which showed extensive asymmetric sclerosis of the left mastoid air cells with opacification of the left mastoid air cells, left middle ear cavity, and most of the left external auditory canal. The official radiological impression included extensive chronic mastoiditis, otitis media, and malignant otitis externa (Figure 1), and at the recommendation of the otolaryngologist, the patient was administered ciprofloxacin and rifamycin.

After the discharge from the ear was cultured, the patient's condition was presumptively diagnosed as malignant otitis externa. Although the left facial nerve paralysis was nearly complete, the Hilger nerve stimulation test failed to show any

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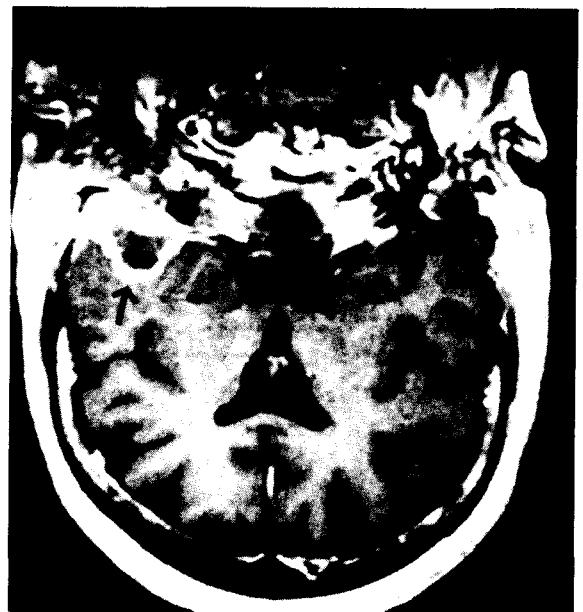


Figure 1. Scintiscan of the head. Left: axial view. Right: coronal view.

evidence of nerve degeneration. Within 6 days of antibiotic treatment, the facial nerve palsy resolved completely. Culture results from the ear showed the presence of *Staphylococcus aureus*, streptococci group G, and a rare *Pseudomonas* that was sensitive to trimethoprim and sulfamethoxazole, so this antibiotic was added. With continued treatment, the patient's ear pain, erythema, and drainage resolved.

Initial biopsy on the day of admission showed only acantholysis and failed to provide a diagnosis; repeat biopsy performed on day 10 of her hospital stay showed squamous cell carcinoma of the left external auditory canal and temporal bone. Subsequent magnetic resonance imaging showed tumor erosion and mass effect on the left temporal bone, which extended into the floor of the left middle cranial fossa.

Management of the squamous cell carcinoma was discussed among the otolaryngologic, neurosurgical, and hematology-oncology consultants.

Because of the risks of surgical resection and the poor prognosis for surgical cure, palliative radiation therapy and chemotherapy were chosen. The patient was scheduled for treatment with mitomycin-C and 28 full courses of radiation therapy for a total of 5040 rads.

Otitis Externa

Acute Otitis Externa

Otitis externa is a clinical entity commonly seen in primary care practice. It is particularly common in the pediatric age group and among swimmers. In the infant, external otitis is often caused by a variety of organisms (Table 1). Swimmer's ear is caused by the loss of the protective cerumen and chronic irritation and maceration of the epithelial lining as a result of persistent and excessive moisture in the ear canal. *Pseudomonas* is the organism most commonly isolated. Herpes and varicella viruses are often involved.¹

Earache is a symptom in diffuse otitis externa, which is accentuated by movement of the external ear, especially with pressure on the tragus. Itching often precedes or accompanies the pain. Edema, redness, and serous or purulent discharge are common features. Edema of the skin and tympanic membrane in combination with skin thickening and serous or purulent secretions can produce a conductive hearing loss. Secondary infection can produce lymphade-

Table 1. Common Organisms Associated with Otitis Externa.

Bacteria	Fungi	Viruses
<i>Pseudomonas aeruginosa</i>	<i>Candida</i>	Herpes
<i>Enterobacter aerogenes</i>	<i>Aspergillus</i>	Varicella
<i>Proteus mirabilis</i>		
<i>Klebsiella pneumoniae</i>		
<i>Staphylococcus epidermis</i>		

nititis with preauricular or postauricular lymphadenopathy.^{1,2} Treatment generally consists of instilling a suspension of polymyxin-B, neomycin, and hydrocortisone (Cortisporin™) in the canal four times daily for 10 to 14 days. Oral antibiotic therapy can be used if the patient is febrile or if lymphadenitis or cellulitis is present.

Chronic Otitis Externa

Chronic otitis externa appears as a continuum of clinical events resulting from nonresolving acute otitis externa. Variables that promote progression of the disease include water exposure and the presence of a fungus or other resistant organism within the auditory canal. Treatment failure can lead to persistent symptoms accompanied by scaling, debris, or granulation tissue formation, which can cause sloughing of the epithelial layer and chronic drainage.²

Malignant Otitis Externa

Malignant otitis externa is a term used for a severe, invasive bacterial infection of the external ear canal. It frequently progresses to osteomyelitis of the base of the skull and can result in cranial neuropathies. The condition typically occurs in elderly diabetic or immunocompromised patients and is caused almost exclusively by *Pseudomonas aeruginosa*. The clinical course of the cranial neuropathies can best be understood by examining the sequential anatomical involvement of the infection.³ The organism characteristically spreads from the external auditory canal into the temporal bone and then through the fissures of Santorini in the external meatus to the mastoid. The facial nerve (cranial nerve VII) is often involved at this location as it exits the stylomastoid foramen. The infection can then proceed along the base of the skull to affect the glossopharyngeal (IX), vagus (X), and spinal accessory (XI) nerves in the region of the jugular foramen. Less commonly, the hypoglossal nerve (XII) can be affected at the hypoglossal canal; the abducent (VI) and trigeminal (V) nerves at the apex of the petrous portion of the temporal bone; and the optic nerve (II) at the orbital apex. Because of its proximity to the site of initial infection, the facial nerve is the first and most commonly affected.

Table 2. Differential Diagnosis of the Inflamed Ear.

Condition	Diagnosis
Infections	Otitis externa
	Furunculosis
	Mastoiditis
	Otitis media, chronic suppurative
	Acute cellulitis
Dermatoses	Herpes simplex
	Herpes zoster oticus
	Bullous myringitis
	Keratitis obturans
Radiation-induced necrosis	
Cerebral spinal fluid leakage	
Neoplasms of the external auditory canal	

Clinical Presentation

Malignant otitis externa can cause a severe otalgia that is characteristically more intense than the pain of acute otitis externa or otitis media. Severe headaches located in the temporal, occipital, or temporomandibular joint region can be present, too. The external ear canal is usually characterized by erythema, edema, a purulent otorrhea, and the presence of granulation tissue within the canal.³ Tenderness and edema in the periauricular region can also be present.

Treatment

Although controlled clinical trials have not been used to evaluate the best antibiotic therapy, the recommended treatment for malignant otitis externa has traditionally been parenteral antibiotics. The most widely accepted therapy consists of an aminoglycoside combined with a second intravenous antipseudomonal agent, with the course of therapy lasting from 4 to 12 weeks. More recently, the oral antibiotics ciprofloxacin and rifampin in combination have produced good results.⁴

Table 3. Clinical Features of Malignant Otitis Externa.

Severe otalgia
Purulent otorrhea
Periauricular tenderness
Periauricular edema
Erythema of external auditory canal
Edema of external auditory canal
Granulation tissue within external auditory canal
Severe headaches
Cranial neuropathies

Squamous Cell Carcinoma of the External Auditory Canal

Squamous cell carcinoma is the most common neoplasm of the external auditory canal and temporal bone. Tumors of the temporal bone are thought to originate in the external auditory canal rather than from the bone itself.⁵ Chronic inflammation such as that resulting from chronic otitis externa can be a predisposing factor. The first symptoms of carcinoma of the auditory canal include otalgia, otorrhea, and pruritus. Late symptoms include vertigo, trismus, sensorineural hearing loss, and cranial nerve paralysis.⁵

The path of tumor extension follows a course similar to that of infectious spread. Tumors that originate in the external auditory canal can spread to the cartilaginous portion of the canal and extend through the fissures of Santorini, leading to the involvement of the parotid gland or temporomandibular joint. An advancing tumor can extend into the mastoid region or extend directly through the tympanic membrane to the middle ear, eustachian tubes, and the cranial nerves.⁵ Again, the facial nerve is most commonly affected, but the glossopharyngeal or vagus nerve can also be affected at the jugular foramen. An extensive tumor could involve the entire temporal bone. Metastatic spread is confined usually to the preauricular nodes first, with subsequent spread to the cervical nodes.

Of the malignancies of the external auditory canal, only those that arise from the skin of the pinna and extend locally have a fair prognosis. Tumors that arise deep within the external auditory canal have a universally poor prognosis, and surgery and radiation therapy are largely palliative. Only those lesions isolated to the soft tissues of the external auditory canal have a reasonable chance of surgical cure.⁶

Discussion

The case presented envelops the broad spectrum of disease related to the chronically draining ear, as well as the potential pitfalls of presumptive diagnoses. Initially, such a "wet ear" is presumed all too often to represent an acute otitis externa. When symptoms persist for weeks despite therapy, the working diagnoses might then be changed to chronic otitis externa. The otalgia; otorrhea, erythema, and edema of the external auditory canal; and periauricular tenderness and

edema might all be consistent with this diagnosis. When progression of symptoms and facial nerve paralysis occur, the term malignant otitis externa might then be invoked. The similarity of the symptoms of nonresolving chronic otitis externa and malignant otitis externa and their progression with the clinical presentation of the slowly progressive squamous cell carcinoma of the external auditory canal should warrant further investigation at an earlier stage. Biopsy may be the only means of differentiating benign from cancerous lesions.

Failure to consider or disprove the diagnosis of squamous cell carcinoma can result in the progression of the carcinoma to an extensive, incurable stage. The differential diagnosis of nonresolving chronic otitis externa or malignant otitis externa, therefore, must include squamous cell carcinoma. Moreover, serious consideration should be given to the early biopsy of nonresolving chronic otitis externa, even in the absence of neural involvement, a late sign in carcinoma.⁷

The case presented here serves as a reminder that squamous cell carcinoma of the ear should be suspected in the presence of prolonged symptoms despite appropriate treatment with confirmed patient compliance. It would be difficult to speculate when cancer developed in this patient. Any chronic irritation is a setting for such an occurrence. As external otitis usually resolves within a few weeks of appropriate treatment, cancer could have occurred after the first 1 to 2 years. Biopsy would have been appropriate as early as 6 to 12 months after the initial symptoms failed to resolve.

Management of unresolving external otitis should include appropriate culture, investigation of concurrent illnesses, such as diabetes mellitus, and biopsy. Biopsy of skin lesions is risk-free provided appropriate asepsis is used. Highly suitable areas for biopsy include ulcerations, edges, and those areas surrounding erythematous tissue that are prone to bleeding.

When external otitis fails to resolve with the initial trial of antibiotics, samples of the inflamed tissue should be cultured; if after treatment with appropriate drugs, there is no resolution and the lesion continues to progress, biopsy is the most appropriate measure for definitive tissue diagnosis.

Finally, the diagnosis of malignant otitis externa should be questioned in nondiabetic pa-

tients and in cases in which organisms other than *Pseudomonas aeruginosa* are isolated, as these two characteristics are very highly correlated with malignant otitis externa. Only by being alert to this condition and initiating early investigation with biopsy can the primary care physician expect to diagnose squamous cell carcinoma of the external auditory canal at a curable stage.

References

1. Behrman RE, Vaughan VC, editors. Nelson textbook of pediatrics. 12th ed. Philadelphia: W.B. Saunders, 1983:1024-5.
2. Graham MD, Kemink JL. The wet ear. *Otolaryngol Clin North Am* 1986; 19:39-44.
3. Rubin J, Yu VL. Malignant external otitis: insights into pathogenesis, clinical manifestations, diagnosis, and therapy. *Am J Med* 1988; 85:391-8.
4. Rubin J, Stoehr G, Yu VL, Muder RR, Matador A, Kamerer DB. Efficacy of oral ciprofloxacin plus rifampin for treatment of malignant external otitis. *Arch Otolaryngol Head Neck Surg* 1989; 115: 1063-9.
5. Olsen KD, Desanto LW, Forbes GS. Radiographic assessment of squamous cell carcinoma of the temporal bone. *Laryngoscope* 1983; 93:1162-7.
6. Chakeres DW, Kapila A, LaMasters D. Soft-tissue abnormalities of the external auditory canal: subject review of CT findings. *Radiology* 1985; 156:105-9.
7. Mattucci KF, Setzen M, Golantich P. Necrotizing otitis externa occurring concurrently with epidermoid carcinoma. *Laryngoscope* 1986; 96:264-6.