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Active squamous chronic otitis media and labyrinthine fistula: The importance of vertigo symptoms

Two clinical cases demonstrate the need for physicians to maintain their vigilance for chronic otitis media and its complications, which can be life threatening, even in patients who do not have poor determinants of health, and to provide early intervention.

ABSTRACT: Chronic otitis media is a persistent middle ear inflammatory disease. One form of the disease, squamous chronic otitis media, features an accumulation of keratinizing squamous epithelium in the middle ear, known as a cholesteatoma, which can result in erosion of the ear's bony structures. Labyrinthine erosion leads to vertigo and permanent sensorineural hearing loss. In this study, two patients in their mid-80s presented with hearing loss and vertigo. In the first case, the patient was treated for osteomyelitis but later reported additional symptoms. An urgent tympanomastoidectomy improved his vertigo symptoms, but mastoid cavity related otorrhea requires repeated treatment with topical antibiotic ear drops and cleaning of the mastoid cavity. In the second case, the patient was first referred to a neurotologist when she developed symptoms of vertigo. An urgent tympanomastoidectomy relieved her vertigo, but her hearing was unchanged. Given that a chronic otitis media diagnosis is seldom considered by many clinicians in more economically developed countries, despite the disease's potential to cause life-changing complications or death, physicians are encouraged to consider it as a diagnosis in patients presenting with hearing loss, vertigo, and other features of ear disease.

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Otitis media, inflammation of the middle ear, comprises a spectrum of diseases, the most common of which affects children within their first 5 years of life.[1] Otitis media can be broadly categorized as acute or chronic, or otitis media with effusion.[2] Chronic otitis media is differentiated from the others by the presence of long-term tympanic membrane changes, such as a perforation or retraction pocket.[3] *Pseudomonas*, *Proteus*, and other anaerobes are the common bacteriological isolates in chronic otitis media. Risk factors for its development include low socioeconomic status, tobacco smoke exposure, malnutrition, and family history.[4] Chronic otitis media is significantly less prevalent in non-Indigenous populations in more economically developed countries such as Canada relative to less economically developed countries.[1] The prevalence varies from 2% to 4% in low-prevalence populations to 43% in high-prevalence populations.[2,5,6]

The common presenting features of chronic otitis media are hearing loss and otorrhea.[1] The World Health Organization Department of Child and Adolescent Health and Development and Team for Prevention of Blindness and Deafness recommends that patients with chronic otitis media who present with a newly discharging untreated ear or persistently discharging ear despite initial treatment can be managed in primary care with topical antibiotics for 2 to 4 weeks. They also recommend that the diagnosis be confirmed by a trained otoscopist.[6] Topical quinolones like ciprofloxacin are the preferred first-line antibiotics.[4] Antibiotic treatment should be initiated at the first consultation, providing that patients clean their ears prior to applying the medication. In cases of persistent or recurrent otorrhea, antibiotic resistance should be considered, and a bacterial culture should be obtained if available.[6]

Patients who present with a discharging ear accompanied by symptoms of headache, fever, and dizziness or other danger signs require urgent otolaryngology referral for consideration of mastoidectomy because of the possibility of intra- or extracranial disease extension.[6] Untreated chronic

otitis media can lead to several complications, including life-threatening intracranial abscesses and meningitis, as well as life-changing facial paralysis and labyrinthitis due to permanent sensorineural hearing loss.[7-9] The disease is subclassified into mucosal and cholesteatoma (squamous) associated subtypes. A cholesteatoma is an abnormal growth of keratinized squamous epithelium in the middle ear or mastoid.[10,11] The pathogenesis of cholesteatomas is uncertain; however, one explanation suggests that they form from retraction pockets of the pars flaccida that accumulate desquamated keratin.[12] Cholesteatomas erode bone by upregulating osteoclast-activation pathways, and by pressure necrosis, among other mechanisms, thereby leading to labyrinthine facial nerve bony canal and skull base erosions.[13-15] Progressive labyrinthine bone erosion can ultimately result in a labyrinthine fistula that presents classically with symptoms of hearing loss, vertigo, and disequilibrium. Nystagmus elicited by the fistula test, which stimulates vestibular receptors by applying intermittent positive pressure to the outer ear canal, is one sign of a labyrinthine fistula.[3] Facial nerve bony canal erosion by cholesteatoma can present with facial weakness, and some authors recommend limited surgical decompression of the canal in addition to cholesteatoma removal.[9] Severe otalgia or headache in a patient with otorrhea is particularly ominous because it is suggestive of skull base erosion with intracranial complications.[16]

Initially, squamous chronic otitis media can be asymptomatic or mildly symptomatic, resulting in missed early diagnoses. It is diagnosed by the typical appearance of a keratin collection in the pars flaccida on otoscopy and supported by CT temporal bone imaging. It can be easy for the clinician who is unfamiliar with the otoscopic appearance of an attic cholesteatoma to miss the diagnosis either through failing to visualize the entire pars flaccida of the tympanic membrane and adjacent deep ear canal bone or to underestimate the importance of crusting in this region. The cholesteatoma can be obscured by a superficial layer of what looks like simple ear wax **[Figure 1]**. The presence of opacification in the middle ear and mastoid with focal areas of bony erosion such as the scutum on CT scan is diagnostic of chronic otitis media with cholesteatoma.[6,11]

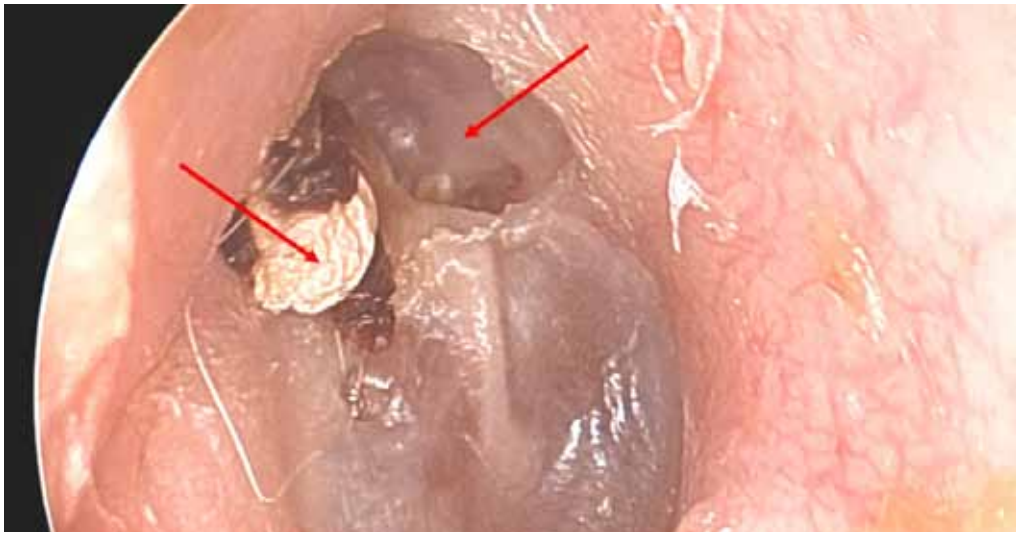

FIGURE 1. Otoscopic image demonstrating an extensive defect in the pars flaccida and pearly white keratin of a cholesteatoma obscured in part by what appears as simple ear wax.

Surgery is the mainstay of treatment of squamous chronic otitis media. Traditionally, mastoidectomy has been the procedure of choice and can be categorized into canal wall up (also known as intact canal wall) surgery, which preserves the posterior bony wall of the external ear canal, or canal wall down mastoidectomy surgery, which partially or completely sacrifices the posterior bony wall of the external ear canal.[10] Expanded endoscopic tympanoplasty is increasingly advocated for cholesteatomas with limited attic extension.[17,18] The surgical procedure selected depends on patient, disease, and surgical factors, including expertise in the range of available procedures.

We report two cases of a labyrinthine fistula complicating squamous chronic otitis media in patients who were not considered to be at risk of suffering from the disease. In both cases, the correct diagnosis was not made until dizziness or vertigo had developed secondary to semicircular canal erosion. We highlight the need for physicians to make a timely diagnosis of chronic otitis media and initiate treatment early to prevent the development of complications of the disease.

Case data

Case 1

An 82-year-old male presented to the emergency department with right-sided hearing loss and rhythmic oscillation of objects to the right in his visual field, accompanied by a "side-to-side rocking sensation." His medical history included a similar episode 2 years earlier of visual symptoms and right-sided hearing loss, though with right otorrhea, tinnitus, fever, lassitude, and decreased appetite. A course of antibiotics at that time resolved his symptoms except for the right-sided hearing loss and intermittent tinnitus. A head CT scan with contrast demonstrated opacification of the right mastoid air cells, and a diagnosis of osteomyelitis was made by an infectious diseases specialist. The patient was prescribed a 6-week course of clindamycin and ciprofloxacin and was referred to an otolaryngology clinic following its completion.

When seen in the otolaryngology clinic, he reported gradual resolution of the initial symptoms but new onset mild morning headaches, momentary dizziness on turning his head to the right, and unsteadiness when standing solely on his right leg. On otoscopy, the right ear demonstrated a cholesteatoma filling the attic, while the rest of the tympanic membrane appeared intact. Halmagyi-Curthoys head impulse and fistula tests were negative. Tuning fork testing at 512 Hz showed a positive bilateral Rinne response and Weber lateralization to the right. A temporal bone CT scan was ordered and demonstrated opacification of the right attic and mastoid air cell system, and erosion of the scutum, ossicular chain, lateral semicircular canal, and bone over the tympanic segment of

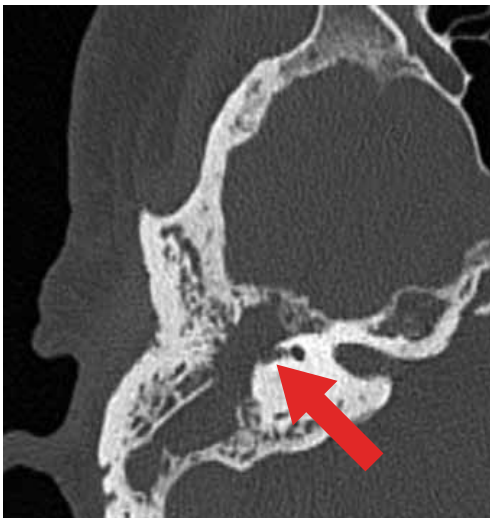

FIGURE 2. Head CT scan axial view showing opacification of the right mastoid air cells and erosion of the right lateral semicircular canal.

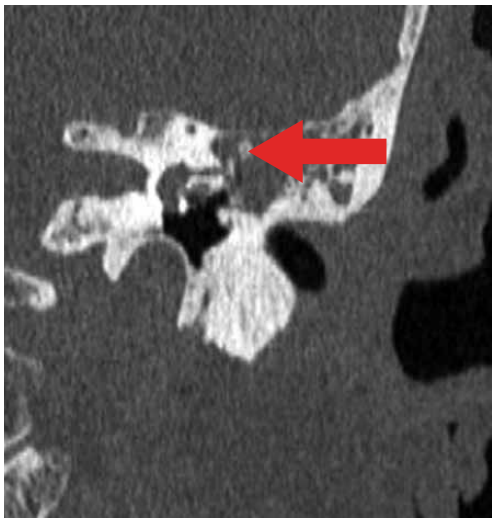

FIGURE 3. Temporal bone CT scan coronal view showing erosion of the left lateral semicircular canal. There is evidence of a previous cortical mastoidectomy, presumably performed in childhood, as the patient denies a previous operation.

the facial nerve canal, in keeping with chronic otitis media with cholesteatoma **[Figure 2]**. The patient was scheduled for an urgent tympanomastoidectomy. At surgery, a canal wall down mastoidectomy exposed a cholesteatoma. The head of the malleus and body of the incus bones were removed. Cholesteatoma was cleared from the mastoid air cells along with the cholesteatoma sac, including that covering the lateral semicircular bony canal dehiscence. The canal dehiscence was repaired with perichondrium. Harvested temporalis fascia, cartilage, and perichondrium were used to line the mastoid cavity and reconstruct the tympanic membrane. The patient was discharged the same day with no complications.

Seven months following the surgery, the patient reported slight improvement in his balance, no change in the hearing in his right ear, and only occasional right-sided tinnitus. He still experienced right otorrhea, which required topical antibiotic ear drops and repeat attendance for cleaning of the mastoid cavity.

Case 2

An 83-year-old female presented to a general otolaryngologist head and neck surgeon with a 5-year history of bilateral hearing loss and 1-month history of intermittent left-sided tinnitus. Additionally, she reported two episodes of vertigo in the previous month, described as a sudden tilt of the environment triggered by having the left ear syringed or by undergoing a hearing test. The patient had presented 2 years previously to the same otolaryngologist, who identified a moderate to severe left conductive hearing loss and mild to moderate right sensorineural hearing loss with a normal left pars flaccida. A left myringotomy and tympanostomy tube placement were undertaken at that time when polypoid tissue was found to be filling the middle ear space. On follow-up, the middle ear had returned to normal, though a residual tympanic membrane perforation persisted. A hearing aid was prescribed.

Otoscopy on the patient's current presentation demonstrated an attic retraction pocket of the left tympanic membrane with granulation tissue. A temporal bone CT scan was done, which identified soft tissue in the epitympanum, erosion of the left lateral semicircular canal, and possible erosion of the bone overlying the horizontal segment of the facial nerve canal and the mastoid bone adjacent to the middle and posterior fossa dura. The left mastoid air cells were opacified **[Figure 3]**. The patient was urgently referred to an otologist/neurotologist for consideration of surgery.

Otomicroscopy in the otology clinic demonstrated a left attic retraction pocket with granulation tissue, cholesteatoma, and discharge, along with a less than 5% perforation of the posterior inferior quadrant of the left tympanic membrane. The patient was scheduled for an urgent tympanomastoidectomy. Preoperative masked bone conduction pure tone audiometry demonstrated a mild to moderate downward sloping sensorineural hearing loss in the right ear. The hearing loss was mixed and moderate to severe in the left ear, with bone conduction thresholds identical to those in the right ear, but there was an added air-bone gap. Tympanometry recorded a Jerger type A tympanogram in the right ear and type B in the left ear, with ear canal volumes within the normal range bilaterally.

At surgery, an extensive cholesteatoma was removed via an intact canal wall tympanomastoidectomy. The skull base was eroded, which exposed dura in the tegmen of the middle ear and mastoid. The lateral and superior semicircular canals were partly eroded. Harvested temporalis fascia and bone chips were used to repair the canal dehiscences. The patient was observed as an inpatient for 24 hours, then was discharged home without complication.

Six months following the surgery, the patient had no otorrhea or vertigo. An audiogram confirmed that her hearing was unchanged. Her left tympanic membrane was intact. Follow-up imaging, including a temporal bone CT scan, at 1 year demonstrated a resolution of the middle ear and mastoid opacification **[Figure 4]**.

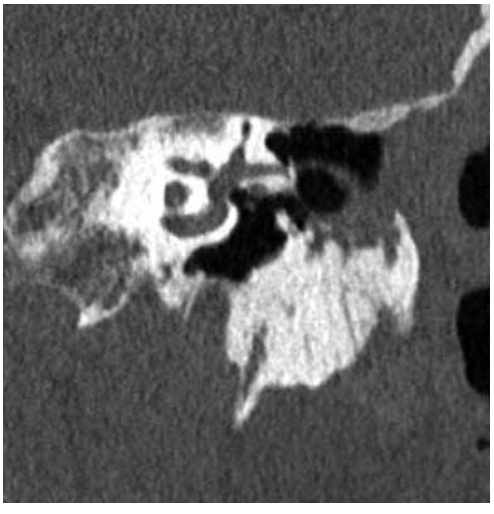

FIGURE 4. Postoperative temporal bone CT scan coronal view showing a normally aerated left middle ear and mastoid.

Discussion

In Case 1, the patient initially presented to the emergency department with symptoms suggestive of vestibular complications of chronic otitis media. In particular, the patient reported hearing loss and otorrhea, the primary diagnostic symptoms of chronic otitis media, and symptoms suggestive of nystagmus, which he reported as a perception of jerking motions when he looked at objects. The new onset of symptoms of nystagmus, coupled with a background of long-standing hearing loss and intermittent otorrhea, was indicative of a lateral semicircular canal fistula complication of squamous chronic otitis media. The patient was ultimately referred to an otolaryngology clinic, where microotoscopic examination confirmed a diagnosis of chronic squamous otitis media, and high-resolution temporal bone CT scan images better delineated the lateral semicircular canal fistula that was evident on the initial head CT scan. The lateral semicircular canal fistula was previously interpreted to be due to skull base osteomyelitis. This case highlights the importance of thorough otoscopic evaluation, including microotoscopy, in the workup of patients with features of ear discharge and hearing loss. Early identification of the characteristic otoscopic findings of squamous chronic otitis media would have expedited appropriate treatment.

In Case 2, the new onset of vertigo triggered by pressure change in the outer ear was highly suggestive of a labyrinthine fistula, as confirmed by the temporal bone CT scan. Canal wall down mastoidectomy with cholesteatoma removal but preservation of the cholesteatoma matrix over the area of eroded lateral semicircular canal bone is the traditional surgical management of a middle ear and mastoid cholesteatoma complicated by a lateral canal fistula.[19] Advancements in MRI techniques and the established safety of controlled removal of the cholesteatoma matrix with preservation of inner ear function makes it unnecessary to undertake canal wall down mastoidectomy routinely in lateral canal fistula cases.[20,21] Follow-up imaging at 1 year in Case 2 demonstrated the safety of an intact canal wall mastoidectomy approach.

The literature shows that chronic otitis media is more prevalent in less economically developed countries and among more marginalized segments of the population in more economically developed countries. These two cases illustrate the need for physicians to watch for this disease and its complications, even in patients who do not have poor determinants of health. The complications of chronic otitis media are still life threatening or life changing, and require early intervention.

Maintaining a high degree of vigilance will help ensure that patients with complications are identified early, and the required surgical interventions are expedited to achieve satisfactory patient outcomes.

Summary

Chronic otitis media can occur in economically developed countries in patients who have no significant comorbidities or poor determinants of health. Given the disease's ability to cause severe complications, including death, competent otoscopic evaluation, including microotoscopy, should be undertaken or arranged urgently in patients with the characteristic symptoms of otorrhea and hearing loss who present with features of complications such as nystagmus and vertigo. Maintaining a high degree of vigilance will help ensure that patients with complications are identified early, and the required surgical interventions are expedited to achieve satisfactory patient outcomes. ■

Competing interests

Dr Nunez receives remuneration for providing independent medical expert neurotological opinions. Mr Abdelmalek has declared no competing interests.

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