Symptom: Plugging and Drainage

By Hamid R. Djalilian, MD

A 72-year-old male presents with a history of chronic ear plugging and intermittent drainage. The drainage episodes generally respond to antibiotic ear drops.

He has seen his primary care physician, who has attempted to flush his ear or pick out the debris, occasionally resulting in bleeding or infections.

The patient states that he has not had previous surgery or trauma to his head or ear. His past medical history is significant for high blood pressure and high cholesterol, for which he takes medication.

Examination of the ear shows an accumulation of dead skin in the ear canal, with destruction of the ear canal floor.

What is your diagnosis? See p. 6.

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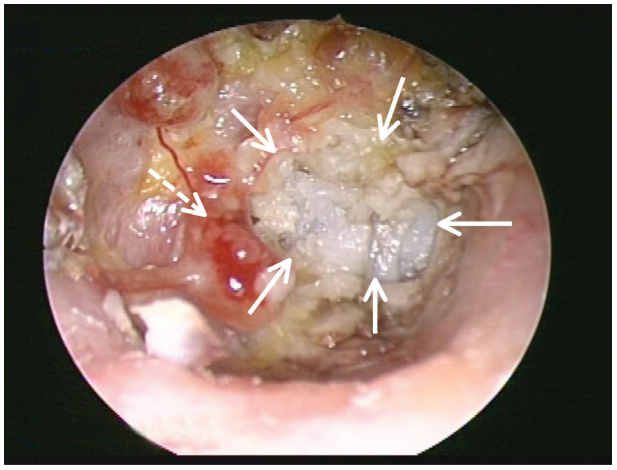

In this view of the patient's ear on otoscopy, solid arrows indicate the tympanic membrane area, and the dashed arrow indicates granulation tissue in the canal, seen posteriorly.

Diagnosis: Canal Cholesteatoma

By Hamid R. Djalilian, MD

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The finding of accumulated dead skin in the ear canal along with destruction of ear canal bone is unusual. The differential diagnosis included external auditory canal cholesteatoma, keratosis obturans, malignant otitis externa (skull base osteomyelitis), and cancerous tumors of the ear canal:

• **Keratosis obturans** generally does not cause significant bony destruction or contain floating dead bone pieces (sequestrum).

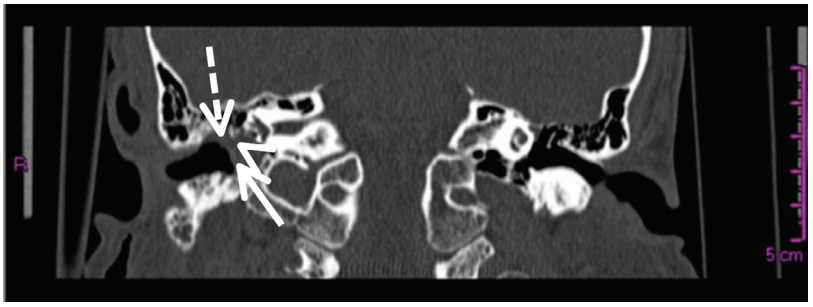
• **Skull base osteomyelitis** is generally accompanied by significant pain and occurs in the setting of immune suppression.

• **Malignant tumors of the external auditory canal skin** are most commonly associated with a friable soft tissue mass and are easily ruled out by a biopsy.

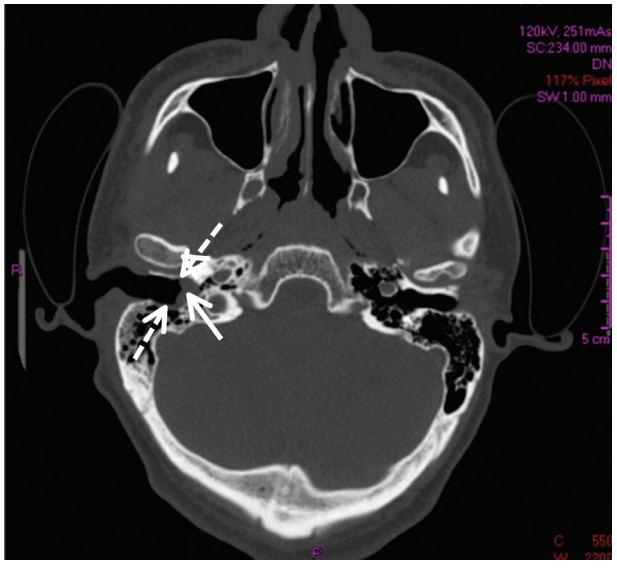
• This patient had an **external canal cholesteatoma**.

Diagnosis of external auditory canal cholesteatoma requires thorough cleaning of the ear canal debris and visualization of the destroyed bone. The presence of devitalized bone (sequestrum) intermixed with the keratin debris is highly suggestive of the diagnosis.

One other disease entity that can cause the same picture is osteoradionecrosis. This condition is caused by a loss of bone tissue vascularity. In general, a biopsy may be needed to rule out cancer.



This coronal CT image of the temporal bone shows the canal cholesteatoma (arrow) and the ear canal destruction (dashed arrow). In this patient, the destruction of bone has involved the mastoid medially and caused middle ear inflammation as well (arrowhead). The cholesteatoma did not involve the middle ear.



This axial CT image of the temporal bone demonstrates the canal cholesteatoma, indicated by the solid arrow, causing destruction of the ear canal, indicated by the dashed arrows.

Cholesteatomas are most commonly found in the middle ear and mastoid, and are a result of chronic Eustachian tube dysfunction. External canal cholesteatoma is rare, with an estimated incidence of 0.1 percent to 0.5 percent of all otologic patients.

> The finding of accumulated dead skin in the ear canal along with destruction of ear canal bone is unusual.

UNCERTAIN CAUSE

The cause of external canal cholesteatoma is unknown. Generally, spontaneous canal cholesteatomas are seen in elderly patients, though they can occur in younger patients.

While most cases of canal cholesteatoma occur spontaneously, they can result from trauma or surgery. During surgery, a piece of skin can get implanted or turn under such that it starts growing inward and destroys the canal bone. Sometimes, external canal cholesteatoma can be caused by stenosis of the external auditory canal, with trapped epithelium (skin) behind the stenosed or closed canal.

Typically, the skin of the ear canal and the outer layer of the tympanic membrane produce new layers, shedding the outer dead layer of skin.

This dead skin layer (keratin layer) normally moves outward during a natural migration that starts at the tympanic membrane and moves to the outside canal. This movement is responsible for the extrusion of normal cerumen from the ear.

It has been hypothesized that the process slows in the setting of canal cholesteatoma.

However, this theory does not explain the destruction of canal bone that occurs in this setting, and why young people may also be affected.

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Another theory for the pathogenesis of canal cholesteatoma states that local accumulation of keratin debris in the setting of slowed migration can alter the underlying epithelium and cause bony destruction. The slower epithelial migration of the lower part of the external auditory canal may explain why a majority of cases occur in the inferior canal.

TREATMENT VARIES

Patients with external canal cholesteatomas generally present with ear drainage. Sometimes, they also have a dull pain. They may have infections from water entering the ear and getting trapped within the destroyed portion of the ear canal. *Pseudomonas aeruginosa* tends to grow in moist environments, such as wet keratin debris accumulating within the canal cholesteatoma.

In rare cases with great expansion, external canal cholesteatomas can involve the facial nerve in the mastoid and cause facial paralysis.

Imaging is sometimes used to distinguish external canal cholesteatoma from other entities, such as keratosis obturans. As mentioned previously, the presence of floating bone in the midst of the keratin debris in the canal is highly suggestive of canal cholesteatoma. The absence of radiation therapy history and a negative biopsy help establish the diagnosis.

Treatment of canal cholesteatomas depends on the extent of the lesion and the areas involved with the disease. Small lesions with limited involvement of the mastoid can be managed with alcohol/vinegar irrigations and in-office cleaning to clear the debris. This treatment is the best option for elderly patients and those with significant medical problems.

When the lesion is causing complications or extends beyond the view of the examiner, surgical therapy is indicated. The goal of surgery is to remove all the squamous epithelium (skin) lining the cholesteatoma and to repair the mastoid defect to prevent re-formation of the disease.