Spontaneous posterior wall external canal cholesteatoma

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A 44-year-old woman with a history negative for trauma, inflammation, surgery, or radiotherapy of the head or neck was referred for cerumen removal. Following the cerumen removal, a dehiscence of the posterior wall of the bony canal with squamous debris suggestive of external canal cholesteatoma was observed (figure).

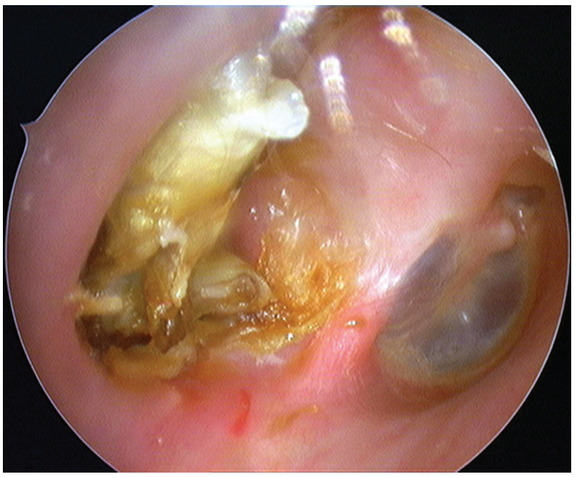

Figure. Photograph shows a cholesteatoma in the posterior wall of the right external auditory canal. The squamous debris and bony canal defect are evident in the front, and a normal tympanic membrane is seen in the back.

This is a rare condition usually involving the inferior auditory canal.[1] Its incidence is 0.30 cases per 100,000 persons per year and 1.2 to 7.1 cases per 1,000 new otologic patients per year.[2,3] This disease is distinguished from keratosis obturans because cholesteatoma develops after a primary canal bone disorder.[4] Ear discharge and otalgia of varying severity (from no pain to severe pain) characterize the clinical picture; hearing loss is rarely observed, primarily because keratin debris obstructs the external canal.[3]

Four progressively worse clinical and histologic stages have been described: simple epithelial hyperplasia, periosteitis, erosion of the bony canal, and erosion of adjacent structures.[5] The first stage is characterized by focal hyperplasia of the external canal epithelium with accumulation of inflammatory cells in adjacent stromal tissues. During subsequent stages, the lesion invades deeper into mesenchymal tissues, leading to accumulation of necrobiotic keratin debris in the newly formed central cavity; bone erosion and bone sequestra may also occur.[1]

There are two basic types of external auditory canal cholesteatoma: *secondary* and *spontaneous*. Secondary cases may arise following congenital, post-traumatic, iatrogenic, or postinflammatory occlusion or narrowing of the external meatus, which results in retention of squamous debris in the medial portion of the external canal. Secondary cholesteatomas may also occur after iatrogenic or traumatic entrapment of squamous epithelium, which leads to cyst formation deep to the skin of the external canal. Finally, secondary lesions may arise following trauma, inflammation, radiation, or iatrogenic means that erode the external canal bone and allow squamous epithelial growth in the defect.[6]

The pathogenesis of spontaneous ear canal cholesteatoma is still debated. Several hypotheses have been proposed, including (1) microtrauma to the external canal skin; (2) retention of hard or adulterated cerumen; (3) focal osteitis; (4) hypoxia leading to angiogenesis via hepatocyte growth factor/scatter factor, tyrosine-kinase c-Met receptor, and vascular endothelial growth factor; and (5) decreased epithelial migration secondary to aging or poor blood supply.[5,7-9] A recent

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for surgery. It was difficult to completely remove the tumor from the facial nerve via the translabyrinth approach because the tumor closely adhered to the facial nerve. The histopathologic examination showed meningeal whorls and nuclear pseudoinclusions, and the mass was confirmed to be a meningioma.

Vestibular schwannoma is the most common tumor in the IAC. The differential diagnosis of an IAC mass includes arachnoid cysts, hemangiomas, cholesteatomas, and lipomas.[1] Meningiomas can be confined to the IAC because the arachnoid villi can be distributed within the IAC and along the superficial petrosal nerve.[2] Vestibular schwannomas are isohyperintense, but meningiomas have variable signal intensity on T2-weighted MR images.

Meningiomas more commonly have calcifications and the dural tail sign, and they tend to have more hyperostosis at the petrous bone or in the IAC and a ragged tumor border. Vestibular schwannomas have a more spherical shape.[3] Meningiomas adhere tightly to the facial nerve, rendering a meticulous, sharp dissection difficult, even with small intracanalicular meningiomas.[4] Although meningioma in the IAC is very rare, it should be included in the differential diagnosis of tumors in the IAC.

References

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report showed that epithelial migration is not substantially changed in patients with canal cholesteatoma, however.[10]

The differential diagnosis of ear canal cholesteatoma includes malignant tumor, aseptic necrosis, and malignant otitis externa.[1] Computed tomography of the temporal bone is recommended to assess middle ear or mastoid lesions for erosion of the labyrinth, facial canal, and tegmina.[9]

Ear canal cholesteatomas may require surgical intervention if any of the following are present: (1) extensive middle ear or mastoid invasion, (2) present or potential complications, (3) otorrhea uncontrolled by local medical treatment, or (4) significant hearing loss.[7] Surgical intervention may include canaloplasty, intact canal wall mastoidectomy with canal wall reconstruction, or canal-wall-down mastoidectomy, depending on the lesion site and extent of bone destruction.[7]

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