# **External Auditory Canal Cholesteatoma Mimicking Malignancy: a case report**

Hui-Chung Yang<sup>1</sup> Chia-Yuen Chen<sup>1,2</sup> Wing P. Chan<sup>1,2</sup> Chi-Long Chen<sup>3</sup> Chun Yu<sup>1,2</sup>

School of Medicine<sup>1</sup>, Taipei Medical University
Department of Radiology<sup>2</sup>, Taipei Medical University-Municipal Wan Fang Hospital
Department of Pathology<sup>3</sup>, Taipei Medical University-Municipal Wan Fang Hospital

External auditory canal cholesteatoma is a rare disease entity. Here we present a case of external auditory canal cholesteatoma with adjacent bone destruction mimicking malignancy on high-resolution computed tomography. The final diagnoses is external auditory canal cholesteatoma.

*Key words:* Cholesteatoma; Ear, CT; External auditory canal

External auditory canal cholesteatoma is a rare disease entity [1]. The cause of this disease is not well understood. Desquamating epithelium normally migrates laterally in the external auditory canal and is exteriorized, but this process fails in external auditory canal cholesteatoma. The trapped keratinous debris accumulates in the lumen of external auditory canal and causes local bone erosion and destruction. Clinical differential diagnosis may include a range of operable and inoperable conditions, including neoplasms and inflammatory and infectious conditions of the external auditory canal [2].

We report a case of an external auditory canal cholesteatoma causing adjacent bone erosion and destruction and involving the mastoid and temporomandibular joint.

#### **CASE REPORT**

A 36-year-old woman suffered from progressive dull pain with hearing impairment in the left ear for 2 months. Physical examination revealed a large mass occupying her left external auditory canal.

High resolution computed tomography(CT) scan of the temporal bone showed a lobulated soft tissue mass occupying the entire left external auditory canal with destruction of the adjacent bony structures (Fig. 1a, 1b). Increased density in the left middle ear cavity and fluid accumulation in the left mastoid air antrum were noted. There was no ossicular chain destruction or erosion to suggest middle ear cholesteatoma. Initial diagnosis was an external auditory canal tumor with aggressive behavior coexisting with chronic otitis media. The patient then underwent excisional biopsy of the lesion and modified mastoidectomy.

Surgical findings revealed a mass in the left external auditory canal with invasion of adjacent bony portion. A large amount of pus and granulation tissue coating on the tumor surface was noted. Grossly, the tumor was a brownish cystic lesion with muddy kerati-

Reprint requests to: Dr. Chia-Yuen Chen Department of Radiology, Taipei Medical University-Municipal Wan Fang Hospital. No. 111, Sec. 3, Hsing Long Road, Taipei 116, Taiwan,

R.O.C.

nous material, measuring  $1.2 \times 1.0 \times 0.8$  cm. Histological examination revealed cholesteatoma composed of keratinized squamous epithelium and peripheral granulation tissue (Fig. 2a, 2b). No bone sequestrum was seen.

### DISCUSSION

Acquired cholesteatoma is an inflammatory mass of the petrous part of temporal bone. It is most commonly encountered in the middle ear cavity. External auditory canal cholesteatoma is a rare disease entity, accounting for 0.1-0.5% of new otolaryngology patients [1]. Patients with external auditory canal cholesteatoma typically present with otorrhea and a chronic dull pain due to the local invasion of squamous tissue into the osseous structures [3]. Clinical examination and otoscopy are commonly used to diagnose cholesteatoma. High resolution CT(HRCT) scan can demonstrate the extent of cholesteatoma and bone erosion with 80% specificity [4, 5]. However, the cholesteatoma sac, associated granulation tissue, mucosal edema and effusion can be indistinguishable on CT images [6, 7].

A soft-tissue mass in the external auditory canal with adjacent bone erosion on HRCT is typical findings of external auditory canal cholesteatoma. With use of high resolution CT scan, Heilbrun et al. [2] reported that 7 of their 13 patients with external auditory canal cholesteatoma showed a soft-tissue mass with adjacent bone erosion and intramural bone fragments. In their study, eight cases arose inferiorly, eight cases arose posteriorly, and two cases were circumferential. Bone fragments are often present within the mass. The external auditory canal cholesteatoma may extend deeply into the middle ear, mastoid, facial

nerve canal, or the tegmen tympani.

The bone erosion adjacent to the soft-tissue mass may be smooth, similar to that of a middle ear cholesteatoma. However, the erosion may be irregular, secondary to the necrotic bone and periostitis [8]. External auditory canal cholesteatoma often involves multiple sites of the external auditory canal, with the posterior and inferior segments most commonly affected. Circumferential involvement is also common [2, 3, 9]. As in our case, the lesion showed circumferential involvement occupying the entire external auditory canal and extension to the middle ear, sparing the ossicular chain. The external auditory canal was focally eroded and the left temporomandibular joint was involved. However, no intramural bone fragment was identified, as documented in the histologic examination.

Gross pathologic finding of external auditory canal cholesteatoma usually demonstrates extensive erosion of the bony external auditory canal by a widemouthed stratified squamous keratinizing epithelial sac with localized periostitis and sequestration of bone [10]. The tympanic membrane is typically normal [10]. This erosion is thought to be related to proteolytic enzymes along the margin of the lesion produced within the cyst lining; this weakens the bone and results in periostitis and sequestration of bone. The erosion can also be partly related to the accumulation of keratin debris, which traps moisture and results in a bacterial infection that can cause ulceration of the epithelial layer and granulation tissue formation in patients who have a superimposed infection [10]. As in our case, granulation tissue with necrotic debris coating and dense inflammatory cell infiltration were seen at initial biopsy.

On otoscopic examination, it can be difficult to distinguish external auditory canal cholesteatoma from

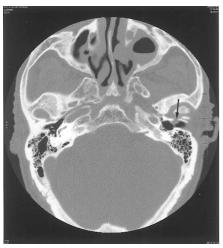
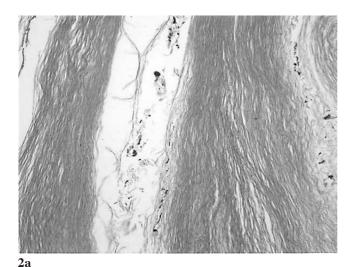




Figure 1. Axial a. and coronal b. high resolution CT scans of temporal bone show a soft tissue mass occupying the left external auditory canal with bone destruction at the inferior (white arrow) and anterior walls (black arrow). The left temporomandibular joint is involved. The middle ear cavity and ossicular chains are intact. The scutum (curved arrow) and eardrum are preserved.

1a 1b



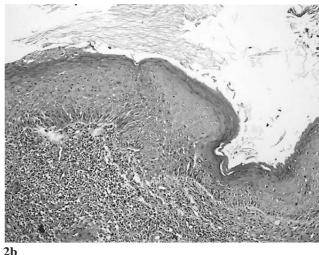


Figure 2. Photographs of the lesion specimen. Large keratin flask a. and chronic inflammation with granulation tissue formation b. are noted in the external canal tissue. Keratinization is noted in overlying squamous epithelium (b).

other inflammatory, infectious, or neoplastic processes. Examples of these include keratosis obturans, malignant otitis externa, and squamous cell carcinoma. The most closely related non-neoplastic condition may be keratosis obturans [2, 3]. Keratosis obturans generally occurs in a youger age group. It is often bilateral, and it has a definite relationship to bronchiectasis and sinusitis [3]. CT imaging of keratosis obturans typically demonstrates a soft-tissue plug in bilateral external auditory canals, without focal bone erosion, which may be different from external auditory canal cholesteatoma [2]. Malignant otitis externa is a severe infectious disease involving the external auditory canal. The disease classically occurs in elderly patients with diabetes mellitus. The diagnosis is generally based on clinical examination, abnormal laboratory and imaging studies. CT imaging shows cortical bone erosion with abnormal soft tissue, which may be indistinguishable from malignancy [2, 8]. Malignant tumors originating from the external auditory canal are rare, with squamous cell carcinoma being the most commonly encountered. It may be difficult to fifferentiate squamous cell carcinoma in the external auditory canal from external auditory canal cholesteatoma by imaging findings alone [2, 8].

In conclusion, cholesteatoma in the external auditory canal presenting as a soft-tissue mass with bone destruction may mimic malignancy. Tissue biopsy is required to obtain a correct diagnosis. High resolution CT scan is helpful in evaluation of deep extension of the tumor into the mastoid air cells and other structures, and thus is also helpful in surgical planning.

#### REFERENCES

- Anthony PF, Anthony WP. Surgical treatment of external auditory canal cholesteatoma. Laryngoscope 1982; 92: 70-75
- 2. Heilbrun ME, Salzman KL, Glastonbury CM, Harnsberger HR, Kennedy RJ, Shelton C. External auditory canal cholesteatoma: clinical and imaging spectrum. AJNR Am J Neuroradiol 2003; 24: 751-756
- 3. Piepergerdes MC, Kramer BM, Behnke EE. Keratosis obturans and external auditory canal cholesteatoma. Laryngoscope 1980; 90: 383-391
- 4. O'Donoghue GM, Bates GJ, Anslow P, Rothera MP. The predictive value of high resolution computed tomography in chronic suppurative ear disease. Clin Otol 1987; 12: 89-96
- O'Reilly BJ, Chevretton EB, Wylie I, et al. The value of CT scanning in suppurative otitis media. J Laryngol Otol 1991; 105: 990-994
- Jackler RK, Dillon WP, Schindler RA. Computed tomography in suppurative ear disease: a correlation of surgical and radiographic findings. Laryngoscope 1984; 94: 746-752
- 7. Phelps PD, Wright A. Imaging cholesteatoma. Clin Radiol 1990; 41: 156-162
- 8. Swartz JD, Harnsberger HR. The external auditory canal. In: Imaging of the temporal bone. 3rd Ed. New York: Thieme 1998: 16-46
- Garin P, Degols JC, Delos M. External auditory canal cholesteatoma. Arch Otolaryngol Head Neck Surg 1997; 123: 62-65
- 10. Applebaum EL, Duff BE. Ear and temporal bone, I: clinical considerations for non-neoplastic lesions of the ear and temporal bone. In: Fu YS, Wenig BM, Abemayor E, Wenig BL, eds. Head and neck pathology with clinical correlations. Philadelphia: Churchill Livingstone 2001: 668-678

## 外耳道珍珠瘤似爲惡性瘤:病例報告

楊惠中1 陳家媛1,2 陳榮邦1,2 陳志榮3 于 俊1,2

臺北醫學大學 醫學系<sup>1</sup> 臺北醫學大學市立萬芳醫院 放射線科<sup>2</sup> 病理科<sup>3</sup>

外耳道珍珠瘤為耳鼻喉臨床少見病例。其成因尚未充分了解。我們報告一位36歲女性罹患 外耳道珍珠瘤,高解像電腦斷層表現為外耳道腫瘤,以及鄰近的骨破壞,最初診斷懷疑外耳道 惡性腫瘤。

關鍵詞:珍珠瘤;耳,電腦斷層;外耳道