

Ceruminous gland carcinoma of the external auditory canal presenting as chronic otitis media

A. Selcuk*, S. Ensari*, M. A. Cetin*, S. Dizbay Sak** and H. Dere*

*Ankara Numune Educational and Research Hospital, 4th ENT Clinic and **Ankara University Faculty of Medicine, Department of Pathology, Ankara, Turkey

Key-words. External auditory canal; ceruminous; carcinoma

Abstract. *Ceruminous gland carcinoma of the external auditory canal presenting as chronic otitis media. Objective:* Ceruminous neoplasms of the external ear canal are rare. Classification, clinical behavior, and management of these tumours are controversial. We report a case of carcinoma originating from the ceruminous glands of the external ear canal (EAC), operated based on a diagnosis of chronic otitis media with polyp.

Case report: A 48-year-old man presented with left ear discharge and hearing loss. Clinical examination showed a well-circumscribed polypoid mass limited to the EAC. There was no history of bloody ear discharge or radiological findings of bony erosion suggestive of malignancy. Our preliminary diagnosis was chronic otitis media with polyp formation. Tympanoplasty was performed. Histopathology revealed a ceruminous carcinoma, and an additional operation involving lateral temporal bone dissection was performed, followed by 60 Gy radiation therapy.

Conclusion: Ceruminous carcinomas should be considered in the differential diagnosis of middle and external ear pathologies in cases of soft tissue mass in the EAC.

Introduction

Ceruminous neoplasms arising in the external auditory canal (EAC) are extremely rare. The true incidence, nomenclature, behavior, and histopathological classification remain unclear, making management of these tumours the subject of controversy.¹

Here we describe a case of ceruminous gland carcinoma of the EAC that was operated based on a diagnosis of chronic otitis media with polyposis. Ceruminous neoplasms should be considered in evaluation of cases with soft tissue mass in the EAC or chronic otitis media with polyposis.

Case report

A 48-year-old man presented to our clinic with complaints of

intermittent left ear discharge and a gradual hearing loss since childhood. He also reported experiencing otalgia and a feeling of blockage in the ear canal for 6 months. Clinical examination revealed a reddish polypoid mass and mucopurulent discharge in the left ear canal. There was no facial weakness, enlarged lymph node, or parotid mass. Pure tone audiometry revealed an air-bone conduction gap in the left ear. A fine-section temporal bone CT scan showed a mass with soft tissue density in the external ear canal enclosing the eardrum and the ossicles. Demineralization and destruction of the ossicles also were observed, and mastoid cells were sclerotic (Figure 1). With these clinical and radiological findings, our preliminary diagnosis was chronic otitis media com-



Figure 1

Temporal bone CT scan of the patient shows a mass with soft tissue density in the external ear canal enclosing the eardrum and the ossicles. Demineralization and destruction of ossicles are noted. Mastoid cells are sclerotic.

plicated with polyp formation, and we performed an intact-canal wall down tympanoplasty.

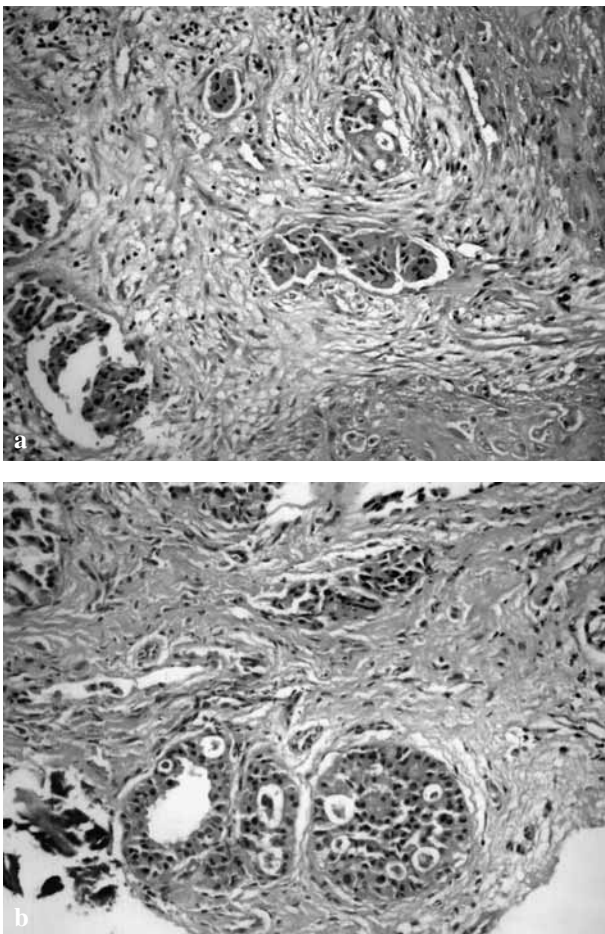


Figure 2

(a) Small nests of cells within the stroma (HE); (b) cribriform area (HE).

Histopathological evaluation of the specimen was reported as carcinoma of the ceruminous glands with adenoid cystic carcinoma-like differentiation. The patient was evaluated for local or distant metastasis. Neck CT showed no lymphadenopathy, and distant metastasis was absent. The patient was re-operated, and a lateral temporal bone dissection was performed. Postoperative radiation therapy was delivered to the tumour area (60 Gy, fractionized, 6 weeks). At 6 months postoperatively, the patient was evaluated with temporal, cranial, and thorax CTs and showed no evidence of

local recurrence or distant metastasis.

Pathological findings

Histopathological examination of the lesion demonstrated an infiltrative neoplasm composed of small nests and islands of epithelial cells within the stroma with focal areas of cribriform organization resembling adenoid cystic carcinoma (Figure 2). Invaginations from the squamous epithelium from the surface and non-neoplastic ceruminous glands were observed in between the tumour islands and nests. Nuclear

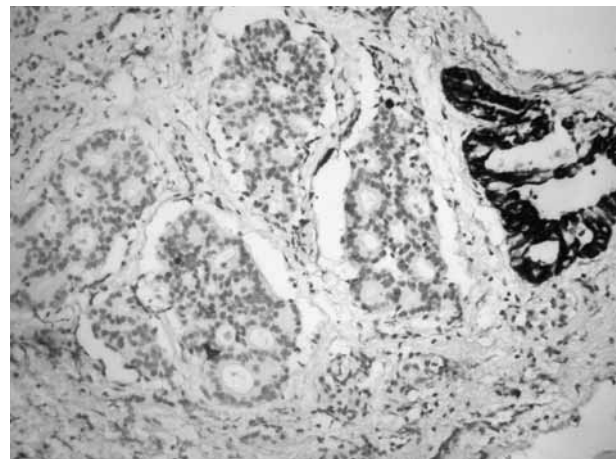


Figure 3

Cribriform organization is more prominent in the deeper levels of the block, which were used for immunohistochemical stains. Tumour cells were not stained with CK7, in contrast with the neighboring CK7-positive glands.

pleomorphism was low to moderate, and mitotic activity was low. Apocrine differentiation was not observed, and no lymphovascular or perineural invasion was identified.

Immunohistochemical staining for p63 (Neomarkers, MS1081-P, 1:200), S-100 (Neomarkers, MS296-P1, 1:200), cytokeratin (CK) 7 (Neomarkers, MS1352-P, 1:150), CK5/6 (Dako, M7237, 1:50), and Ki-67 (Neomarkers, RM9106-S, 1:200) was performed using the streptavidin-biotin peroxidase technique with appropriate positive and negative controls. CK7 was positive in non-neoplastic glands but negative in the tumour cells (Figure 3). CK5/6 was positive in the squamous epithelium on the surface and in the myoepithelial layer of the glands but negative in the tumour cells. With p63, staining was observed in the squamous epithelium, in the myoepithelial layer of the non-neoplastic glands, and in the peripheral cells of the cribriform tumour groups. With Ki67, 5-10% of the tumour nuclei were

labeled. The tumour was diagnosed as carcinoma of the ceruminous glands.

Discussion

The skin of the cartilaginous portion of the EAC contains two types of glandular structures: ceruminous and sebaceous glands. Ceruminous glands lie deep within the dermis, consisting of columnar cells surrounded by a layer of myoepithelial cells containing intensely eosinophilic cytoplasm. They are responsible for the watery secretion of the ear canal. This fluid mixes with the secretion of the sebaceous glands to compose cerumen.^{2,3}

Ceruminous neoplasms of the EAC were reported as "ceruminoma" for the first time by Haugh⁴ in 1894. The term ceruminoma has long been used for both benign and malignant ceruminous gland neoplasms, which has caused confusion. In 1972, Wetli *et al.*⁵ proposed a new classification based on pathological findings, with the classifications of pleomorphic adenoma, ceruminous adenoma, ceruminous adenocarcinoma, and ceruminous adenoid cystic carcinoma. Additional variants have been proposed as part of this classification, including syringocystadenoma papilliferum, mucoepidermoid carcinoma, and benign-malignant eccrine cylindroma.³⁻⁶ The term "ceruminoma" was excluded from the World Health Organization (WHO) classification in 1991. In the current WHO classification, malignant tumours of ceruminous glands are covered in three categories: adenocarcinoma (low and high grades), adenoid cystic carcinoma, and mucoepidermoid carcinoma.⁷ Because of the rarity of neoplasms of the

ceruminous glands, these three categories may not be adequate for classifying all histological types that arise in this area. The present case, consisting of scattered tumour nests and focal cribriform areas, was somewhat reminiscent of adenoid cystic carcinoma; however, it was not completely consistent with any of the current WHO categories and was descriptively diagnosed as a carcinoma of the ceruminous glands with adenoid cystic carcinoma-like differentiation.

The etiopathogenesis of ceruminous carcinoma has not been fully clarified, but presumptive progenitors are embryonic anlage, ectopic remnants of salivary glands, and the ceruminous gland itself.²

Patients usually present with otalgia and a mass in the external ear canal. A sense of aural fullness, bloody ear discharge, and conductive-type hearing loss are frequent accompanying symptoms. External otitis media may also be present. Tumour extension may progress laterally to the auricle along the skin, medially to the tympanic membrane, middle ear, and mastoid air cells, or to the parotid through holes in the external canal cartilage. The facial nerve may be involved. A CT scan is helpful in detecting bone erosion and the extent of the mass.^{1-6,8-9}

Our patient had no history of bloody ear discharge, facial weakness, or radiological findings of bone erosion that could suggest malignancy. There was a history of ear discharge and gradual hearing loss since childhood. CT scanning of the temporal bone in this patient demonstrated soft tissue in the external ear canal, mastoid, and middle ear cleft, indicating

otitis media. The history of former otologic disease further complicated the diagnosis. All of these findings directed us towards a conclusion of chronic otitis media rather than an EAC neoplasm.

No universally accepted staging system for ceruminous EAC malignancies exists. Several factors impede the process of developing a staging system, including the rarity of the tumours, the impossibility of evaluating disease extent by physical examination alone, and the unreliability of radiographic studies to determine the extent of disease. A staging system for malignancies of the EAC proposed by Arriaga *et al.*¹⁰ and modified by Moffat *et al.*¹¹ has been useful and has gained support. In general, tumours that are limited to the EAC are defined as early-stage disease, and those that extend beyond the external canal and invade the surrounding soft tissues, the middle ear, the mastoid, or cranial nerves are recognized as late-stage disease.

Local excision is the treatment of benign ceruminous neoplasms. More radical surgery and radiotherapy should be applied for malignant tumours. Treatment is based on the feasibility of resection, and the extent of resection depends on the extent of tumour involvement. The limits of resection may include lateral, subtotal, or total temporal bone resection. The lateral temporal bone dissection includes resection of the EAC, tympanic membrane, malleus, and incus. The boundaries are the middle ear cavity and stapes medially, the mastoid cavity posteriorly, the epitympanum and zygomatic root superiorly, the temporomandibular joint capsule anteriorly, and the medial tympanic

ring or infratemporal fossa inferiorly. Lateral temporal bone dissection is appropriate for T1 and T2 tumours. A subtotal temporal bone resection is performed when invasion medial to the tympanic membrane or into the mastoid (T3 disease) is evident. In this case, the medial margin may be obtained in a piecemeal fashion, usually with a drill. The specimen includes the lateral temporal bone dissection with additional dissection of the otic capsule and the medial bony wall of the middle ear and mastoid. The margins of resection are the sigmoid sinus and posterior fossa dura posteriorly, middle fossa dura superiorly, internal carotid artery anteriorly, jugular bulb inferiorly, and petrous apex medially. A total temporal bone dissection can be used to address T4 disease. This approach includes a subtotal temporal bone dissection with the additional resection of the petrous apex. The internal carotid artery may be isolated, mobilized, and preserved or resected. The sigmoid sinus, jugular vein, carotid artery, dura, and cranial nerves are removed as indicated by the extent of the tumour.¹²

Adjuvant radiation therapy may also be required, depending on the extent of the lesion. Recurrence may be seen within months or years. Distant metastases of malignant types have been reported,^{9-11,13-16} and their presence heralds a poor prognosis in these types of cancer. Death usually is caused by intracranial extension of the tumour or by pulmonary metastases. Therefore, all patients who have a histologically confirmed adenoid cystic carcinoma of the external ear canal should have lifelong follow-up. Cranial and thorax CT is recommended at

6-month intervals.^{9-11,13-16} Distant metastases should be excised when possible.

We performed a tympanoplasty, approaching the case as otitis media complicated with polyp formation. Mastoid air cells were infected and partially sclerotic, and the ossicles were defective. Intraoperative findings resembled findings of chronic otitis media except for excess bleeding. In several studies, it has been reported that chronic otitis media and cholesteatoma are common in patients with temporal bone cancers and have been implicated as etiologic factors. Chronic suppurative otitis media and the resulting chronic inflammation may lead to epithelial metaplasia.^{11,17} The presentation of a polypoid mass and purulent discharge, which also accompany the more commonly occurring chronic otitis media, led to the wrong clinical diagnosis.

In the literature, cases similar to ours have been reported. Kloeck *et al.*¹⁸ reported a case of Wegener's granulomatosis presenting with polypoid tissue in the EAC. Elsurer *et al.*¹⁹ reported a case of ceruminous adenoma mimicking furunculosis in the EAC. Neoplasms of the EAC may present as a polypoid mass, which can obscure the lumen and protrude through the tympanic membrane and middle ear. Histologic evaluation is necessary to differentiate simple inflammatory polyps from other underlying disease processes or as neoplasia, as in the present case.

Conclusion

The ENT surgeon should keep the possibility of neoplasia in mind when dealing with patients with

chronic otitis media. Discharge, hearing loss, and an aural polyp, although commonly associated with chronic otitis media, should prompt biopsy, particularly if associated with pain, bloody discharge, or facial palsy. Although malignant tumours of ceruminous glands are very rare, they should be considered in the differential diagnosis of middle and external ear pathologies.

References

1. Batsakis JG. *Tumors of the Head and Neck*. Williams & Wilkins, Baltimore; 1979:429-430.
2. Thompson LD, Nelson BL, Barnes EL. Ceruminous adenomas: a clinicopathologic study of 41 cases with a review of the literature. *Am J Surg Pathol*. 2004;28:308-318.
3. Garin P, Degols JC, Delos M, Marbaix E. Benign ceruminous tumours of the external ear canal. *J Otolaryngol*. 1999;28:99-101.
4. Tzagaroulakis A, Paxsalidis J, Papadimitriou N, *et al*. Recurrent ceruminous adenocarcinoma of the external auditory canal. *ORL J Otorhinolaryngol Relat Spec*. 2003;65:300-302.
5. Wetli CV, Pardo V, Millard M, Gerston K. Tumors of ceruminous glands. *Cancer*. 1972;29:1169-1178.
6. Koyuncu M, Karagoz F, Kiliaçarslan H. Pleomorphic adenoma of the external auditory canal. *Eur Arch Otorhinolaryngol*. 2005;262:969-971.
7. Michaels L, Thompson LD. Ceruminous gland neoplasms of external auditory canal and cylindroma. In: Barnes L, Eweson JW, Reichart P, Sidransky D, eds. *World Health Organization Classification of Tumors Pathology and Genetics Head and Neck Tumors*. IARC Press, Lyon, France; 2005:331-333.
8. Lassaletta L, Patron M, Oloriz J, Perez R, Gavilan J. Avoiding misdiagnosis in ceruminous gland tumours. *Auris Nasus Larynx*. 2003;30:287-290.
9. Conlin PA, Mira JL, Graham SC, Kaye KS, Cordero J. Ceruminous gland adenoid cystic carcinoma with

- contralateral metastasis to the brain. *Arch Pathol Lab Med.* 2002;126:87-89.
10. Arriaga M, Curtin H, Takahashi H, Hirsch BE, Kamerer DB. Staging proposal for external auditory meatus carcinoma based on preoperative clinical examination and computed tomography findings. *Ann Otol Rhinol Laryngol.* 1990;99:714-721.
 11. Moffat DA, Wagstaff SA, Hardy DG. The outcome of radical surgery and postoperative radiotherapy for squamous carcinoma of the temporal bone. *Laryngoscope.* 2005;115:341-347.
 12. Antonio SM, Hirsch, EB. Malignant Tumors of the Temporal Bone - Otolaryngology and Facial Plastic Surgery. Medicine Clinical Reference, Updated Continually. Available at: [Http://www.emedicine.com/ent](http://www.emedicine.com/ent). Article Last Updated April 30, 2007.
 13. Schwentner I, Obrist P, Thumfart W, Sprinzl G. Distant metastasis of parotid gland tumors. *Acta Otolaryngol.* 2006;126:340-345.
 14. Perzin KH, Gullane P, Conley J. Adenoid cystic carcinoma involving the external auditory canal. A clinico-pathologic study of 16 cases. *Cancer.* 1982;50:2873-2883.
 15. Koopot R, Reyes C, Pifarré R. Multiple pulmonary metastases from adenoid cystic carcinoma of ceruminous glands of external auditory canal. A case report and review of the literature. *J Thorac Cardiovasc Surg.* 1973;65:909-913.
 16. Turner HA, Carter H, Neptune WB. Pulmonary metastases from ceruminous adenocarcinoma (cylindroma) of external auditory canal. *Cancer.* 1971;28:775-780.
 17. Takahashi K, Yamamoto Y, Sato K, Sato Y, Takahashi S. Middle ear carcinoma originating from a primary acquired cholesteatoma: a case report. *Otol Neurotol.* 2005;26:105-108.
 18. Kloeck I, Crols R, De Belder T, Vandist V, Schmelzer B. Wegener's granulomatosis presenting as otomastoiditis. A case report. *B-ENT.* 2006;2:7-12.
 19. Elsurer C, Senkal HA, Baydar DE, Sennaroglu L. Ceruminous adenoma mimicking furunculosis in the external auditory canal. *Eur Arch Otorhinolaryngol.* 2007;264:223-225.

Adin Selcuk
 50. Sok 1/10
 Bahcelievler 06500
 Ankara, Turkey
 Fax: +00 312 310 3460
 E-mail: sadin27@yahoo.com