



Case Report

Ceruminous Adenocarcinoma of External Auditory Canal: A Case Report

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The external auditory canal contains ceruminous glands, which are modified apocrine sweat glands, along with sebaceous glands. Tumors that originate from ceruminous glands are very rare; thus, the classification, clinical behavior, and management of these tumors remain debatable. Here we present a case of ceruminous adenocarcinoma arising from the external auditory canal. Although most authors advise more aggressive therapy, our patient was treated with local en bloc resection of the tumor followed by intensity modulated radiotherapy and had no recurrence for 3 years. We suggest that limited surgery with safe margins followed by radiotherapy is an alternative choice of treatment in selected patients with ceruminous adenocarcinoma. Further reports are required to support this outcome.

Keywords: Ear neoplasms, adenocarcinoma, ear canal

INTRODUCTION

Ceruminous glands are modified apocrine glands located within the dermis of the skin overlaying the cartilaginous portion of the external auditory canal (EAC) ^[1]. Watery secretions of ceruminous glands, along with sebaceous gland secretions, are drained into the hair sacs of fine hairs in EAC, together forming the cerumen (wax) ^[2]. Ceruminous gland-originated tumors of EAC are rare neoplasms. Because of the varied clinical and histological manifestations of these tumors, controversies currently exist regarding their nomenclature, classification, histopathological features, diagnosis and treatment.

Until Wetli et al. ^[3] proposed a classification based on histological features in 1972, these tumors were referred to as ceruminoma, a term which does not discriminate between benign and malign lesions. Ceruminous adenocarcinomas are malign glandular tumors of EAC that do not show specific clinical symptoms. As most cases are advanced, treatment results are unsatisfactory ^[3,4]. Herein we report our case of ceruminous adenocarcinoma of EAC, which presented as a polypoid mass. The patient was treated with local en bloc resection of the tumor followed by radiotherapy.

CASE PRESENTATION

A male patient aged 54 years, without any chronic disease, previous history of surgery, or trauma, was referred to our clinic with mild hearing loss in his left ear for about 2 months. He had no discharge, dizziness, tinnitus, vertigo, earache, or facial paralysis. Physical examination showed a red-violet-colored, polypoid, non-tender mass emerging from the posterior wall of left EAC that prevented the visualization of the tympanic membrane. Examination of the nose and throat showed no abnormality, and no pathological lymphadenopathy on the neck was detected by ultrasonography. Pure-tone audiometry revealed mild conductive hearing loss in the left ear. Magnetic resonance imaging (MRI) of the temporal bones revealed an isointense polypoid lesion, approximately 15×10 mm in greatest dimensions (Figure 1). Signs of erosion on bony EAC were also observed with computerized tomography (CT) (Figure 2). An incisional biopsy was performed and the histopathological diagnosis was initially reported as ceruminous adenoma. As there were signs of bone erosion, a frozen section was performed during the planned surgery to prevent misdiagnosis and inadequate surgery. As the frozen section was indecisive, local en bloc resection of the lesion along with cortical mastoidectomy was performed. The posterior wall of EAC, adjacent to the tumor, was also drilled superficially. Intraoperative visualization of the lesion revealed that despite bone erosion, there was no tumor extension in the mastoid cavity. The tumor was classified as T2 N0 M0 according to the modified Pittsburgh (revision 2002) classification. After pathological examination of the specimen, it was reported as ceruminous adenocarcinoma. Marginal skin biopsies did not reveal any abnormalities. After the surgery, the patient experienced an uneventful recovery. The patient also received 30 sessions of intensity-modulated radiotherapy (IMRT) (Synergy; Elektra, Sweden) at a total dose of 60 Gy. The patient's hearing was well preserved after therapy, and despite mild sensorineural hearing loss in the high frequencies, pure-tone average was normal in the left ear. Long term follow-up was planned and there was no evidence of

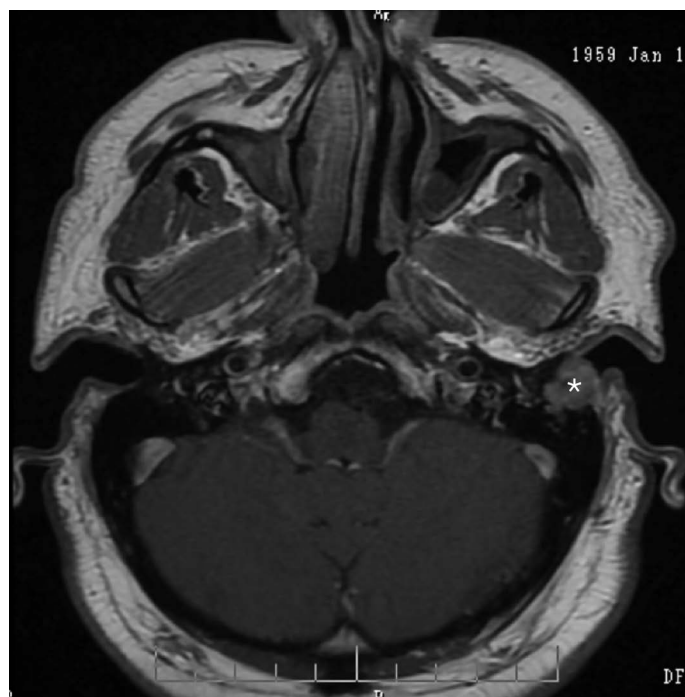


Figure 1. Axial contrast-enhanced T1 MRI of the tumor on the left external auditory canal (*: tumor)

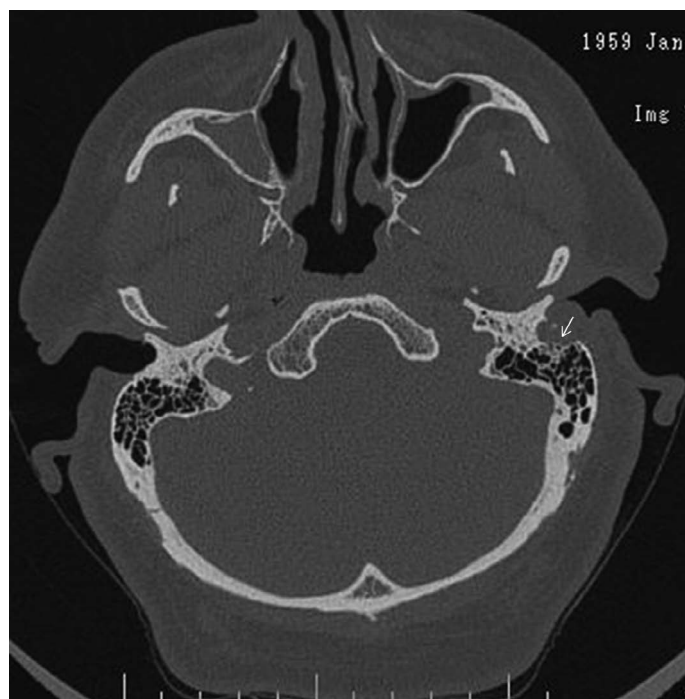


Figure 2. Axial CT image of the tumor on the left external auditory canal (✓: erosion on the posterior wall of the left external auditory canal)

recurrence for 3 years after the surgery (Figure 3-5). Written informed consent was obtained from the patient for publication of the report.

DISCUSSION

For a long time, ceruminous gland-originated tumors were lumped together as ceruminoma, a term that does not discriminate between benign and malign lesions, until Wetli et al. [3] proposed a classification based on histological features in 1972. There are four categories



Figure 3. Endoscopic view of the external auditory canal after 3-year follow-up (*: tympanic membrane)

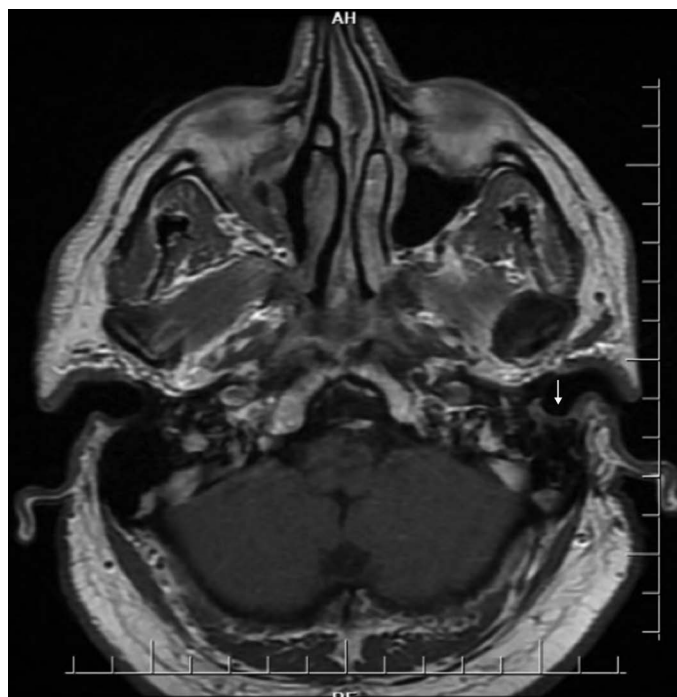


Figure 4. Despite postoperative changes, contrast enhanced MRI of left ear showed no sign of recurrence 3 years after surgery (↓: surgery site)

in this classification: ceruminous adenomas, ceruminous adenocarcinomas, adenoid cystic carcinomas, and pleomorphic adenomas [3]. Some additional entities such as syringocystadenoma papilliferum and mucoepidermoid carcinoma have been proposed for inclusion in this classification [5,6].

Ceruminous adenocarcinomas are rare malignant tumors that originate from the ceruminous glands of EAC. Differential diagnosis between ceruminous adenocarcinoma and ceruminous adenoma is



Figure 5. No sign of recurrence was observed in the axial CT image of the temporal bone (↓ : surgery site)

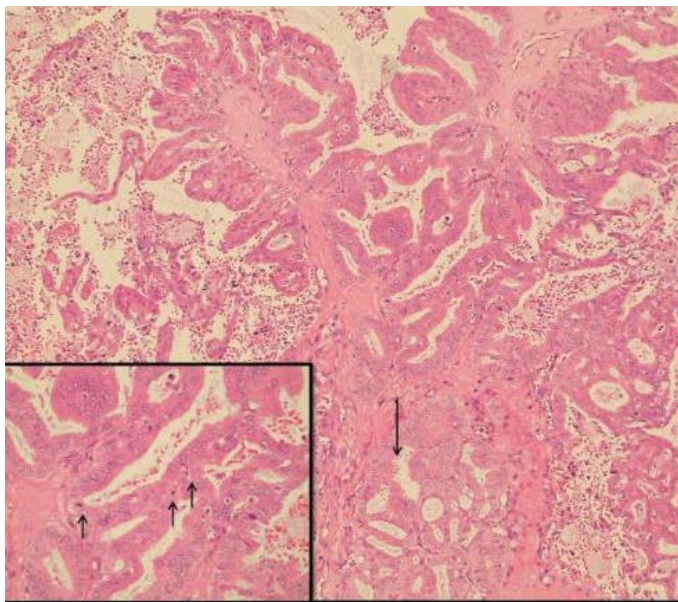


Figure 6. Adenocarcinoma with papillary and glandular structures. Within some tumor cells that formed glandular structures, apocrine secretion (↓) could be seen (hematoxylin–eosin stain, original magnification ×40). Tumor cells with a wide eosinophilic cytoplasm, large nucleus, and vesicular chromatin showed cellular pleomorphism and frequent mitoses (↑) (hematoxylin–eosin stain, original magnification ×400).

difficult. Although pathological evaluations of ceruminous adenocarcinomas demonstrate significantly more infiltration, perineural invasion, irregular gland formation, pleomorphism, prominent nucleoli, increased mitotic figures, atypical mitotic figures, and tumor necrosis, only few of these features are usually observed, which complicates the distinction between adenocarcinoma and adenoma^[7]. Primary lung, breast, or kidney cancer metastasis should be excluded when diagnosing this tumor^[8].

In our case, pathological examination showed an infiltrative tumor consisting of pleomorphic and atypical cells with a wide eosinophilic cytoplasm, large nucleus, vesicular chromatin, apocrine snout, and prominent nucleoli that formed glandular, cribriform, and papillary structures (Figure 6). Increased mitotic figures were observed. Despite bone erosion, perineural and vascular invasion was not observed. Immunohistochemical stains were performed; the tumor stained positive for keratin-7 and negative for keratin-20, p63, s-100, and CD117. Ki 67 showed mild positivity. With these findings, the tumor was considered to be ceruminous adenocarcinoma.

Otalgia, mass, and hearing changes are the most common symptoms reported for ceruminous gland tumors in the literature^[9]. In addition, facial nerve involvement may result in facial nerve paralysis^[6].

Although cervical lymph node metastasis is uncommon, metastasis to the bones, lungs, and brain may be possible^[4,8]. CT is helpful in detecting bone erosion and destruction as well as tumor extension^[10]. The main treatment is surgery. Most authors recommend aggressive surgery that may be followed by radiotherapy^[4,10]. As a surgical procedure, Hicks suggested initial wide en bloc surgical resection. If there is tumor extension to the middle ear, resection of the temporal bone and adjacent structures is recommended^[4]. Although radiation therapy is recommended for other types of ceruminous gland carcinomas only for palliation and recurrence, it is thought to be beneficial for the ceruminous adenocarcinoma NOS group as part of the initial therapy^[9].

In our case, we preferred limited surgical therapy because the disease was not at an advanced stage. After total resection of the tumor, the patient received 60 Gy IMRT. Although recurrences usually occur within months after the initial treatment, it is reported that some may develop up to 7 years after the surgery^[4]. Therefore, cautious long term follow-up on a 3-monthly basis is planned. Limited surgery with healthy margins followed by radiotherapy may be an alternative choice of treatment in selected patients with ceruminous adenocarcinoma to prevent morbidity associated with aggressive surgery. Further reports and studies are required to compare survival outcomes and treatment options.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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