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Case Report

Adenoid cystic carcinoma of the external auditory canal

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Abstract

Primary cancers of the external auditory canal (EAC) are rare and most are squamous cell carcinomas. We report the case of a 78-year-old man who visited our institution with a 5-month history of right-side intermittent otalgia and ear fullness. Otoscopic examination showed a bulging mass arising from the superior and posterior aspects of the right EAC, and incision biopsy confirmed the lesion as adenoid cystic carcinoma (ACC). Lateral temporal bone resectionin conjunction with total parotidectomy and neck dissection was subsequently performed. Postoperative adjuvant radiotherapy was administered and no recurrence was noted at a 26-month follow-up. We review the medical literature on the topic and suggest that early diagnosis is still the best option for successful treatment of this neoplasm. ACC arising in the EACmust be removed using radical procedures to increase the chance of local control. Subsequent metastasis that tends to develop in the lungs and regional lymph nodes is best evaluated regularly using computed tomography examination.

Keywords: adenoid cystic carcinoma; external auditory canal; surgery; temporal bone resection

1. Introduction

Malignant tumors involving the external auditory canal (EAC) and temporal bone are exceedingly rare, and the most common type is squamous cell carcinoma.[1] The presence of adenoid cystic carcinoma (ACC) arising in the EAC is therefore infrequently encountered. ACC of the head or neck is usually found in the salivary glands, oral cavity, palate, nasal cavity, and nasopharynx.[2] Since 1894, there have been only 106 cases of ACC involving the EAC reported the English literature. The natural history of ACC of the EAC is characterized by an indolent clinical course, which usually leads to a late diagnosis. The treatment goal includes complete surgical extirpation and a clear margin because of the high risk of repeat local recurrence. It is not uncommon for distant metastasis, mainly to the lungs, to occur over the course of many years.[3,4]

Because of the rarity of ACC of the EAC, most of the observations drawn from various reports lack detailed comparisons of pathological findings and long-term outcome follow-up. Here, we present our own case along with a review and discussion of the literature to date.

2. Case report

A 78-year-old man presented with a 5-month history of right-side intermittent otalgia and ear fullness. His past history was unremarkable. Otoscopic examination showed a bulging mass with an irregular surface arising in the right EAC (Fig. 1A). High-resolution computed tomography (CT) of the temporal bone showed an ill-defined, moderately enhanced, soft tissue mass protruding from the superior and anterior walls of the EAC (Fig. 1B), measuring 10 mm at the widest point, with adjacent bone erosion. Incision biopsy confirmed the diagnosis of ACC. Further information from magnetic resonance imaging (MRI) with gadolinium suggested that the superficial lobe of the parotid gland and the anterior portion of the temporal bone were also

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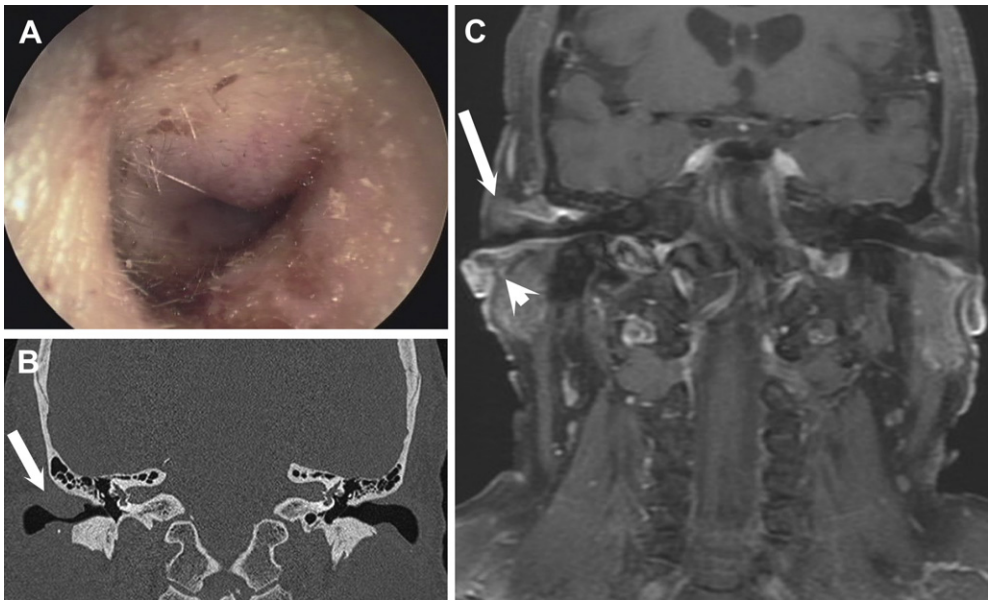

Fig. 1. (A) Subcutaneous mass bulging from the superior and posterior aspects of the external auditory canal (EAC). (B) Coronal computed tomography scan of the temporal bone demonstrating a soft tissue mass (arrow) protruding from the superior wall of the EAC. (C) Magnetic resonance imaging shows that the superficial lobe of the parotid gland (arrowhead) and the anterior portion of the temporal bone (arrow) may have been invaded by the mass lesion.

invaded (Fig. 1C). In view of the fact that the tumor had eroded the osseous EAC but had not invaded the middle ear cavity, and based on a presumed clinical T3N0M0 stage using University of Pittsburgh TNM staging for EAC carcinoma,[5,6] surgery entailing lateral temporal bone resection in conjunction with total parotidectomy and selective neck dissection levels I–III was performed. In brief, the lateral temporal bone resection was approached with a wide postauricular incision followed by a separate incision made around the tragus and a portion of the conchal cartilage to isolate the tumor from the normal auricle. The facial nerve was skeletonized in its course through the stylomastoid foramen up to the geniculate ganglion and was found to be tumor-free. The malleus–incus joint was then disarticulated. The EAC, with the tumor lesion in its osseous and cartilaginous parts, was completely isolated and removed en bloc, along with the tympanic membrane and the ossicles (Fig. 2). The defect left over the lateral temporal region was reconstructed by rotation of the sternocleidomastoid (SCM) muscle and temporalis muscle flap.

Histologically, the tumor cells were in sheets and showed a predominantly cribriform pattern with scant intervening stroma. The cells were basaloid, with a scanty cytoplasm and hyperchromatic nuclei (Fig. 3). The lumina in the cribriform areas contained material that was positive in a periodic acid Schiff test. Few areas of necrosis were noted. Immunohistochemistry analysis revealed intense and diffuse positive staining for cytokeratin, vimentin, and muscle actin, and focal staining for S-100 protein. These findings confirmed the diagnosis of ACC of the EAC with perineural invasion and involvement of the tympanic portion of the temporal bone. The parotid gland and neck nodes were not invaded. Although clear surgical margins were achieved, postoperative adjuvant radiotherapy with doses of 6000 cGy in 34 fractions was administered. Good progress was noted during the patient's 26-month follow-up, and the reconstructed external ear showed excellent esthetic and structural support (Fig. 4).

3. Discussion

ACC (also called cylindroma in the older literature), although it seldom arises in the EAC, is the most common malignant lesion of glandular origin.[3,7,8] Our review of 106 cases of ACC arising from the EAC is based on the PubMed indexed English literature. Of the 106 ACC cases, 93 had clearly identified gender data, with a female-to-male ratio of 1.58 (57 vs. 36), indicating that females are more prone to being affected than males. Although ACC may appear at any age, the peak incidence is around the fourth and fifth decades of life. This tumor has an indolent clinical course and usually grows for years before causing symptoms, and thus tends to result in late diagnosis.

The most common clinical manifestations are otorrhea, pain, hearing loss, bleeding, and a mass or polyp in the ear canal.[9–11] Differential diagnosis of a mass lesion in the EAC includes adenoma, papilloma, tuberculosis, and other neoplasms such as mucoepidermoid carcinoma, adenocarcinoma, basal cell carcinoma, and squamous cell carcinoma. Because of the propensity of ACC toward aggressive repeated local recurrences and distant metastases, it is important for clinicians to determine the extent of tumor invasion for adequate surgical planning and management.[12] The use of MRI in cases of ACC of the EAC has not been fully delineated.[13] ACC occurring elsewhere in the head and neck may show low-signal intensity on T1-weighted (T1w) MRI images and high or low intensity on T2-weighted (T2w) images, depending on its cellularity. Our case presented with low intensity on T1w but high intensity on T2w images, indicating low cellularity, which was compatible with predominantly cribriform histological features.