Mucoepidermoid Carcinoma of the External Auditory Canal: Case Report

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This study reports a case of mucoepidermoid carcinoma (MEC) of the external auditory canal, which to date has only been described once in literature. Because the lesion is extremely rare, it is particularly difficult to classify it into stages following normal diagnostic parameters. This obviously limits the possibilities of treatment that consequently are either empirical or based on those of squamous cell carcinoma. The problems in the diagnosis and the possible methods of treatment of mucoepidermoid carcinoma are discussed.
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Neoplastic lesions of the external auditory canal (EAC) are quite rare. In fact they occur in 1 or 2 cases in 5 million people and affect in particular those between 50 and 60 years of age.[1-3]

In the past the term *ceruminoma* comprehended benign and malignant tumors of the EAC whether they were of epithelial or glandular origin.[4] Recently, the Armed Forces Institute of Pathology (Bethesda, MD, 1991) and the World Health Organization have reclassified the histological pathologies, forming a new simplified version in substitution of the old method. Squamous cell carcinomas followed by adenoidal cystic carcinomas and basaliomas are respectively the most frequent forms of tumor,[5] whereas mucoepidermoid tumors are extremely rare. From a thorough review of the literature available, we found that only 4 cases of this type of tumor have been reported, and of these only 1 was situated in the EAC.[6-9] The aim of this study is to present a second case of mucoepidermoid carcinoma (MEC) of the EAC that are diagnosed and treated surgically and to propose the possible methods of diagnosis and treatment.

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CASE REPORT

A 62-year-old woman was seen in March 2000 complaining of slight left-sided otalgia. The patient stated that she had 2 small lumps that she felt during palpation of the EAC and that they had been present for about 6 years. On otoscopy, they were seen to be bright red in color, situated on the inferior wall of the cartilaginous portion of the EAC, and less than 8 mm in size. They caused her some pain, which slightly increased on palpation. A biopsy was performed that revealed a medium-grade malignant MEC (Fig 1). A computed tomography scan with contrast medium and magnetic resonance imaging of the head and neck were performed to establish the extent of the lesion and the possibility of metastases. These confirmed the position of the lesion and showed that it had irregular margins and a tendency to infiltration, without it, however, having invaded the cartilage. Some of the cervical lymph nodes seemed to be slightly swollen but had regular margins and were judged by the radiologist to be free of metastases. Surgery with sleeve resection of the external auditory canal was planned. Frozen section documented an involvement of the chondro-osseous junction obliging us to manage the neoplasm with a lateral temporal bone resection. In view of the medium-grade malignancy of the MEC and the risk of hidden metastases in the lymph nodes, a total parotidectomy and unilateral functional neck dissection from level I to level IV were also performed. The final histological diagnosis confirmed medium grade of MEC that had reached but not infiltrated the cartilaginous area. The extir-

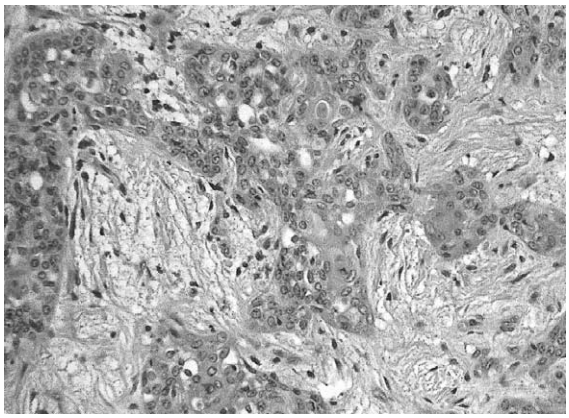

Fig 1. Hematoxylin eosin staining (original magnification 25×). Both the epidermoid and mucous components are clearly shown.

pated parotid parenchyma was free of neoplastic infiltration, whereas one of the lymph nodes from the functional neck dissection (level I) showed signs of metastases. The patient has been seen repeatedly since surgery and in the 2-year follow-up period has shown no signs of relapse.

DISCUSSION

MECs are malignant tumors that frequently originate in the mayor and minor salivary glands (11% and 8%, respectively).[10] They may also occur in other glandular structures in their normal anatomic sites such as the anal, esophageal, tracheobronchial mucous glands, and the biliary and ceruminous duct, or they may arise in ectopic sites such as the hypophysis, the mandible, and the neck.[9]

The ceruminous glands are known to be situated in the deep dermis of the EAC. The mechanism that provokes tumoral degeneration is not clearly understood, but Soh et al[9] have advanced 4 etiopathogenetic hypotheses: those originating from the seromucous glands because of metastatic or functional alterations of the mucosal epithelium, those caused by chronic irritation that causes squamous metaplasia of the mucosa, seromucous and minor salivary glands implanted in the ear during embryogenesis, and finally those caused by secondary invasion of a tumor located in an adjacent primary site. Each single hypothesis is valid and reliable on its own but the possibility that they may act together should also be taken into consideration. Soh et al's[9] interpretations of glandular tumors of both the middle and external ear clearly also applies to MECs. In the case of the latter, the only possible means of diagnosis is histological. Under histological examination, MECs appear to be composed of scattered solid and cystic mucinous areas. In the solid parts, the cells are arranged in nests, are tubular in structure, and have different characteristics very similar to those of mucous squamous cells, whereas other cells have intermediate histopathological characteristics. It is exactly this percentage of the different cells that makes it possible to establish a low-, medium-, or high-grade malignancy. In low-grade tumors, there is a higher percentage of mucous cells, and, in high-grade tumors of squamous cells and in medium-grade tumors, there is an equal percentage of glandular and epithelial cells. The histological diagnosis of MECs is often difficult and not always completely reliable. For example, low-grade MECs may be confused with adenomas because they have a low mitotic rate and very little atypia; thus, the diagnosis of MEC may be established only if intermediate and epithelioid cells are present. High-grade MECs instead are very similar to poorly differentiated squamous carcinoma except for the fact that the former has intermediate and mucin-secreting cells, a difference that gives a useful indication in the histological diagnosis. The clinical diagnosis is based on classical otoscopy. MECs do not particularly differ from other neoplasms found in the external auditory canal. They appear as solid reddish masses that are variable in shape (usually irregular) and may be located in any part of the EAC. They are normally neither painful nor tender, but pain is sometimes present as a result of concomitant infection, compression (as in our case), or infiltration of the nerves. Imaging techniques serve to establish the extent of the lesion and to assess possible metastatic involvement of the lymph nodes. Their informations are essentially not specific. The images of high-grade malignant tumors both on computed tomography scan and magnetic resonance imaging are identical to those of other malignant neoformations in the EAC with irregular infiltrating margins. Moreover, low-grade malignant tumors with a regular surface and well-

TABLE 1. Review of the Literature

Authors	Site	TNM	Grade of Malignancy	Treatment	Follow-up (months)	Outcome
Kinney and Wood (1987)	External auditory canal	n.p.	high	Ear canal resection + superficial parotidectomy resection	18	Persistent disease
Landman and Farmer (1991)	Posterior mid helix	T1N0M0	low	Excision by Mohs surgical technique	8	Free from disease
Shotton et Al. (1993)	Cheek involving ear	T4N0M0	n.p.	Superficial parotidectomy + external radiotherapy	0	Dead
Soh et Al. (1996)	Middle ear	T4N1M0	n.p.	Radiotherapy (not specified	13	Dead

circumscribed margins are very similar in appearance to benign lesions.[11] The very fact that MECs in the EAC are extremely rare makes it highly difficult if not impossible to establish a reliable system of classification into stages. In fact, of the 4 cases of MECs in the ear described in literature, only 1 reported by Kinney and Wood[6] involved the EAC (Table 1).

The lack of precise guidelines in the classification and treatment of this form of tumor obliged us to adopt the same criteria for squamous cell carcinomas of the external auditory canal. Several methods of classification do exist; however, the one proposed by Pittsburgh University, that is a combination of the clinical and imaging technique results before surgery, has recently come to be considered the most useful (Table 2). [12]

Our case involved the chondro-osseous junction and may be classified as a T2. Because the algorithm for high-grade squamous cell carcinomas indicates partial lateral temporal bone resection, the same form of surgery also applies to this form of lesion. This modality is combined with functional neck dissection in those cases in which these are involved. The effective necessity of the latter form of surgery is based on the results of the preoperative imaging techniques, but these unfortunately are somewhat unreliable and give rise to doubt because they do not always clearly manifest nodal involvement. At this point, it should be kept in mind that metastases of the lymph nodes indicates a worse prognosis and automatically puts the patients into the category of advanced lesions. Furthermore, as Jesse et al[13] already stressed, the first form of surgical treatment of malignant neoplasms of the EAC must necessarily be radical because subsequent surgery is virtually ineffective and only palliative. It is therefore particularly important to adopt a highly aggressive strategy at the very beginning. We

TABLE 2. University of Pittsburgh Staging System for Squamous Cell Carcinoma of the Temporal Bone and Stage

T1	Tumour limited to the external auditory canal without bony erosion or evidence of soft-tissue extension
T2	Tumour with limited external auditory canal erosion (not full thickness) or radiologic findings consistent with limited (<0.5 cm) soft-tissue involvement
T3	Tumour eroding the osseous external auditory canal (full thickness) with limited (<0.5 cm) soft tissue involvement, or involving middle ear and/or mastoid or causing facial paralysis at presentation
T4	Tumour eroding the cochlea, petrous apex, medial wall of middle ear, carotid canal, jugular foramen, or dura, or with extensive (>0.5 cm) soft-tissue involvement
N status	Involvement of lymph nodes is a poor prognostic finding and automatically places the patient in a higher category (ie, stage III (T1, N1) or stage IV (T2,3,4, N1)
M status	M1 immediately places a patient in the stage IV category

performed functional neck dissection even though the results of the imaging scans were negative, basing our decision on the histological appearance of the lesion. The tumor in question was a medium-grade MEC with a high potential risk of metastasis. The final histological examination proved our choice to be right. Although it is not possible to draw final conclusions, it would, however, seem reasonable not to postpone lymph node dissection when the surgeon is faced with a clinically and histologically proved highly aggressive squamous cell or mucoepidermoid tumour.

Alternative measures would only apply to low-grade MECs. In these cases, lymph node dissection could be delayed, but in the meantime the patient should be kept under close surveillance. Because of the clinical features of the MEC described in this study, we are not in a position to be able to judge the validity of postoperative radiotherapy. It would, however, seem logical to reserve this form of treatment for the particularly aggressive forms of tumor.

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