Microcystic Adnexal Carcinoma of the External Auditory Canal

Hsiu-Hui Chiu, MD; Chieh-Shan Wu, MD; Chen-Yu Chien, MD; Kun-Bow Tsai, MD; Leong-Perng Chan, MD

PRACTICE **POINTS**

- Microcystic adnexal carcinoma is a locally aggressive malignant adnexal tumor with a high potential for local recurrence.
- The skin of the head, particularly in the nasolabial and periorbital regions, most often is involved.
- Once diagnosed, the tumor should be surgically excised. Because local recurrence is common and may occur several decades after excision, lifetime follow-up for recurrence is essential.

To the Editor:

Microcystic adnexal carcinoma (MAC), described by Goldstein et al[1] in 1982, is a relatively uncommon cutaneous neoplasm. This locally aggressive malignant adnexal tumor has high potential for local recurrence. The skin of the head, particularly in the nasolabial and periorbital regions, most often is involved.[2] Involvement of the external auditory canal (EAC) is relatively rare. We report a case of MAC of the EAC.

A 52-year-old man presented with 1 palpable nodule on the right EAC of approximately 1 year's duration. The lesion was asymptomatic, and the patient had no history of radiation exposure. The patient was an airport employee required to wear an earplug in the right ear. Endoscopic examination identified a 1×1 cm^2 erythematous nodule on the anterior inferior quadrant of the right external ear canal orifice (Figure 1). Axial and coronal computed tomography demonstrated a soft tissue mass in the right EAC without any bony erosion. No clinical signs of regional lymphadenopathy or distant metastasis were present. Excision was performed under microscopic visualization.

Histopathology of the nodule showed marked proliferation of multiple keratin-containing cysts, irregular ductal structures, and solid epithelial nests in the deep dermis (Figure 2). Irregular ductal structures with 2 cell layer walls and several epithelial strands or small nests of tumor cells within desmoplastic stroma were noted (Figure 3). No perineural infiltration or tumor infiltration existed at the margin. Based on the clinical and histopathologic findings, the final diagnosis was MAC. Complete resolution was noted after the excision. The patient returned for regular follow-up and no signs of recurrence were noted for 7 years postoperatively.

FIGURE 1. Erythematous 1×1 cm^2 nodule on the anterior inferior quadrant of the right external ear canal orifice.

Dr. Chiu is from the Department of Dermatology, Pingtung Christian Hospital, Taiwan. Dr. Wu is from the Department of Dermatology, Kaohsiung Veterans General Hospital, Taiwan, and the Department of Dermatology, Faculty of Medicine, College of Medicine, Kaohsiung Medical University. Drs. Chien and Chan are from the Department of Otolaryngology-Head and Neck Surgery, Kaohsiung Medical University Hospital, and Kaohsiung Municipal Ta-Tung Hospital, Kaohsiung Medical University. Dr. Chan also is from the Institute of Clinical Medicine, Kaohsiung Medical University. Dr. Tsai is from the Department of Pathology, Kaohsiung Municipal Hsiao-Kang Hospital, Kaohsiung Medical University.

The authors report no conflict of interest.

Correspondence: Leong-Perng Chan, MD, No.100, Tzyou 1st Rd, Kaohsiung 807, Taiwan (oleon24@yahoo.com.tw).

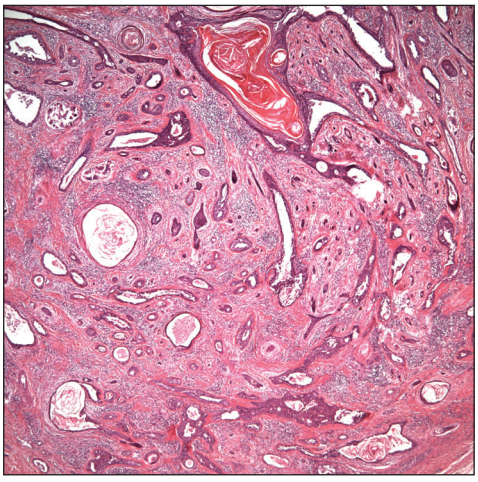

FIGURE 2. Microcystic adnexal carcinoma in the external auditory canal with marked proliferation of multiple keratin-containing cysts, irregular ductal structures, and solid epithelial nests in the deep dermis (H&E, original magnification ×20).

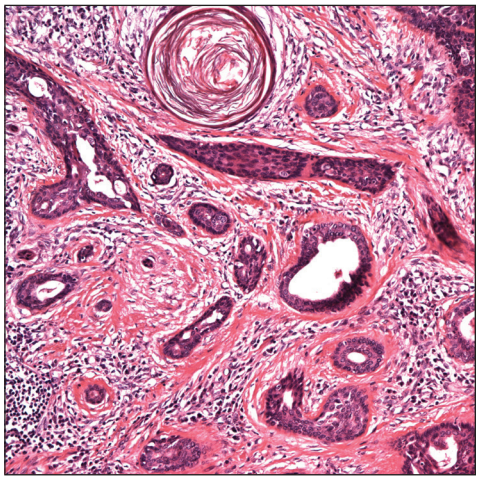

FIGURE 3. Microcystic adnexal carcinoma in the external auditory canal with irregular ductal structures with 2 cell layer walls and several epithelial strands or small nests of tumor cells within desmoplastic stroma (H&E, original magnification ×200).

Microcystic Adnexal Carcinoma of the External Auditory Canal

Reference (Year)	Sex/Age, y	History of Radiation	Location	Treatment
Hunt et al[5] (1995)	F/49	NM	EAC and spreading along facial nerve to parotid gland	Surgical excision, radiotherapy
Chi et al[6] (2002)	F/46	None	EAC	Surgical excision
Ozbek et al[7] (2004)	M/22	NM	EAC, middle ear	Surgical excision
Beer et al[8] (2005)	M/28	>15 y ago	EAC, parotid gland, neck	Surgical excision, radiotherapy
Current report	M/52	None	EAC	Surgical excision

Abbreviations: F, female; NM, not mentioned; EAC, external auditory canal; M, male.

Microcystic adnexal carcinoma, also known as sclerosing sweat duct (syringomatous) carcinoma, malignant syringoma, and syringoid eccrine carcinoma, is characterized by slow and locally aggressive growth with high likelihood of perineural invasion and frequent recurrence.[2] Regional lymph node metastasis is uncommon, and systemic metastasis is rare.[2-4]

Although the head most often is involved, a PubMed search of articles indexed for MEDLINE using the terms *microcystic adnexal carcinoma* and *external auditory canal* revealed 4 cases (Table).[5-8] Our report adds another case of MAC arising solely in the EAC. Although the etiology of MAC is unknown, prior studies indicated that radiotherapy is a risk factor for MAC. Other possible risk factors include UV light exposure and immunodeficiency.[2] Our patient had no history of these factors and experienced chronic friction caused by use of an occupational unilateral earplug, which may be a notable factor. Locations of MAC arising outside the head region include the axilla, vulva, breast, palm, toe, perianal skin, buttock, chest, and an ovarian cystic teratoma.[3,9] Friction commonly occurs in many of these areas. Therefore, we propose that friction may be a risk factor for MAC.

Microcystic adnexal carcinoma should be included in the differential diagnosis of any slowly growing cutaneous tumor, even in the EAC. Once diagnosed, the tumor should be surgically excised. Because local recurrence is common and may occur several decades after excision, lifetime follow-up for recurrence signs is essential.

REFERENCES

1. Goldstein DJ, Barr RJ, Santa Cruz DJ. Microcystic adnexal carcinoma: a distinct clinicopathologic entity. *Cancer*. 1982;50:566-572.
2. Brenn T, Mckee PH. Tumors of the sweat glands. In: McKee PH, Calonje E, Granter SR, eds. *Pathology of the Skin With Clinical Correlations*. 3rd ed. Philadelphia, PA: Elsevier Mosby; 2005:1647-1651.

3. Ohtsuka H, Nagamatsu S. Microcystic adnexal carcinoma: review of 51 Japanese patients. *Dermatology*. 2002;204:190-193.
4. Yu JB, Blitzblau RC, Patel SC, et al. Surveillance, Epidemiology, and End Results (SEER) database analysis of microcystic adnexal carcinoma (sclerosing sweat duct carcinoma) of the skin. *Am J Clin Oncol*. 2010;33:125-127.
5. Hunt JT, Stack BC Jr, Futran ND, et al. Pathologic quiz case 1. microcystic adnexal carcinoma (MAC). *Arch Otolaryngol Head Neck Surg*. 1995;121:1430-1433.
6. Chi J, Jung YG, Rho YS, et al. Microcystic adnexal carcinoma of external auditory canal: report of a case. *Otolaryngol Head Neck Surg*. 2002;127:241-242.
7. Ozbek C, Celikkanat S, Beriat K, et al. Microcystic adnexal carcinoma of the external ear canal. *Otolaryngol Head Neck Surg*. 2004;130:148-150.
8. Beer KT, Bühler SS, Mullis P, et al. A microcystic adnexal carcinoma in the auditory canal 15 years after radiotherapy of a 12-year-old boy with nasopharynx carcinoma. *Strahlenther Onkol*. 2005;181:405-410.
9. Nadiminti H, Nadiminti U, Washington C. Microcystic adnexal carcinoma in African Americans. *Dermatol Surg*. 2007;33:1384-1387.