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Epstein Barr Virus–Associated Lymphoepithelial Carcinoma in the Middle Ear

Leh-Kiong Huon, MD, Pa-Chun Wang, MD, MSc, and Shih-Hung Huang, MD

Case Report

Histopathologically, lymphoepithelial carcinoma (LEC) is characterized by individual, sheets, or nests of poorly to undifferentiated epithelial cells. The malignant epithelial cells are surrounded and infiltrated by prominent components of small mature lymphocytes and plasma cells. The most frequent site of LEC occurrence is the nasopharynx (nasopharyngeal carcinoma [NPC]), where it is almost invariably associated with Epstein-Barr virus (EBV) infection. NPC is an endemic disease prevalent in southeastern coastal provinces of China. Isolated nonnasopharyngeal LEC occurrence is rare, and its association with EBV remains controversial.

Primary temporal bone LEC is a rare disease entity.1 To the best of our knowledge, only 4 patients have been reported in the literature to date, all with confirmed EBV infection.2 In this report, we present the diagnosis and management of a rare LEC case that originated from the middle ear. Its association with EBV is also addressed.

A 56-year-old otherwise healthy woman (a native of Guangdong Province, China) presented to our clinic with aural fullness and hearing loss on her right ear for 1 year. Otoscopic examination revealed a lobulated mass extending from the middle ear space (Figure 1). There was no history of otorrhea. Her brother had had cured NPC for several years. The pure-tone audiometry (PTA; 45 dB) revealed a mixed-type hearing loss with a 30-dB air-bone gap on the right side. The left-side PTA was 18 dB, showing a high-tone sensorineural loss with no conductive component. High-resolution temporal bone computed tomography showed a middle ear mass with no bone or ossicle erosion.

Exploratory tympanotomy revealed that the multilobulated lesion originated from the promontory wall, filling the whole mesotympanum and partially extending into the Eustachian tube. The tumor was removed piece by piece without disrupting the ossicular chain. The patient received adjuvant radiotherapy (62 Gy, 31 fractions) over a period of 7 weeks.

Histopathological examination demonstrated irregular sheets of undifferentiated carcinoma intermingled with abundant lymphoid infiltrate (Figure 2). Immunohistochemical staining was positive for keratin and negative for leukocyte common antigen. The diagnosis of LEC was confirmed. In situ hybridization for EBV-encoded RNA was strongly positive. However, serum

Figure 1. Otoscopic examination revealed a lobulated mass extending from the middle ear space.

Keywords
middle ear, lymphoepithelial carcinoma, Epstein-Barr virus, nasopharyngeal carcinoma

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1Department of Otolaryngology, Head and Neck Surgery, Cathay General Hospital, Taipei, Taiwan
2School of Medicine, Fu Jen Catholic University, Taipei, Taiwan
3School of Public Health, China Medical University, Taichung, Taiwan
4Department of Pathology, Cathay General Hospital, Taipei, Taiwan

Corresponding Author:
Shih-Hung Huang, Department of Pathology, Cathay General Hospital, 280 Sec.4 Jen-Ai Rd, 106 Taipei, Taiwan
Email: drshhuang@gmail.com
immunoglobulin A (IgA) antibodies against EBV capsid antigen (EBV VCA IgA) were negative. Nasopharynx biopsy revealed no evidence of LEC. Magnetic resonance imaging excluded extratemporal invasion and distant metastasis.

Discussion
Tumor occurrences at the temporal bone region are not commonly seen; most are squamous cell carcinoma. Primary LEC of the middle ear is a rare entity. Nonnasopharyngeal LEC has been previously reported under various names, including undifferentiated carcinoma of nasopharyngeal type, undifferentiated carcinoma with lymphoid stroma, lymphoepithelioma, and lymphoepithelioma-like carcinoma. The profound infiltration of lymphoid cells in the stroma makes it difficult to indentify the malignant epithelial cells with routine hematoxylin and eosin staining.

The association between EBV and nasopharyngeal LEC has been well established. Serum EBV titer check is a routine NPC screening test in the endemic area of China. However, the role of EBV in the pathogenesis of nonnasopharyngeal LEC remains unclear. Even though the serum EBV titer was negative, we were able to demonstrate EBV infection in the present case using highly sensitive polymerase chain reaction amplification technique. The presence of EBV-encoded RNA in the middle ear suggests that EBV infection may indeed play some role in the development of middle ear LEC.

Owing to the paucity of middle ear LEC, the optimal therapy regimen remains unknown. Nonnasopharyngeal LEC of the head and neck is radiosensitive with high regional control rates. From the published data, surgery and postoperative radiotherapy are considered the appropriate protocol for middle ear LEC.

In this report, we present the clinicopathologic manifestations of middle ear LEC and its association with EBV. Further study is warranted to establish the appropriate radiation field and dose for the treatment of middle ear LEC.

The preparation of this report is in compliance with the Institutional Review Board regulations in our hospital.

Author Contributions
Leh-Kiong Huon, principal manuscript drafting, patient care; Pa-Chun Wang, patient care, manuscript editing, and finalization; Shih-Hung Huang, histopathological confirmation of diagnosis.

Disclosures
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Figure 2. Hematoxylin and eosin staining demonstrated irregular sheets of undifferentiated carcinoma intermingled with abundant lymphoid infiltrate.