Lymphoepithelial Carcinoma of the Parotid Gland: A Case Report and a Sort Review

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Abstract

Background: We report a case of a 76-year-old woman, presented with a primary lymphoepithelial carcinoma of the parotid gland. Lymphoepithelial carcinoma (LEC) is a rare variant of undifferentiated carcinoma that accounts for less than 0.4% of all salivary gland malignancies. There is strong association with Epstein-Barr virus (EBV) infection. These tumors are histologically indistinguishable from metastatic undifferentiated nasopharyngeal carcinoma and meticulous examination of the pharynx is necessary to exclude metastatic disease and establish diagnosis. Primary surgical excision followed by adjuvant radiotherapy is the most widely accepted therapeutic approach; however, controversies still exist in the current literature regarding the proper management.

Conclusion: Lymphoepithelial carcinoma is a rare tumor and beyond the differential diagnosis further studies and multiclinical trials seem necessary to effectively guide treatment decisions.

Keywords: Carcinoma; Epstein-Barr virus; Lymphoepithelial; Parotid Gland

Introduction

Parotid gland tumors represent approximately 3% of all head and neck neoplasms. Mucoepidermoid carcinoma is the most often encountered neoplasm of the parotid gland with 40% incidence [1].

LEC is an uncommon malignant neoplasm consisted of undifferentiated malignant epithelial cells into lymphoid stroma cells. Schmincke and Regou were independent describers a nasopharyngeal neoplasm with the above mentioned histologic features [2-4]. Later in 1962, Hildermann used the term lymphoepithelial carcinoma (LEC) to describe lesions with these histologic characteristics [5]. Tumors with the same histologic features have been described at various other sites such as tonsil, lung, breast, uterus and other organs [2].

LEC is a subtype of undifferentiated carcinoma with histologic features of nasopharyngeal carcinoma and benign lymphoepithelial lesion. In our paper we describe a new case of LEC of the parotid gland with clinical information on treatment and outcome [6].

Case Presentation

A 76-year-old woman, presented with a 12-month history of a relative firm, gradually enlarging left-sided parotid mass. Her medical and social history was non-contributory. The patient denied pain and the facial nerve was intact (House-Brackmann grade I). A contrast-enhanced magnetic resonance imaging (MRI) revealed an ill-defined lesion in the superficial parotid lobe with no signs of cervical lymphadenopathy. Fine-needle aspiration (FNA) biopsy results indicated a poorly differentiated neoplasm of epithelial origin. The patient received a superficial left parotidectomy with facial nerve preservation (Figure 1A-1C). Histopathologic analysis revealed non-neoplastic lymphocytic infiltration surrounding nests of undifferentiated cells of epithelial origin (Figure 2A, 2B), that stained positive for cytokeratin stain (CK 5/7) and Epstein-Barr encoded RNA (EBER); features compatible with lymphoepithelial carcinoma (LEC). A meticulous endoscopic examination of the nasopharynx and oropharynx was performed to rule out metastatic undifferentiated nasopharyngeal carcinoma and confirm diagnosis of primary parotid gland LEC. Postoperatively, the patient received adjuvant radiotherapy (5400 cGy) to the gland area, according to the institutional
multidisciplinary head and neck tumor board recommendations. During a 36-month follow-up period no evidence of recurrence did reported.

**Discussion**

Lymphoepithelial carcinoma is a rare variant of anaplastic carcinoma, that accounts for less than 0.4% of all salivary gland malignancies and predominantly encounters in major salivary glands (83.5% in the parotid and 15% in the submandibular gland) [7]. There is marked predilection for Greenland Eskimos and Asians (southeastern Chinese, and Japanese) and a strong association with Epstein-Barr virus (EBV) infection [8]. EBV genome is detected by hybridization, in the majority of LEC cases, indicating its potential role in tumorigenesis [9].

LEC of the parotid is usually presented as a painless, rapidly increasing mass, with significant heterogeneous or homogeneous enhancement in CT or MRI scans [10]. Facial nerve is rarely involved, while more than 40% of patients present with cervical dissemination [11]. Histopathologically, it is characterized by poorly differentiated tumor cells of epithelioid origin separated by a mixture of B- and T-lymphocytes (lymphoid stroma). Tumor cells are typically stained positive for cytokeratin and epithelial membrane antigen. A “starry-sky” appearance associated with histiocytes may be demonstrated in some cases [9].

Differential diagnosis includes metastatic undifferentiated nasopharyngeal carcinoma, lymphoma, lymphadenoma, lymphoepithelial sialadenitis and large cell carcinoma [9,11]. As LEC is hardly distinguishable from metastatic undifferentiated nasopharyngeal carcinoma, a meticulous endoscopic examination of the nasopharynx and oropharynx seems necessary to exclude metastatic disease and establish diagnosis [12].

Superficial or total (for deep lobe tumors) parotidectomy with facial nerve preservation is the mainstay of therapeutic approach, based on the literature review [11,12]. Postoperative adjuvant radiotherapy is widely advocated to maximize oncologic control [11]. Neck dissection is generally reserved for neck positive cases [11]. Given its radiosensitive behavior, controversies still exist regarding proper management with Dubey et al. [13] from MD Anderson Cancer Center (Houston) considering primary radiotherapy as a reliable alternative. Chemotherapy has failed to improve survival and it is not routinely performed. Compared with the other types of salivary undifferentiated carcinomas, LEC has a significant better prognosis with the estimated 5-year overall survival to reach up to 90% [11].

Due to its rarity, lymphoepithelial carcinoma is not an adequately-studied malignancy and further studies and multi-clinical trials seem necessary to ascertain proper management and effectively guide decision making.

**References**


