CASE REPORT

# Lymphoepithelial Carcinoma of Parotid Gland- A Case Report

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**Abstract:**

Lymphoepithelial carcinoma (LEC) is most commonly seen in the nasopharynx. Very rarely it is found in the salivary gland, preferably in parotid gland followed by submandibular gland where it accounts for 0.4% of all malignant salivary gland tumours. Most commonly it is seen in fifth decade with female predominance. Significant correlation has been reported between this tumour and the Epstein Barr virus (EBV).It has a racial predilection for Inuits,Chinese and Japanese. Very rarely it is found In Indians. So we present a case of LEC of parotid gland in a 23 year old male Indian patient. As this is a very radiosensitive tumour, surgery followed by radiotherapy remains the treatment of choice.

**Keywords:** Carcinoma, Lymphoepithelial Carcinoma, Parotid Gland, Salivary Gland.

## Introduction

Lymphoepithelial carcinoma is a specific subtype of undifferentiated carcinoma with characteristic dense lymphoid stroma. The most frequent location is the nasopharynx. Identical tumours have been rarely described in the major salivary glands where they account for approximately 0.4% of all malignant salivary gland tumours [1]. The parotid gland is affected in approximately 80% of the cases, followed by the submandibular gland and minor salivary glands [2]. Very rarely they are found in lungs, thymus, stomach, larynx, trachea and skin [3]. Majority of LECs are seen in fifth decade with female predominance [4]. It shows a striking racial predilection for Inuits (Eskimo) in the Arctic regions (Greenland, Canada, Alaska), South-eastern Chinese, and Japanese [2]. In Indians it is very rarely found. Significant correlation has been reported between this tumour and the Epstein Barr virus (EBV) [5]. Due to a high radiosensitivity for LEC, surgery combined with Radio Therapy (RT) is considered as the first choice for treatment of this disease [6]. Very few cases have been reported in medical literature till now. We present a case of LEC of parotid gland in a 23 year old male Indian patient.

## Case Report:

A 23 year old male patient presented with painful swelling of 4x3 cm in left preauricular region. The swelling was hard in consistency, tender, having irregular borders and bosselated surface. His facial nerve functions were intact. All finding in general examination and laboratory investigations were normal. CT scan revealed a mass in left parotid gland. We had performed whole body CT Scan to rule out metastasis in other organs of the body and on that we didn't get any areas of metastasis. Patient was treated with removal of mass by a surgical procedure of superficial parotidectomy. We received superficial parotidectomy specimen measuring 5x4x2.5 cm. On cut section a gray white tumour measuring 4x3x2.5 cm was seen along with areas of hemorrhage and necrosis. Histopathological examination showed salivary gland with tumour composed of tumour cells growing in nests and trabecular sheets intermingled with lymphoid stroma (Fig.1)

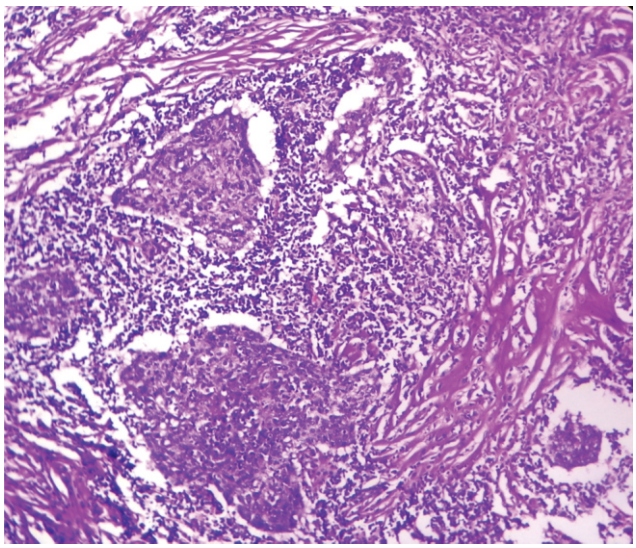

**Fig. 1: Tumour Cells Growing in Nests Intermingled with Lymphoid Stroma(H &E,10X)**

Neoplastic cells exhibited syncytial appearance with indistinct cell border, eosinophilic cytoplasm, large vesicular nuclei and prominent nucleoli (Fig. 2).

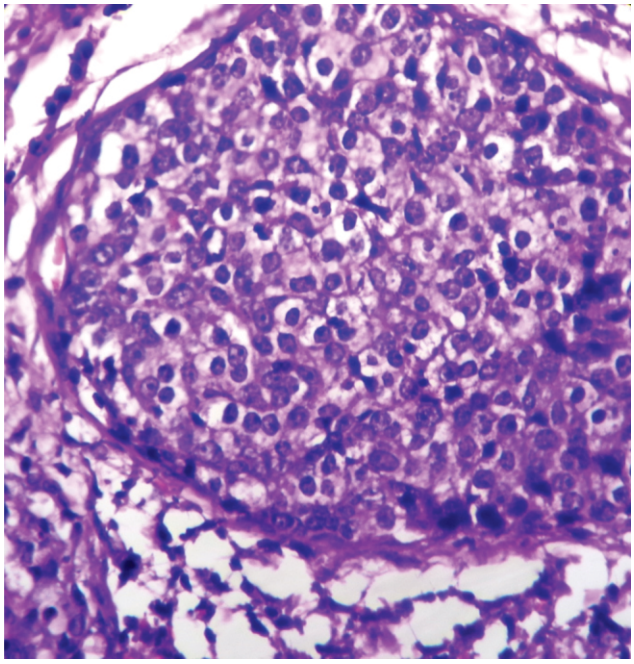

**Fig. 2: Neoplastic Cells Exhibited Syncytial Appearance with Indistinct Cell Border, Eosinophilic Cytoplasm, Large Vesicular Nuclei and Prominent Nucleoli (H&E, 40X)**

Few mitosis and areas of hemorrhage and necrosis were also seen. In immunohistochemistry epithelial cells and lymphocytes showed positivity for cytokeratin and leukocyte common antigen respectively. So the diagnosis of LEC of parotid gland was given. We didn't get the patient for follow up to opine about the outcome of the treatment.

**Discussion:**

According to the classification by Ellis and Auclair, undifferentiated carcinomas of the salivary glands can be subtyped further into small cell undifferentiated carcinoma, large cell undifferentiated carcinoma, and LEC [7]. LEC accounts for approximately 0.4% of malignant salivary gland tumours [1]. This tumour affects the parotid gland in approximately 80% of the cases [2]. Most cases occur in the fifth decade of life with age range of first to ninth decade. It is most commonly seen in females [4]. There are few reports showing its occurrence in males as seen in our case [8]. Most of parotid LEC cases arises *de novo*, but they may rarely develop within lymphoepithelial sialadenitis [9]. The exact origin and pathogenesis of parotid LEC remain unknown. Among the possible etiologies are a malignant transformation of the glandular and ductal inclusions in the intra-parotid lymph nodes and a malignant transformation of the epi-myoepithelial island [3, 10]. It seems there is a complex interaction between genetic factors, environmental factors, and EBV infection in the oncogenic process of LEC of the salivary glands [2]. EBV may be directly shed from lymphocytes into the salivary gland or may replicate in epithelial cells. The viral episome is maintained in the infected epithelial cell that continues to proliferate and does not differentiate. This focus of latent infected, metaplastic epithelial cells rapidly become malignant and invade the basement membrane.

Parotid LEC usually presents as an enlarging parotid lump, occasionally painful and with facial nerve involvement in approximately 20% of cases [4]. Distant metastases usually involve the lung, liver, bone, and brain [1]. Macroscopically, these tumours are firm, 1–10 cm masses, multinodular, circumscribed, or clearly infiltrative into adjacent salivary gland, fat, muscle, or skin, with a cut surface that varies from a grey-tan to yellow-gray. Histologically, it is characterized by a syncitial growth pattern and a dense stroma made of non-neoplastic lymphoplasmacytic cells; the lymphoid cells include a mixture of B and T cells and are sometimes associated with germinal centers. The epithelial component is composed of irregular shaped islands, cords, trabeculae of pleomorphic, large, malignant cells with abundant lightly eosinophilic cytoplasm and vesicular nuclei. Mitotic rate is variable. LEC is indistinguishable from undifferentiated nasopharyngeal carcinoma which is much more common or other LECs that develop in various parts of the body [4].

Therefore, to confirm the diagnosis of primary LEC in the major salivary glands, metastatic nasopharyngeal carcinoma to the salivary glands should be eliminated through examination of the upper aerodigestive tract with endoscopy, CT scan and even random biopsy of the nasopharynx [4]. Nasopharynx was absolutely normal on CT scan in our case.

In reality, the parotid gland is the predominant site of occurrence of LEC and an exceptional site of metastasis from nasopharyngeal carcinoma, which more typically metastasizes to the cervical or sub-mandibular lymph nodes [1]. Immunohistochemistry shows neoplastic cells that stain positive for cytokeratin and epithelial membrane antigen [9]. Lymphoid cells are reactive for leukocyte common antigen, CD20 and CD3 markers. Histologically, the LEC must be distinguished from benign lesions such as lymphoepithelial sialadenitis and from other malignancies, such as primary or metastatic poorly differentiated squamous cell carcinoma, adenocarcinoma, amelanotic melanoma and large cell or anaplastic types of lymphoma [7]. Lymphoepithelial carcinoma seems to have a better prognosis than the other undifferentiated carcinomas of the salivary glands, in part because of the lymphoid stroma that has a role in limiting the aggressiveness of this carcinoma. Advanced disease, the presence of metastases on diagnosis, and histological features such as high mitotic rate, anaplasia, and necrosis are predictors of a worse prognosis [1, 2]. However, the presence or absence of EBV in LEC does not appear to be of prognostic significance [2, 7]. Because most LECs of the parotid gland are radiosensitive, the optimal management is complete excision with clear surgical margins followed by adjuvant radiotherapy to the tumour bed and neck. In conclusion, LEC of parotid gland is a very rare tumour to found in Indian population that requires surgical excision and postoperative radiotherapy.

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