Primary Lymphoepithelial carcinoma of the parotid gland - A case report

BHONSLE VAISHALI SHARAD SHARADSHIVAJIROBHONSLE
Department of Pathology,
CHRISTIAN MEDICAL COLLEGE

Abstract:
We present a case of a 54 year old female with lymphoepithelial carcinoma of the parotid gland. Primary lymphoepithelial carcinoma is a rare tumour of the salivary glands and metastatic deposits from a nasopharyngeal tract primary need to be excluded before this diagnosis is made.

Keyword: Lymphoepithelial carcinoma, Parotid gland

Figure 1 – Tumour nodules with salivary gland tissue seen at the periphery (star) (H&E X 5)
Figure 2 – Tumour cells with ill defined cell borders, vesicular nuclei and prominent nucleoli with closely admixed lymphoplasmacytic infiltrate (H&E X 40).

Figure 3 – Positive immunostaining with pancytokeratin; Pancytokeratin (X 20).

Introduction
Lymphoepithelial carcinoma (LEC) is an undifferentiated carcinoma with a prominent non neoplastic lymphoplasmacytic infiltrate. It accounts for less than 1% of all salivary gland tumors. It is known to be associated with EBV infection in endemic areas. (1)

Case report:
History, physical examination and imaging findings:
54 year old lady presented with history of progressive swelling of the left parotid region for 2 years, which rapidly increased in size over the past 6 months and associated with mild dull aching pain in the parotid region with decreased hearing in the left ear. She was a known diabetic on oral hypoglycaemic drugs. There were no other ENT complaints. On examination there was a 2.5 x 2.5 cm oval, firm swelling smooth surface in the left parotid area. It was nontender and immobile. The skin overlying the swelling was normal. There was no cervical adenopathy. ENT examination by indirect laryngoscopy was normal. Facial nerve and sensation over the parotid and left pinna was normal. USG screening showed an ill defined hypoechogenic lesion in the left parotid. MRI revealed STIR hyperintense area in the superficial lobe of the left parotid gland with contrast enhancement on the post contrast study with associated multiple intra-parotid nodes. FNAC correlation was suggested. FNA was reported as carcinoma, and biopsy was suggested for definite characterisation of the lesion. Left total conservative parotidectomy with selective neck dissection was done. Intraoperatively there was a large firm to hard mass involving the superficial and deep lobes of the parotid gland and also infiltrating the cartilagenous portion of the external auditory canal and adjacent soft tissue. The upper division of facial nerve was encased by the tumour. There were multiple level II and level III lymph nodes.

Pathology
Gross – The parotidectomy specimen weighed 20gms and measured 5x4x2cm. The cut surface was lobulated firm grey white cut surface. Microscopy revealed salivary gland tissue infiltrated by a tumour composed of syncitial sheets, islands and cords displaying polygonal cells with round to oval, moderately pleomorphic, vesicular nuclei with prominent nucleoli and moderate amounts of eosinophilic cytoplasm with ill defined cell borders. There was increased mitotic activity including atypical mitotic figures. Tumour nests were densely infiltrated by lymphoid cells rich stroma composed of lymphocytes, plasma cells, histiocytes and many reactive lymphoid follicles with germinal centre formation. Tumour reached up to inked resection margin. Perineural invasion was evident. Lymphovascular invasion was not seen. Tumour cells were immunopositive for Pan-CK. EBV LMP-1 staining was non-contributory. A diagnosis of primary lymphoepithelial carcinoma of the parotid gland was made.

Discussion
Lymphoepithelial carcinoma (LEC) is an undifferentiated carcinoma with a prominent non neoplastic lymphoplasmacytic infiltrate. Schmincke first described lymphoepithelial carcinoma in the nasopharynx in the early 1920’s. LEC accounts for less than 1% of all salivary gland tumors and 0.4% of malignant
salivary gland tumours. (2) It is known to be associated with EBV infection in endemic areas. (1) Among the salivary glands the parotid gland is the site of origin in 80% of cases followed by the submandibular gland. LEC of the salivary glands and nasopharyngeal carcinoma are morphologically indistinguishable from each other. Hence it is necessary to thoroughly examine the nasopharynx before accepting a diagnosis of primary LEC of the salivary gland. (3) Precursor lesion - Most cases arise de novo but lymphoepithelial sialadenitis may precede the development of LEC in some cases. (4)

Grossly these tumours may be circumscribed or show infiltration. The size ranges from 1-10cm. The consistency is fleshy to firm. (5) There is a tendency to spread to cervical lymph nodes. Distant metastasis occurs to the lungs, liver, bone and brain. The dense lymphoplasmacytic infiltrate may or may not be present in the metastatic deposits.(6) Histology reveals infiltrative sheets, islands and cords of neoplastic cells with indistinct cell borders, pale eosinophilic cytoplasm, oval vesicular nuclei with open chromatin, and conspicuous nucleoli. The nuclei are usually moderately pleomorphic but may also be uniform appearing in a few cases. The tumour cells may also be spindly and form fascicles.(6) There may be areas of squamous differentiation. Mitotic activity is high. Necrosis is usually present. The tumour is richly infiltrated by lymphocytes and plasma cells with frequent formation of reactive lymphoid follicles. The lymphoid cells include a mixture of B cells and T cells. Abundant histiocytes in some cases impart a “starry sky” appearance. (7) Other inconsistent findings include non-caseating granulomas with or without multinucleated giant cells, amyloid deposition, cyst formation, perineural and lymphovascular invasion. (8)

In endemic cases, EBV-encoded RNA (EBER) and EBV-DNA can be detected in the tumour cells by in-situ hybridization. Immunohistochemical expression of EBV latent membrane protein 1 is more variable.(9) The differential diagnosis includes metastatic undifferentiated carcinoma, malignant lymphoma, lymphoepithelial sialadenitis and large cell undifferentiated carcinoma. (6) The prognosis is related to tumour stage and grade. Five-year survival rate of 75-86% has been reported in patients treated by combined surgery (including neck dissection) and radiation therapy, although local recurrence can occur. (10)

**Conclusion**

LEC of the parotid gland is a rare tumour and since it is morphologically indistinguishable from nasopharyngeal carcinoma which is a commoner tumour it is essential to rule out this entity before making a diagnosis of primary LEC of the parotid gland. In our case a thorough examination of the upper aerodigestive tract excluded a primary from these sites.


