

# A rare case of lymphoepithelial carcinoma of the parotid gland

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## ABSTRACT

Lymphoepithelial carcinoma (LEC) of salivary gland is uncommon and accounts for 0.4% of malignant salivary gland tumours. Diagnosis can be difficult. We report case describes a 43-year-old indigenous gentleman who presented with a year history of left parotid swelling. Fine needle aspiration was inconclusive and computed tomography of neck was consistent with a benign parotid lesion. The patient proceeded with surgery and histopathology revealed LEC. Adjuvant radiotherapy was given and no recurrence was detected during his two years follow up. Our case highlights that diagnosis of LEC can be difficult and surgical resection or tissue sampling other than fine needle aspiration may need to be considered to make a diagnosis.

**Keywords:** Lymphoepithelial carcinoma, parotid gland, head and neck cancer

## INTRODUCTION

Lymphoepithelial carcinoma (LEC) of salivary gland is uncommon. LEC accounts for 0.4% of malignant salivary gland tumours.<sup>1</sup> In about 80% of the salivary gland LEC, the parotid gland are involved.<sup>2</sup> We would like to report a case of parotid LEC which was treated in our centre. Looking at the rarity of primary salivary gland LEC, from our literature review it is crucial to rule out possible primary from the nasopharynx.

## CASE REPORT

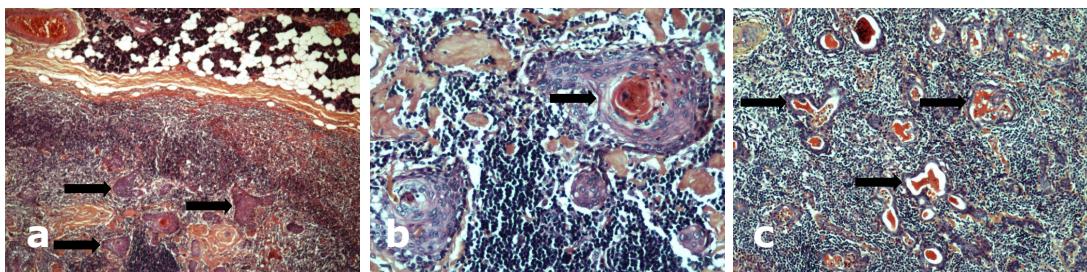
A 43-year-old indigenous man (Iban) was referred to Department of Otorhinolaryngology, Head and Neck Surgery for a gradually enlarging painless left parotid swelling over duration of a year. His medical and surgical histories were unremarkable. He was a non-smoker. He had no family history of malignancy. Upon physical examination, there was

an infra-auricular mass measuring 5 cm x 3 cm, cystic in consistency, mobile and non tender. Facial nerve function was not impaired. There were no palpable cervical lymph nodes. Other systemic examinations were unremarkable.

A fine needle aspiration cytology (FNAC) was performed. Unfortunately, this was not conclusive. Computer tomography (CT) of neck showed a left parotid solid-cystic lesion seen within the superficial lobe with no significant cervical lymphadenopathy. Patient then underwent left superficial parotidectomy with preservation of the facial nerve anatomically and physiologically. Intraoperative findings were left parotid tumour involving superficial lobe which was cystic in nature and well encapsulated. There was no macroscopic invasion of the facial nerve by the tumour.

Histopathology of the surgical specimen revealed a fairly circumscribed parotid tumour composed of sheets, islands and

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**Figs. 1:** Lymphoepithelial carcinoma a) A well encapsulated tumour containing sheets, cords and trabeculae of malignant cells surrounded by abundant lymphoid stroma. Note normal parotid gland acini at the periphery (H&E x 100), b) Squamous morules surrounded by lymphoid cells seen in LEC with keratin pearl formation at the centre (H&E x 200) and c) Foci of glandular differentiation in LEC (H&E x 100).

cords of malignant cells surrounded by abundant lymphoid stroma (Figures 1). The malignant cells possessed uniform oval nuclei with abundant cytoplasm and showed squamous differentiation. Focal areas showed tubular differentiation. Abundant histiocytes, lymphocytes, plasma cells and eosinophils were seen. Mitotic figures were occasionally seen. Focal area showing a cystic space containing debris was noted. There was no perineural or vascular invasion seen. There was no invasion into the surrounding tissue seen. A diagnosis of lymphoepithelial carcinoma was made with the lesion completely excised.

In view of the rare diagnosis of primary LEC of parotid gland, nasoendoscopy was performed to rule out possibility of metastases from nasopharyngeal carcinoma which was insignificant. Chest radiography and ultrasound abdomen done showed no distant metastases. His Epstein Barr Virus Viral Capsid Antigen IgG was reactive.

Patient was referred for radiotherapy six weeks post operation. He refused radiotherapy initially as he had some unsettled family issues. However upon settling his family issues which was eight months after operation, he agreed for commencement of radiotherapy. A repeat CT scan before commencing radiotherapy showed no evidence of recurrence. A total of 60 Gy in 30 fractions

were successfully delivered to the primary site and neck without any long term complication. Surveillance CT neck done one year post completion of radiotherapy had no evidence of recurrence. At two years follow-up, the patient remained disease free.

## DISCUSSION

The term 'Lymphoepithelial Carcinoma' (LEC) was first introduced by Hildermann in 1962.<sup>3</sup> The World Health Organisation (WHO) defines LEC as undifferentiated carcinoma with lymphoid stroma.<sup>4</sup> Primary LEC can also occur in various other sites such as the tonsils, thymus, lung, breast, uterus, stomach, urinary tract, bladder and skin.

LEC is nearly 100% associated with EBV in endemic areas.<sup>5</sup> Based on the striking racial prevalence of this condition in Inuits in the Arctic region, south-eastern Chinese and Japanese, most published studies concluded that the association of salivary gland LEC and EBV is strongly related to racial and geographical factors.<sup>6</sup> In our case, the EBV serology IgG was reactive which indicated previous EBV infection.

Parotid LEC cannot be distinguished from nasopharyngeal carcinoma through its morphology.<sup>2</sup> Therefore, it is important to perform nasoendoscopy to rule out a primary nasopharyngeal lesion. In our case, nasoen-

doscopy was done only after histopathological diagnosis of malignancy. Preoperative CT neck had not shown any mass over the Fossa of Rosenmuller.

Parotid LEC usually presents as an enlarging parotid lump and in approximately 20% of cases, there will be pain and facial nerve involvement.<sup>7</sup> Subtle presentation with absence of typical symptoms of malignancy makes diagnosis difficult resulting in often delayed diagnosis. Involvement of cervical lymph node during presentation has been reported, ranging from 10 to 40% of the cases.<sup>8</sup> In 20% of the reported cases, distant metastasis is documented at presentation.<sup>7</sup> The most common sites for distant metastasis are the lung, liver, bone and brain. In our patient, there were no evidence of nodal or distant metastases.

The main aim of performing FNA for parotid lesion is to differentiate between benign and malignant condition. In our case, FNA was inconclusive. Studies have reported the accuracy of FNA to range from 69% to 96%, the sensitivity of FNA to detect malignancy from 57% to 88%; the specificity of FNA to diagnose malignant lesion ranging from 86% to 100%.<sup>9</sup> A study has shown that preoperative FNA diagnosis of malignancy improves surgical treatment and outcome parameters of parotid cancer and should be strongly considered.<sup>9</sup> However, the gold standard to diagnose LEC lies with histopathological analysis of the resected tumour.

Due to the rarity of primary salivary gland LEC, there are limited cases to study about the optimal treatment for the disease. Since LEC is radiosensitive, a combination of surgery with complete removal of the tumour with wide margin and post operative radiotherapy to the parotid bed is recommended.<sup>7</sup> Involvement of neck nodes requires neck dissection followed by radiotherapy to neck region. In our case, left superficial parotidectomy

with complete excision of the tumour was done and proceeded with adjuvant radiotherapy.

Myers *et al.* reported estimated 2-, 5- and 10 year survival for LEC of salivary gland at 91%, 66% and 29% respectively.<sup>10</sup> The 5-year survival rate has also been reported to range from 50 to 87%.<sup>11</sup> In our patient, the prognosis is good in view of absence of distant metastases at presentation and adequate treatment subsequently.

In conclusion, histopathology is the gold standard to attain correct diagnosis. Primary salivary gland LEC is rare and metastatic disease from nasopharyngeal lesion needs to be ruled out. Rarity of this condition prevents extensive study on its treatment modality but a combination of surgery and radiotherapy is so far the best treatment regime and again highlighted in our case.

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