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CASE REPORT

Lymphoepithelial Carcinoma of the Submandibular Gland: A Rare Case

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ABSTRACT

Lymphoepithelial Carcinoma (LEC) is a rare malignancy of the salivary gland, which is associated with Epstein-Barr virus (EBV). LEC of the head and neck is commonly seen in the nasopharynx. In Asia, the common head and neck cancer associated with dermatomyositis is nasopharyngeal carcinoma. We report a case of a 46-year-old Chinese lady with underlying dermatomyositis who presented with a left submandibular swelling of two years duration that was associated with numbness of the left hemi-tongue. The left submandibular swelling was firm, non-tender, mobile, and measured 2 x 2cm in size. Computed tomography (CT) showed a heterogeneous enhancement of the left submandibular gland with surrounding inflammatory changes suspicious of malignancy. Preoperative fine needle aspiration revealed reactive lymphadenopathy. Left sub-mandibulectomy was performed, which was subsequently confirmed as LEC. Patient then underwent a modified radical left neck dissection followed by chemoradiotherapy. One-year post-treatment the patient showed no evidence of recurrence. LEC is a rare salivary gland tumour that has been associated with the EBV. Patients with dermatomyositis have a high risk of developing nasopharyngeal carcinoma, particularly, in Southeast Asian populations. This case, highlights that clinicians should also have high index of suspicion of salivary gland malignancy in patients with dermatomyositis, as this is rare and not widely seen and reported.

KEYWORDS: dermatomyositis, lymphoepithelial carcinoma, Epstein-Barr virus, submandibular gland

INTRODUCTION

Lymphoepithelial carcinoma (LEC) mainly occurs in the nasopharynx. However, it can also be found in the salivary glands, mainly in the parotid glands followed by submandibular glands [1]. LEC is a relatively uncommon entity when compared to the prevalence of mucoepidermoid carcinoma and adenoid cystic carcinoma of the salivary gland [1]. Majority of the patients with LEC of the salivary gland present with a painless parotid or submandibular swelling, cervical lymphadenopathy, and in some instances facial nerve palsy [2]. It is more common in females than males, with an average age of occurrence of 40 years [1]. Surgical excision is the main stay of treatment followed by radiotherapy. Dermatomyositis, on the other hand, is an idiopathic inflammatory myopathy of the striated muscle with presence of distinctive skin rash [3,4]. Patients with dermatomyositis have a six-fold risk of developing cancer, depending on the origin, age and gender [4]. It is said that 10-20% of patients have increased risk of developing carcinoma at the time of diagnosis. Here we report a case of lymphoepithelial carcinoma of the submandibular gland with an uncommon presentation.

CASE PRESENTATION

A 46-year-old Chinese woman presented to us with a history of painless left submandibular swelling over the past two years, associated with numbness over the left side of the tongue for one year. However, the left submandibular swelling became painful to touch about two months prior to her presentation. Otherwise, she

had no other symptoms. She has underlying dermatomyositis and has been on treatment for about one year. She also has type II diabetes mellitus, hypertension and multinodular goitre. Physical examination showed a 2 x 2cm left submandibular mass, which was firm, non-tender, and mobile with no skin changes. The swelling was not attached to the underlying structures. There was also an anterior neck swelling measuring 5 x 4 cm that moved during deglutition, corresponding to the underlying multinodular goitre. Otherwise, there were no palpable cervical lymph nodes. Examination of the oral cavity showed normal appearance of the tongue and other subsets of the oral cavity. However, upon protrusion, there was deviation of the tongue to the left. A full cranial nerve examination showed isolated left hypoglossal nerve involvement. Naso-endoscopic examination was unremarkable.

A pre-operative fine needle aspiration cytology of the left submandibular mass reported reactive lymphadenopathy. Contrast Enhanced Computed Tomography (Figures 1 & 2) showed a diffusely enlarged left submandibular gland (3.2 x 2.2 x 3.6 cm) with heterogenous enhancement, abutting the left mylohyoid, anterior belly of left digastric and left platysma muscles, with no gross infiltration. There were also a few associated enlarged left submandibular nodes, the largest measuring 1.7 x 1.2 cm. Left sub-mandibulectomy was then performed. The histopathology report showed LEC of the submandibular gland. A post-operative positron emission tomography (PET) scan showed multiple sub-centimetre lymph nodes over the left level II and level III region, with no fluoro-deoxyglucose uptake at fossa of rosenmuller. After a multidisciplinary team discussion together with the oncology team, the patient underwent modified radical left neck dissection, which showed no malignancy in all the lymph nodes excised. Nevertheless, the patient was given concurrent chemoradiotherapy, which consisted of five cycles of chemotherapy and thirty-five fractions of radiotherapy. One-year post treatment evaluation of the patient showed no evidence of recurrence.

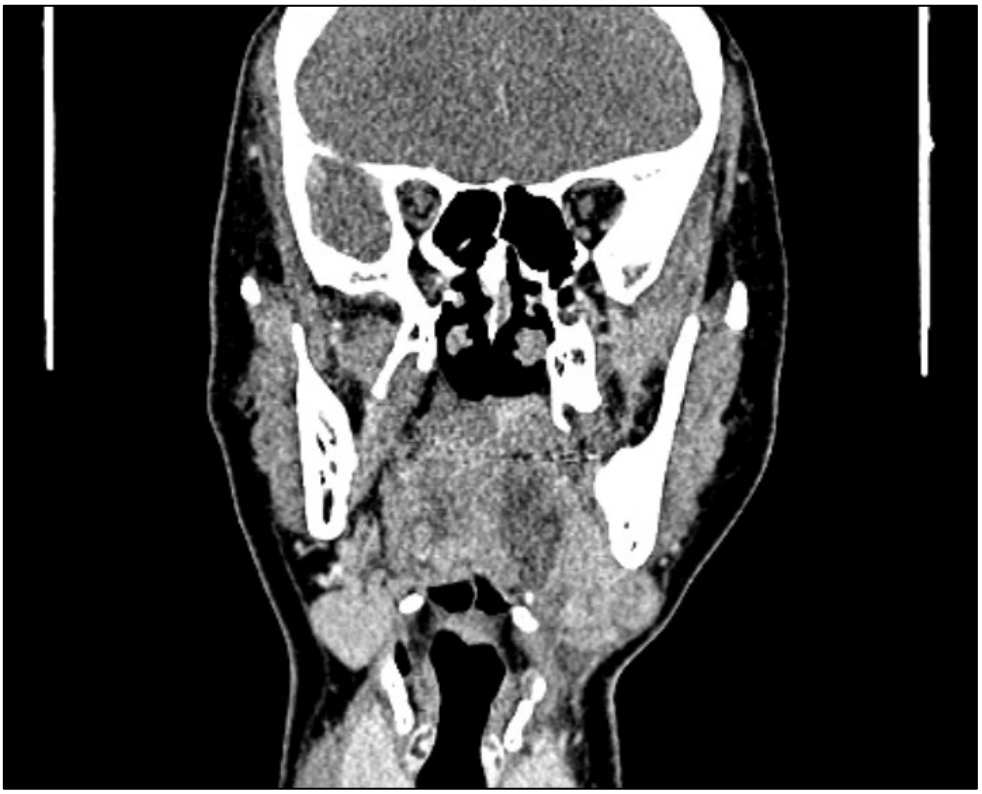

Figure 1 Coronal view shows a diffusely enlarged left submandibular gland with heterogenous enhancement, abutting left mylohyoid (green dot), anterior belly of left digastric (yellow dot) and left platysma muscle (blue dot)

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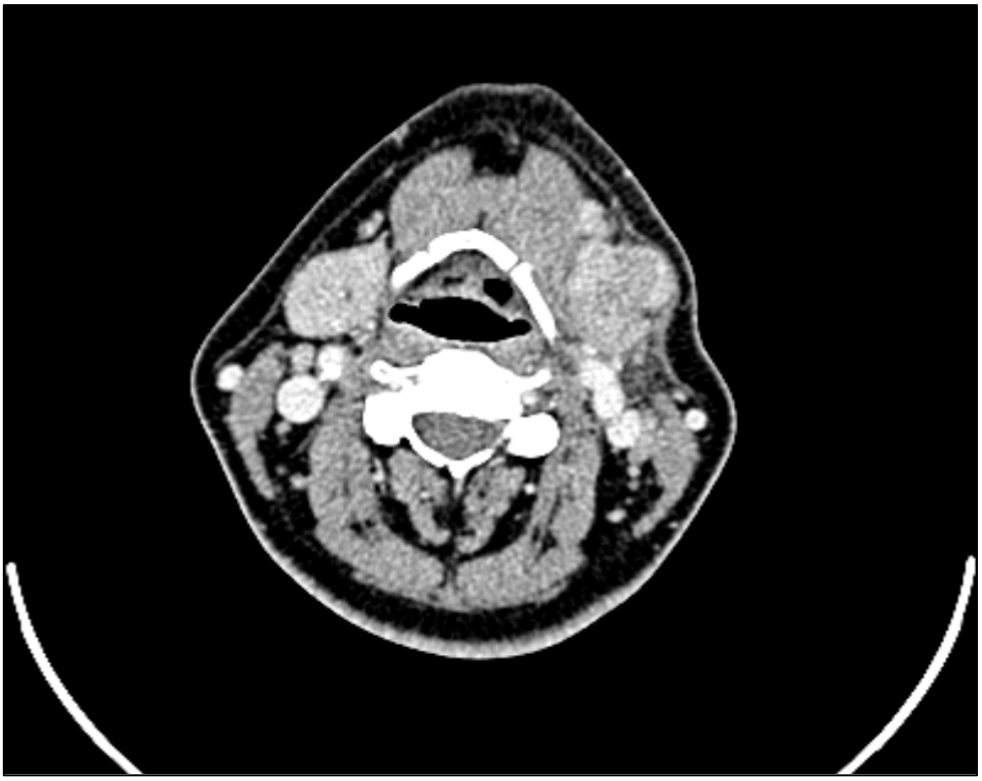

Figure 2 Axial view shows a diffusely enlarged left submandibular gland with heterogenous enhancement and enlarged multiple submandibular nodes (yellow circle)

DISCUSSION

Lymphoepithelial carcinoma (LEC) is commonly found in the nasopharynx [2], and LEC of the salivary gland is rare, accounting for less than 0.4% of all salivary gland malignancies, especially of the parotid gland [1,2]. Histologically, this tumour is almost identical to metastatic undifferentiated nasopharyngeal carcinoma [2,4,5]. This rare entity is common among the Eskimo, Southeast Asian Chinese and Japanese populations [2]. It also has a strong association with EBV [2,3,5,6]. LEC usually presents with unilateral painless swelling, though very occasionally with pain, and is commonly seen between the ages of 30 and 80 years, with a median age of 60 years [6]. Preoperative MRI, CT and ultrasound can be done to differentiate between benign and malignant growths, and to help plan the surgery. MRI will usually show T2 hypodensity and ill-defined margins on a post contrasted image, which is useful for anatomical delineation [2,5]. Tumour excision with post-operative radiotherapy is the treatment of choice as LEC is radiosensitive and has agood outcome [1,5].

Dermatomyositis is a paraneoplastic syndrome and is associated with malignancy. About 20 to 30% of patients with dermatomyositis will develop cancer [7]. Dermatomyositis patients from Western societies may develop cancer of the breast, lung or colon and rectum, whereas, patients from Southeast Asia and Southern China may develop nasopharyngeal carcinoma [4,8]. Many reported cases of dermatomyositis associated with head and neck malignancy were shown to have malignancy in the nasopharynx rather than salivary gland. There is a possibility of interconnection between genetic factors, environmental factors and EBV infection in causing oncogenic process of LEC of salivary gland [9]. Even though LEC of the salivary gland has not been reported to be associated with dermatomyositis, there is a relationship between nasopharyngeal carcinoma and LEC of the salivary gland, which is closely related to EBV infection. Therefore, in a patient with underlying dermatomyositis, there is a risk of developing LEC of the salivary gland as evident from this case. Dermatomyositis, has also been associated with Sjogren Syndrome.

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CONCLUSION

Our patient, who is a Chinese lady in her 40's with underlying dermatomyositis, presented with a left submandibular swelling, which was consistent with LEC. Even though there are no case reports suggesting a correlation between dermatomyositis and LEC of the submandibular gland, there are many case reports showing correlation between dermatomyositis and EBV-positive nasopharyngeal carcinoma. We would like to highlight that in a case of dermatomyositis, when a patient presents with concomitant major salivary gland swelling, it should be further investigated with a view to its excision. This is so that a proper histopathological assessment can be obtained and the patient can be referred to oncology for chemoradiation therapy based on the extend and severity of the disease. This awareness is of utmost importance among dermatologists and otorhinolaryngologists that would enable them to be able to make a diagnosis, as LEC of the salivary gland in dermatomyositis can occur even though it is extremely rare.

Conflict of Interest

Authors declare none.

Authors' contribution

Prempreet Kaur: Drafting and finetuning the manuscript.

Chong Shu Sim: Case selection and editing the manuscript.

Muhammad Hazim Abdul Ghafar: Case selection and editing the manuscript.

Norhafiza Mat Lazim: Case selection and manuscript revision.

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