Case Report

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Pathology Section

A Rare Case of Lymphoepithelial Carcinoma of Salivary Gland in a Patient of Indian Origin

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ABSTRACT

Salivary gland neoplasms comprise 3% of all the head and neck neoplasms. Lymphoepithelial carcinoma of the salivary gland is extremely rare and accounts for less than 1% of all salivary gland neoplasms. It is known to have a racial predilection for Inuits (Eskimos), Orientals and a slight female preponderance. Lymphoepithelial carcinoma is known to be associated with Epstein Barr virus infection.

A report of a lymphoepithelial carcinoma of the parotid salivary gland occurring in a 48 year old female is presented here, in view of the rarity of occurrence in the salivary gland and ethnicity.

Keywords: Lymphoepithelial Undifferentiated Carcinoma, Salivary Gland, Parotid.

INTRODUCTION

Despite their relatively simple morphology, salivary glands give rise to a striking range of histologically distinct tumours. Between 64-80% of all the primary epithelial salivary gland tumours occur in the parotid gland amongst which about 54 to 79% are benign and 21 to 46% malignant. Lymphoepithelial carcinoma (LEC) is an undifferentiated carcinoma which occurs predominantly in the upper aero-digestive tract. It shows a consistent association with Epstein Barr virus infection [1,2]. LEC of the salivary gland is rare and accounts for less than 1% of all the salivary gland tumours and predominantly affects the parotid gland. It is a demographically and histopathologically unique malignancy. It is predominantly a carcinoma of American Eskimos and native Green landers [3] and at one point of time accounted for 25% of all the malignancies in that population [1].

We report a case of LEC in parotid gland in a 48 year old patient, of Indian origin, in view of the rarity of its occurence, with reference to the site and ethnicity.

CASE HISTORY

A 48 year old female patient presented with a painless swelling below the right ear, present since 6 months. The swelling gradually progressed in size during this period to attain a size of 5x4 cm. There was no history suggestive of fever, trauma, tooth extraction or facial weakness.

Physical examination revealed a firm, mobile and non-tender mass below the right ear. The facial nerve function was intact. No enlarged lymph nodes or other masses were palpable. A clinical diagnosis of benign parotid tumour was offered.

Following this, a fine needle aspiration was performed. A cytological diagnosis of Warthin's tumour was given. Right superficial parotidectomy was performed and the specimen was subjected to histopathological examination.

PATHOLOGIC FINDINGS

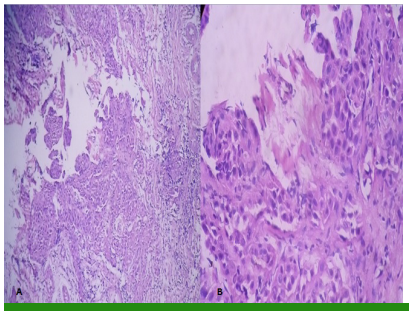
Gross examination: A single, grey-white to grey yellow mass with attached fibro-fatty tissue measuring 4.5 x 3 x 2.5 cm, was received. Cut section showed 3 well encapsulated grey-white nodules surrounded by normal looking areas. The largest nodule measured 1.5 x 1 cm.

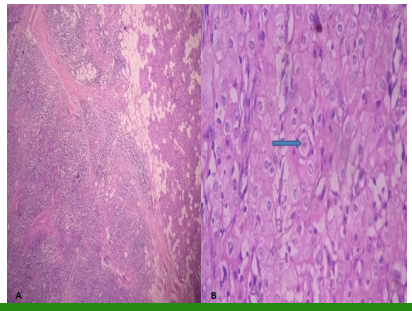
Microscopy: Haematoxylin and eosin stained sections showed an encapsulated malignant epithelial tumour with large irregular islands of tumour cells separated by fibrous septae, which were heavily infiltrated by lymphoid cells. Lymphocytes were also seen infiltrating the tumour nests [Table/Fig-1a,b,2A].

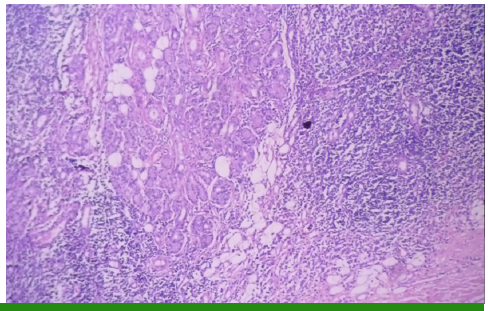
Large tumour cells with eosinophilic cytoplasm, indistinct cell borders, vesicular nuclei and prominent nucleoli were seen. [Table/Fig-2b]. Occasional atypical mitotic figures (1-2/hpf) were noted. Some of the tumour cells were arranged in a glandular pattern surrounded by lymphoid stroma [Table/Fig-3]. Multiple peri-tumoral lymph nodes were seen, one of which showed metastatic tumour deposits. On the basis of histopathological findings, a diagnosis of Lymphoepithelial Carcinoma of the parotid gland was made.

DISCUSSION

The term lymphoepithelioma was introduced in 1921 to refer to an undifferentiated carcinoma of the nasopharynx having a dense lymphocytic component [4].







[Table/Fig-1a]: 10x H&E – Tumour showing squamoid component surrounded by lymphoid stroma
[Table/Fig-1b]: 40x H&E – Shows pleomorphic squamoid cells with large, irregular & hyperchromatic nuclei
[Table/Fig-2a]: 10x H&E – Shows partial encapsulation of the tumour with focal islands of squamoid tumour cells against a dense lymphoid stroma with adjacent normal salivary gland tissue
[Table/Fig-2b]: 40x H&E – shows squamoid tumour cells with vesicular nuclei & prominent eosinophilic nucleoli
[Table/Fig-3]: 10x H&E – Glandular component of the tumour surrounded by lymphoid stroma

In 1952, Godwin in his article titled "Benign lymphoepithelial lesion of the parotid gland" described the potentiality and histological components of this entity in detail [5].

The malignant variant of this neoplasm was first reported by Hilderman et al., in 1962 [6]. Since then, malignant lymphoepithelial lesion has become a distinct entity with well-defined histopathologic features [2].

LEC is an unusual squamous cell neoplasm, with a near 100% association with Epstein Barr virus [1,2].

Over 100 cases of LEC have been reported in world literature till date with the vast majority of these occurring in North American and Greenland Eskimos, and Asian Orientals with a slightly higher female preponderance [1,2]. It is a demographically and histologically unique malignancy. These factors indicate a complex interaction of ethnic, geographic and viral factors in the pathogenesis of salivary gland LEC [1]. In the present case report, we discuss a case of LEC in a female patient of Indian origin.

Although the origin and pathogenesis of LEC is still largely debated, few studies carried out by various authors have confirmed the squamous epithelial origin of the neoplastic cells, through electron microscopy [2,4].

Amongst salivary glands, parotid gland is affected in approximately 80% of the cases, as in the present case, followed by submandibular gland [1].

Clinically, LEC presents as a parotid or submandibular swelling, which may be longstanding with recent rapid increase in size and with or without pain, as in the present case. Facial nerve palsy has been reported in about 20% of the cases [1]. In our case, facial nerve involvement was not seen.

LEC of salivary glands is morphologically indistinguishable from nasopharyngeal carcinoma [1].

On gross examination, the tumour is encapsulated, partially circumscribed, multinodular and infiltrative. It is firm, fleshy and varies in colour from grey-tan to grey yellow [1,2]. Our case had similar findings.

The classical histopathologic pattern of infiltrative sheets and islands of syncytial epithelial cells, separated by lymphoid stroma, was also seen in our case.

The pleomorphic tumour cells possess indistinct cell borders, lightly eosinophilic cytoplasm and vesicular nuclei with prominent eosinophilic nucleoli. The tumour is richly infiltrated by lymphocytes and plasma cells. The presence of all these classic features helped in clinching the diagnosis in the present case.

On immunohistochemistry, the carcinomatous cells show positivity for cytokeratin 7, LMP-1 and vimentin. These cells are found to be negative for cytokeratin 20 and lymphoid markers. In our case, immunohistochemistry could not be performed, as the patient was referred to a higher centre for management and was lost to follow up.

Many authors have implied that, LEC arises from the epithelial component of a pre-existing benign lymphoepithelial lesion. No association with Sjogren's syndrome has so far been reported [2]. No history suggestive of any association with a pre-existing lesion or Sjogren's syndrome was available in the present case.

A 5 year survival rate of 75-86% has been reported in patients treated by combined surgery and radiation therapy. Local recurrence is known to occur [1].

Since it is difficult to distinguish LEC from nasopharyngeal carcinoma, a thorough examination of nasopharynx is mandatory, before considering a primary salivary gland LEC.

Following the histological diagnosis of LEC of parotid gland, a thorough clinical examination was conducted in the present case and involvement of the nasopharynx was ruled out.

CONCLUSION

Since, primary LEC rarely occurs in the salivary gland and with not many reports of its occurrence in Indian ethnic origin, one may overlook its possibility. The present case report highlights the importance of considering LEC in the differential diagnosis of primary salivary gland tumours, despite its rarity.

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