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Lymphoepithelial Carcinoma: A Rare Parotid Gland Tumour

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Abstract: ***Primary lymphoepithelial carcinoma (LEC) is a rare type of undifferentiated carcinoma that accounts for less than 0.4% of all salivary gland malignancies with majority involving the parotid gland. The disease usually associated with Epstein - Barr virus and histopathologically it is indistinguishable from metastatic undifferentiated nasopharyngeal carcinoma. Therefore, meticulous examination of the nasopharynx should be done to rule out metastatic lymphadenopathy of the parotid nodes with nasopharyngeal carcinoma. Once the metastatic disease is excluded, primary diagnosis of lymphoepithelial carcinoma can be made. Primary surgical excision with postoperative radiotherapy is the most widely accepted approach is managing the disease with better outcome. Hereby we present a case report of a 41 - year - old lady with primary lymphoepithelial carcinoma of the parotid gland.***

Keywords: Lymphoepithelial carcinoma, Salivary gland neoplasms, Parotid neoplasms, Histopathology, Immunohistochemistry, Treatment

1. Introduction

Parotid gland constitutes about 3% of all the head and neck neoplasms and the most common type of the parotid gland malignancy is mucoepidermoid carcinoma which comprises about 40% of total cases followed by adenocystic carcinoma. [2] Primary lymphoepithelial carcinoma is a rare type of malignancy which occurs in parotid gland with an increase occurrence among the Eskimos and Chinese populations. Neighbouring pharyngeal malignancy especially from nasopharynx and oropharynx may cause possible parotid nodal metastasis, therefore the upper aerodigestive tract should be evaluated thoroughly before initiating any treatment modality. [2] These tumors mainly affect females in the fifth decade of life. [5] Although it is clinically significant due to its malignant entity, it is difficult to establish a diagnosis or to form a differential diagnosis by preoperative imaging of a salivary gland mass. (3)

Here we present a case report of a middle - aged woman who came with short history of infra auricular swelling who was confirmed with lymphoepithelial carcinoma of the parotid gland based on histopathological examination, imaging and the treatment modality that was provided to her.

2. Case Report

A 41 - year - old lady came to our clinic having left infra auricular swelling which enlarged rapidly in three months. Swelling was painless with no history of head and neck cancers, salivary gland problems, autoimmune diseases or facial trauma, also denies nasal block, epistaxis, hearing disabilities, tinnitus. Physical examination revealed a round, fixed and painless tumour, with approximately 3 cm in diameter over her left infra auricular region, skin appears smooth without numbness and no facial asymmetry noted. There was no other palpable neck mass. Fibre optic nasopharyngoscopy revealed normal nasopharynx and oropharynx findings.

Initial fine needle aspiration cytology of the swelling shows, benign salivary gland element, was proceeded with imaging, Contrast - enhanced computed tomography (CECT) neck, showed left intraparotid mass involving both superficial and deep lobes. [Figure 1, Figure 2]. Patient underwent left subtotal parotidectomy, and the histopathology examination of the intraoperative specimen reported as lymphoepithelial carcinoma of parotid gland. [Figure 3, Figure 4]. Subsequently patient was screened for nasopharyngeal carcinoma by taking blind biopsy from nasopharynx which shows no evidence of malignancy. Proceeded with a CECT staging (Neck, Thorax, Abdomen, Pelvis), which shows no evidence of regional and distant metastasis. Patient underwent completion of left parotidectomy with facial nerve preservation, and left modified radical neck dissection (MRND) type 1 and intraoperatively histopathology examination shows no malignancy seen in residual parotid tissue, regional lymph nodes and adjacent fibrofatty tissue. with no distant metastasis. Facial nerve is intact after the first and second surgery. Patient was subsequently referred to oncology department for radiotherapy/ concurrent chemoradiotherapy.

Figure 1

Figure 2

CECT Neck showing ill defined enhancing mass at left parotid region measures 2.3cmx 2.8cmx3.0cm (AP x W x CC)

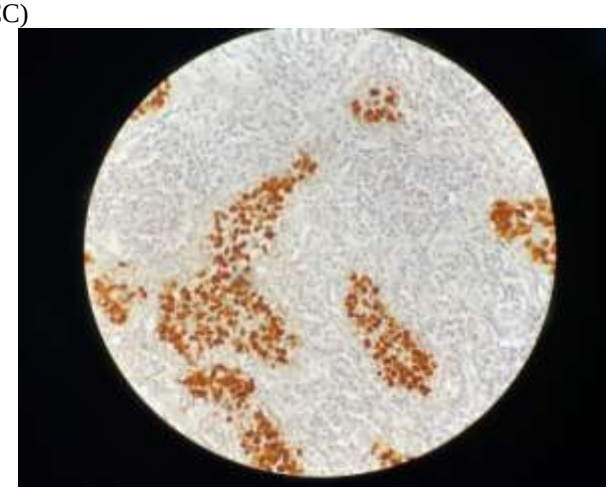

Figure 3: Microscopic appearance: Tumour cells with poorly defined borders, vesicular chromatin with prominent nucleoli and dense lymphoplasmacytic inflammatory infiltrates (using microscope BX43, under magnification x40).

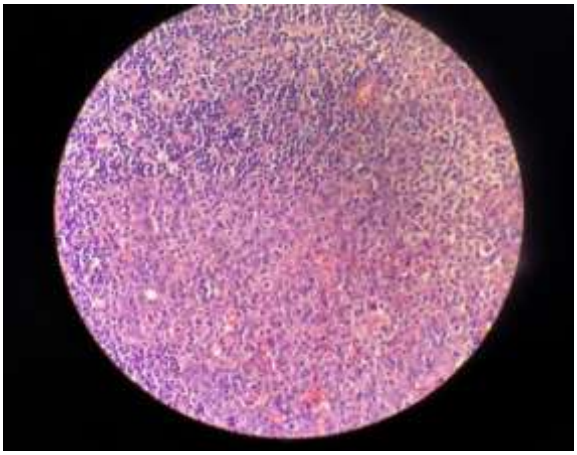

Figure 4: Microscopic appearance: In situ hybridization for EBV - encoded small RNA (EBER) show strong nuclear positivity. (using microscope BX43, under magnification x40).

3. Discussion

Lymphoepithelial carcinoma is a rare tumor of the salivary which may also be found in multiple other parts of soft tissues. (2) Hilderman first described lymphoepithelial carcinoma in 1962 as a rare type of malignant tumour which comprises 0.3% - 5.9% of all the salivary gland tumours. (4). They are categorized into the benign and malignant histologies with a variety of differential diagnoses with different treatment modality and outcome. However, FNA cytology is not definitive in some cases of improper sampling or inaccessible tumor location and therefore, preoperative imaging plays a vital role in the management of salivary gland tumors. LEC of the parotid is usually presented as a non - tender progressively increasing swelling, with significant heterogeneous or homogeneous enhancement seen in CT or MRI scans. Facial nerve is rarely involved in LEC and more than 40% of patients present with cervical node involvement. (1)

Parotid gland is the most common salivary gland tumour involved, which comprises about 80% of the salivary gland tumour. The general principle about salivary gland malignancy is that the smaller the salivary gland involved, the higher the malignancy rate, therefore the malignancy rate increases from 20% to 25% in the parotid gland to 40% to 50% in the submandibular gland whereas, 50% to 81% in the sublingual and minor salivary glands. (3)

Possible regional metastasis from the nasopharynx need to be ruled out while diagnosing lymphoepithelial carcinoma due to completely different therapeutic approach of the both the diseases need to considered. Nonkeratinizing, undifferentiated nasopharyngeal carcinoma (NPC) and salivary primary lymphoepithelial carcinoma share the similar histological presentation and therefore random biopsy of the normal looking nasopharyngeal mucosa is required to establish the diagnosis of lymphoepithelial carcinoma. Histologically, primary lymphoepithelial carcinoma described as a well circumscribed nodules, anaplastic cells with prominent eosinophilic nucleoli arranged in nests, sheets and cords of syncytial - like growth pattern, surrounded by moderate to heavy lymphocyte infiltrates. Immunohistochemical positive staining of neoplastic cells for cytokeratin, epithelial membrane antigen, EBV, lymphoid cells for both CD 20 and CD 3 markers suggestive of presence of B - cell and T - cell needed to establish the initial histological diagnosis. (6)

Fine - needle aspiration cytology (FNAC) is a primary diagnostic tool for salivary gland tumours, however, the diagnostic accuracy, sensitivity, and specificity of fine - needle aspiration cytology in the diagnosis of salivary gland lesions are subjective in respect to diverse morphologic patterns and overlapping features between benign and malignant lesions, also as seen in this cases. (4). Therefore Computed tomography (CT) and magnetic resonance (MR) imaging play an important role for the diagnosis, preoperative evaluation, and biopsy guidance for patients with LEC. (4) Management of the lymphoepithelial carcinoma of the salivary gland includes surgical excision, radiation therapy, and chemotherapy. The treatment of choice at present would be surgical excision followed by

postoperative radiation therapy and about 5 - year survival rate is reported to range from 50% to 90%. (4)

4. Conclusion

The case report highlights the importance of having a high index of suspicion in reaching the diagnosis of primary parotid LEC. LEC can afflict patients without any warning sign, therefore any painless parotid mass should be taken seriously to exclude malignancy at early stage itself regardless at any ethnicity.

5. Acknowledgement

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6. Conflict of interest

There is no conflict of interest

7. Ethic statement/ Confirmation of patient permission

Ethics approval not required. The patient gave her consent for publication of the pictures.

References

[1] Liu, YH., She, YY., Lin, YS., Wu, CC., (2015) Primary Lymphoepithelial Carcinoma of the Parotid Gland: A Rare Case Journal of Medical Sciences 35 (6) pg 267 - 269
[2] Yuen, JK., Hyun, SH. , Sun, HJ. , Eun, HL. , Ming, JJ., (2017) Lymphoepithelial carcinoma of the salivary glands. Pubmed Central 96 (7)
[3] Hammoud M, Serhal J, FakhreddinN, et al. Lymphoepithelial Carcinoma of the Parotid Gland case report. J Otolaryngol ENT Res.2018; 10 (6) 410 - 412
[4] Ban. X, Wu J, Mo Y, Yang Q, Liu X, Xie C, Zhang R, (2014) Lymphoepithelial Carcinoma of the Salivary Gland: Morphologic Patterns and Imaging Features on CT and MRI. American Journal of Neuroradiology September 2014, 35 (9) 1813 - 1819
[5] Christopher GT., Thomas MS., Grace YT., Luke JS., Barry R. (2012) Lymphoepithelial Carcinoma: A Case of a Rare Parotid Gland Tumor. Pubmed Perm J.2012 Summer; 16 (3): 60 - 62
[6] Athanasios P, Evangelos P, Christina N, Aikaterini, (2018) Primary Lymphoepithelial - Like Carcinoma of the Parotid Gland - Case Presentation. Balk J Dent Med, 2018; 43 - 48