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Case Report

Pathology Section

Pathological Portrait of a Submandibular Gland Lymphoepithelial Cyst in a Non HIV Patient: A Case Report

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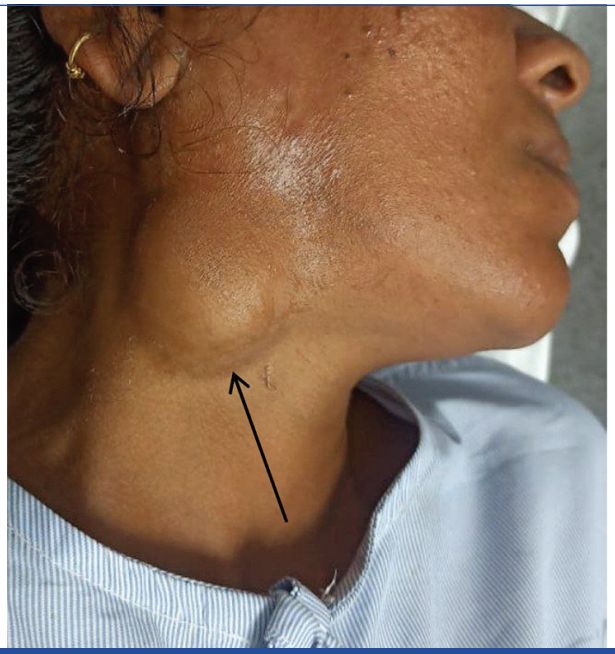
ABSTRACT

Lymphoepithelial Cysts (LEC) manifest as benign, unilocular to multilocular lesions predominantly in the head and neck areas. LEC typically localise in the salivary glands, with the parotid gland being more common and the submandibular gland being a rare occurrence. Additionally, they may be observed in the oral cavity, with a particular affinity for the floor of the mouth. It manifests as a painless, solitary cystic mass situated close to or within the salivary gland. It predominantly occurs in individuals infected with Human Immunodeficiency Virus (HIV). It typically arises as a consequence of lymphocyte-induced dilation of the cystic duct, and in accordance with index case, the definitive diagnosis is consistently confirmed through postoperative histopathological examination. They are infrequently observed in immunocompetent individuals and may have a connection to Sjögren's syndrome. Numerous contemporary investigative methods are at our disposal, with Fine Needle Aspiration Cytology (FNAC) emerging as a swift diagnostic tool for promptly confirming a LEC. Additional diagnostic modalities encompass Ultrasonography (USG) and Computed Tomography (CT). The primary treatment for an LEC continues to be surgical intervention. Understanding the characteristics and behaviour of such cysts in non HIV patients is critical for accurate diagnosis, appropriate therapy, and, perhaps, enhancing the understanding of the spectrum of conditions associated with these cyst formations. Investigating LEC occurring in the rare location of the submandibular salivary gland, this work is distinctive in that it involves an immunocompetent individual. The present case is of a 42-year-old woman who elucidates the diagnostic pathway, encompassing both FNAC and histopathology.

Keywords: Painless swelling, Salivary gland, Submadibular cyst

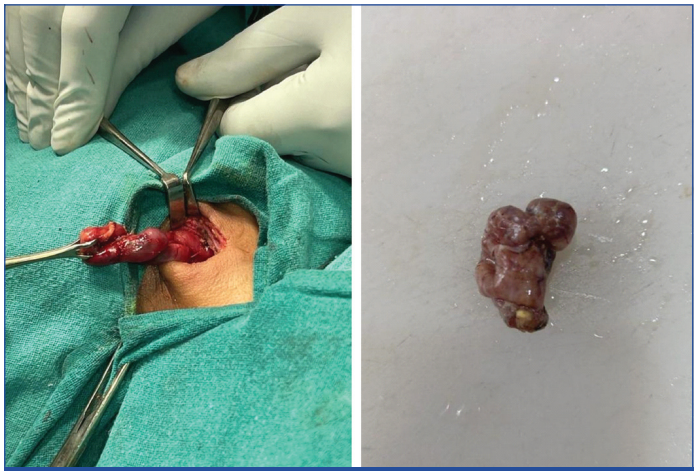
CASE REPORT

A 42-year-old female of Asian origin presented to the Outpatient Department (OPD) seeking medical assistance for a painless swelling beneath her right-sided lower jaw [Table/Fig-1]. The patient noted that the swelling had gradually increased in size over the past six months, accompanied by intermittent pain while swallowing. She denied experiencing significant weight loss, fever, or other systemic symptoms. Upon extraoral examination, a soft, non tender, compressible enlargement in the right submandibular area measuring 8×6×5 cm was observed. No abnormalities were detected during the intraoral examination.



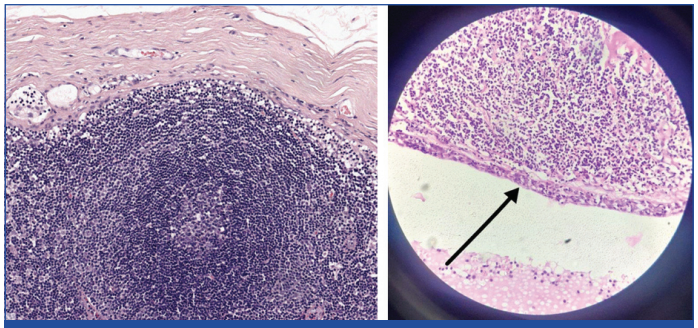
[Table/Fig-1]: Clinical picture of the lesion on the right side of the neck.

The swelling was aspirated with a fine needle, yielding a straw-coloured fluid. Smears revealed numerous lymphocytes and squamous cells dispersed in a proteinaceous background. The cytology strongly suggested a LEC. The patient underwent testing for HIV using both Enzyme-Linked Immunosorbent Assay (ELISA) and Western blot, with results indicating a negative status. Subsequently, the patient underwent complete excision of the lesion [Table/Fig-2]. The postoperative period was uneventful. The excised specimen was then sent for histological analysis. Grossly, it appeared as an irregular, brownish tissue piece measuring 5×3.5×2 cm [Table/Fig-3].

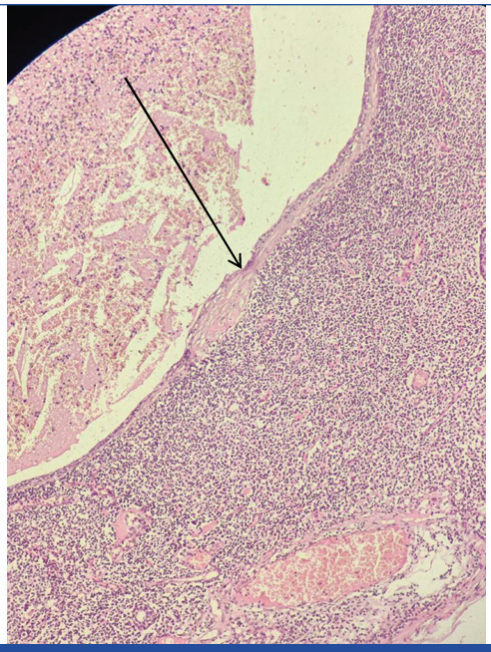


[Table/Fig-2]: Excision of the lesion.
[Table/Fig-3]: Gross specimen of the excised lesion. (Images from left to right)

Microscopically, the section revealed a cyst lined by thin stratified squamous epithelium surrounded by a dense lymphocytic infiltrate and lymphoid follicles with germinal centres [Table/Fig-4,5]. There was evidence of lymphocyte infiltration in the epithelium [Table/Fig-6]. As a result, the final diagnosis of LEC was made.



[Table/Fig-4]: Lymphoid follicle with germinal center (H&E x100).
[Table/Fig-5]: Stratified squamous lining (H&E x400). (Images from left to right)



[Table/Fig-6]: Squamous lined cyst with amorphous debris in the lumen, cholesterol clefts and dense lymphocytic infiltrate in the wall (H&E x100).

DISCUSSION

Bernier JL and Bhaskar SN coined the term "Lymphoepithelial Cyst" (LEC) to emphasise that the lesion is not an embryologic remnant. It is described as single or multiple cysts within lymph nodes connected with salivary glands [1]. LEC, also known as branchial cysts, are most commonly found in the lateral cervical region, just below the mandibular angle and anterior to the sternocleidomastoid muscle. LECs are frequently associated with salivary glands, particularly the parotid gland, with rare occurrences in the submandibular gland. These cysts have a strong affinity for the lateral neck and are less common in the oral cavity or the parotid gland. Branchial cysts that do not involve the salivary component often develop in the lateral cervical region, including the lymph node. LECs range in size from 0.5 to 5.0 cm and can lead to significant cosmetic deformities as well as physical discomfort [2].

The LEC has been associated with HIV infection, particularly as part of the diffuse infiltrative lymphocytosis syndrome. In HIV-positive individuals, HIV-associated salivary gland illness is characterised by lymphoid hyperplasia of the parotid and sometimes submandibular glands, as well as LEC and lymphoepithelial lesions. HIV-associated salivary illness affects roughly 3-10% of HIV-positive patients. It affects individuals of all ages and genders, including children and adults. Salivary gland disorders often precede Acquired Immunodeficiency Syndrome (AIDS), serving as an early indicator of HIV infection. It is common for the condition to manifest bilaterally and exhibit a correlation with cervical lymphadenopathy [3]. The primary cause of concern associated with LEC in HIV-positive individuals is progression to lymphoma [4], which makes investigations for HIV imperative, similar to the case mentioned.

LECs may co-exist with Mikulicz's disease. Additionally, these cysts may also be observed concurrently with myoepithelial sialadenitis. In contrast, Sjögren's syndrome involves the minor salivary glands but lacks a lymphoepithelial component. Another close differential diagnosis is that of Brachial cleft cyst, which is infrequent in the parotid [5]. When comparing an HIV-infected patient to a non HIV patient, CT and Magnetic Resonance Imaging (MRI) demonstrate evidence of bilateral and multicystic lymph nodes, which could potentially be regarded as a significant radiological diagnostic indicator [5].

The treatment of LEC incorporates both conservative and surgical methods. The conservative method involves decompressing the cyst through fluid aspiration, thereby reducing the pressure. This procedure is particularly recommended for immunodeficient patients, such as those with HIV, where the potential risks associated with surgical management outweigh the benefits. Additional conservative treatments may involve external radiotherapy. However, the most efficient and effective treatment is surgical management, which includes full enucleation of the cyst as well as excision of the involved gland. Excision typically results in a complete cure for the majority of patients, with no likelihood of recurrence [6]. To date, LECs have shown no recurrence or metastasis. Consequently, it is crucial to distinguish them from more aggressive lesions such as extranodal marginal zone lymphoma, Warthin tumour, and mucoepidermoid carcinoma [7].

While LECs are primarily associated with HIV infection, they can also occur in non HIV patients, albeit much less frequently. Below are a few reported LEC cases in non HIV patients [Table/Fig-7] [8-16]. As observed, LECs are predominantly situated in the parotid gland among non HIV patients. The scarcity of submandibular occurrences emphasises the importance of considering parotid LECs, particularly in non HIV individuals presenting with swellings in that area.

S. No.	Study	Publication year	Age (years)/ Gender	Site
1.	Kojima M et al., [8]	2009	36/M 53/M 57/F	Parotid gland
2.	Ahamed AS et al., [9]	2014	32/F	Submandibular gland
3.	Pillai S et al., [10]	2016	49/M	Parotid gland
4.	Khadilkar MN et al., [11]	2017	55/F	Parotid gland
5.	Joshi J et al., [12]	2018	35/M	Parotid gland
6.	Sunitha Carnelio MD et al., [13]	2018	28/M	Parotid gland
7.	Thong HK et al., [14]	2019	45/F	Parotid gland
8.	Park YY et al., [15]	2019	45/M	Parotid gland
9.	Liao Y et al., [16]	2023	44/M	Parotid gland
10	Present study	2024	42/F	Submandibular gland

[Table/Fig-7]: Few reported case along with present study [8-16].

CONCLUSION(S)

LEC is traditionally associated with the parotid gland, prevalent in HIV-infected individuals, often progressing to lymphoma. This case highlights its occurrence in an atypical site-the submandibular gland-and in a non HIV-infected individual. Diagnostic methods include both surgical and conservative approaches. LECs exhibit no recurrence or metastasis. This report challenges conventional patterns, emphasising the need for comprehensive diagnostic strategies and raising awareness about LEC's diverse presentations beyond the expected scenario.

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