

CASE REPORT

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Lymphoepithelial cyst of submandibular region: a case report

Sana Iqbal^{1*}, Sufyan Ahmed² and Mehwash Kashif³

Abstract

Background: The oral lymphoepithelial cyst (LEC) is a rare, soft-tissue, developmental cyst, initially presented by Gold in 1962 as a “branchial cleft cyst”. It may occur in the pancreas, tongue, neck, and other regions of the oral cavity. No study has been reported in Pakistan and Karachi reporting a case of lymphoepithelial cyst in the submandibular region. This rare case of LEC has been reported to help the clinicians to bring LEC into their differential diagnosis for lesions affecting the submandibular region.

Case presentation: We report a case of 57 years old female reported to the Maxillofacial OPD of a tertiary care hospital with the complaint of swelling on the right side of the neck for 3 weeks which was rapidly increasing in size. She was having difficulty in mastication. On extra-oral examination, there was swelling on the right side in the submandibular area. The overlying skin is normal with no evidence of pus and discharge. On palpation, the swelling was soft, non-tender, non-fluctuant, non-displaceable into the submandibular area. Lymph nodes were impalpable. Excision of the lesion performed under general anesthesia and biopsy revealed lymphoepithelial cyst associated with submandibular gland. Here we present an attention-grabbing case of swelling in the right submandibular region which was provisionally diagnosed as a malignant submandibular lymph node however later evidence histopathologically as a lymphoepithelial cyst of the submandibular gland.

Conclusion: An unusual cause of swelling in the neck is lymphoepithelial cysts. Submandibular gland appearance is not usual and can prove to be a clinical problem.

Keywords: Lymphoepithelial cyst, Submandibular gland swelling, Etiopathogenesis, Case report

Background

The oral lymphoepithelial cyst (LEC) is a rare, soft-tissue, developmental cyst, initially presented by Gold in 1962 as a “branchial cleft cyst”. It may occur in the pancreas, tongue, neck, and other regions of the oral cavity (Dalal et al. 2016).

The review of the literature published in 2017, resulted in 316 oral LEC cases resulting from twenty-five case reports, three case studies/retrospective studies with comprehensive case-by-case details, and seven studies with summarized results. Out of 31,564 biopsies

accessed during the study period, the twenty-six oral LECs represented 0.08%. Among twenty-five patients, fourteen women, and eleven men, with a mean age of 33.04 ± 9.81 years, were affected. They appeared as soft (92%) nodules, with a soft (24%) or firm (76%) consistency and regular (28%), yellow to normal (20%), yellow (32%), or white (20%) color, in the tongue (69.23%) or mouth floor (30.77%). They've been desquamated epithelial cells, amorphous eosinophilic material, and/or inflammatory cells were protected by parakeratinized squamous (92.31%) or non-keratinized (7.69%) epithelium and contained (100%). The cystic cavity was partly (34.62%) or fully surrounded by the lymphoid tissue (65.38%), mostly in a follicular pattern with prominent germinal centers (53.85%) (Sykara et al. 2017).

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LEC is reported very rarely in the Pakistani population (Yazici et al. 2020). No study has been reported in Pakistan and Karachi reporting a case of lymphoepithelial cyst in the submandibular region. This rare case of LEC has been reported to help clinicians to bring LEC into their differential diagnosis for lesions affecting the submandibular region.

Case presentation

A 57 year old female reported to the Maxillofacial OPD of a tertiary care hospital with the complaint of swelling on the right side of the neck for 3 weeks which was rapidly increasing in size. She was having difficulty with mastication. The patient was a nonsmoker and was not a habitual user of areca nuts, and had no remarkable prior medical, family, and psycho-social history including relevant genetic information. No past surgical inventions. On extra-oral examination, there was swelling on the right side in the submandibular area (Fig. 1).

The overlying skin is normal with no evidence of pus and discharge. On palpation, the swelling was hard, non-tender, non-fluctuant, and non-displaceable into the submandibular area. Lymph nodes were impalpable.

The mouth opening is adequate and intraoral examination revealed bad oral hygiene with multiple broken down roots. No evidence of occlusal derangement or paresthesia. No evidence of extension of swelling intraoral. The swelling was firmly attached and does not move with

deglutination. A provisional diagnosis of malignant submandibular lymph node was made.

The patient has consented to fine-needle aspiration cytology (FNAC) and it was performed. She was also advised to do an orthopantomogram (OPG) and CT scan of the head and face with contrast.

OPG revealed no significant involvement of the bone. It revealed dental problems and periodontitis. FNAC revealed a heavy inflammatory and necrotic background with infiltration of polymorphonuclear leukocytes. A large number of nucleated and anucleated squamous epithelial cells are present. Some of these squamous cells are round to oval in shape. These cells show altered nuclear-cytoplasmic ratio, with hyperchromatic, enlarge, and irregular nuclei and dense organophilic cytoplasm. Excision of the lesion was recommended for further delineation.

CT scan revealed large necrotic lymph nodes at the level I-b, few prominent cervical lymph nodes are identified in the neck bilaterally. They are suggestive of infective and /or neoplastic etiology (Fig. 2).

Therefore, in the light of history, examination, and investigations excisional biopsy was planned. The patient was referred for general anesthesia fitness and has been advised for complete blood count, Hepatitis screening, Covid PCR, Chest radiograph, and Urea creatinine and electrolytes. She was operated on under general anesthesia. Wide excision of the mass was performed; under AAA measures, prep and drape were done. A transverse incision was marked (Fig. 3) and



Fig. 1 The extraoral presentation of swelling in the submandibular region



Fig. 2 Preoperative CT scan outlining a mass in the submandibular area

local anesthesia (Lidocaine with 2% epinephrine) was injected.

An incision was made and the flap was retracted sub-platysmal (Fig. 4).

The cyst dissected out carefully with the submandibular gland along with sparing of the marginal mandibular nerve (Fig. 5).

Hemostasis was achieved and layered closure is done by sutures of vicryl 3-0 and prolene 4-0 (Fig. 6).

After anesthesia recovery, she was hospitalized for 2 days and discharged on oral antibiotics for 5 days. She was followed up after an uneventful week and was pleased with the surgery and outcomes.

The excised mass was sent to the laboratory for histopathological examination. The excised mass was oval and measures $2 \times 2 \times 1.8$ cm. It includes the submandibular gland along with nodes. The histopathology revealed a lesion with the cystic component which is lined by stratified squamous cells showing focal atypia. In some areas, these cells are associated with lymphocytes. The central cystic area sows extensive keratin pearls and flakes of keratin. The squamous cells have round to oval nuclei with vesicular chromatin and occasionally prominent nucleoli. Focal disruption of epithelial lining is present. So final diagnosis of the lymphoepithelial cyst was made (Fig. 7).

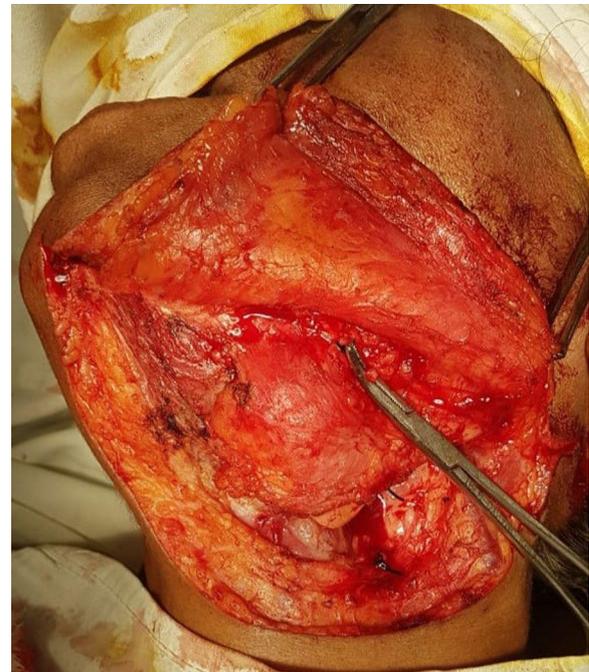


Fig. 4 Flap was raised sub-platysmal



Fig. 3 Marking of a transverse incision



Fig. 5 Excised lesion along with submandibular gland



Fig. 6 Closure of the flap

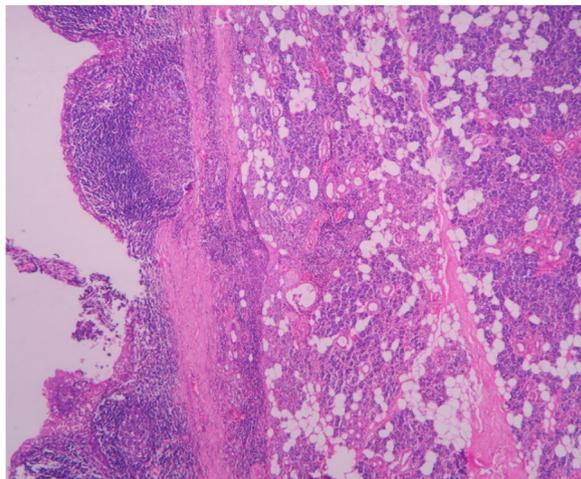


Fig. 7 A histological picture of LEC

Discussion

The lymphoepithelial cyst is typically located in adults in the lateral neck area and is therefore referred to as the lateral cervical cyst and often referred to as the brachial cyst because it is thought to derive from brachial cleft epithelial remains. This injury is generally unilateral, with a 2% bilateral incidence registered. There is no gender preference for the accident (Cheereth et al. 2017).

The majority of the lesions, for unexplained reasons, occur on the left side. The potential clinical differential diagnosis is HIV-associated salivary gland disease, chronic sclerosing sialadenitis, Warthin tumor, extra nodal marginal zone B-cell lymphoma, and salivary duct retention cyst (mucocoele), mucosa-associated lymphoid

tissue lymphoma. By conducting an aspiration, USG, or a CT, a good amount of data is obtained. In identifying the cystic material, aspiration is particularly good. The clear watery fluid is normally obtained in LEC, but pus can also be there if the cyst is infected. Cytology and culture can provide us with useful information on the quality of the type of cells and bacteria and we can prepare the antibiotic coverage as indicated for the patient. Malignant LEC or bronchogenic carcinoma is a rare risk of squamous cell carcinoma arising in the LEC. The cyst is histologically lined with stratified squamous epithelium, respiratory epithelium, or both. The lining epithelium is transformed by persistent infection into a fibrous lining or a granulomatous lining (Ahamed et al. 2014).

The clinician should be cautious in diagnosing if the cyst is diagnosed after the age of 50 years, since the risk of metastatic squamous cell carcinoma in a cervical lymph node with cystic degeneration may mimic neck LEC. The cases are of specific interest because of the rarity of BLEC in HIV-negative patients and highpoint an important differential diagnosis of parotid swelling (Thong et al. 2019). In AIDS patients and the AIDS risk community, LEC is now increasingly diagnosed every day. The pathogenesis of LEC-related aids is still not known. It is proposed that if lymphadenopathy affects the submandibular gland, the ductal wall becomes obstructed by keratin and metaplasia, resulting in the formation of a cyst (Sekikawa and Hongo 2017).

Pure surgical excision is the treatment for LEC. A sudden increase in LEC size is a common complication following an infection of the upper respiratory tract. This is largely due to reactive hyperplasia associated with necrosis, liquefaction, and suppuration of the lymphoid components. It should be recalled that the lymphoepithelial cysts are benign and should be managed as early as possible because they can turn into malignant lesions, such as malignant lymphoma, adenocarcinoma, mucoepidermoid carcinoma, and surgery remains the key stay in the treatment of both lymphoepithelial and lateral cervical cysts. Fine needle aspiration, accompanied by USG, significantly encourages, but not always, the precise diagnosis of swelling. The initial management of the infected LEC is the drainage of the cyst under antibiotic coverage, but there is always a chance of producing chronic discharge from the operated site. Surgery should be postponed until the acute episodes of infection are resolved (Pinheiro et al. 2019).

Conclusion

An unusual cause of swelling in the neck is lymphoepithelial cysts. Submandibular gland appearance is not unusual and can prove to be a clinical problem.

Abbreviations

LEC: lymphoepithelial cysts; BLEC: benign lymphoepithelial cysts.

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Authors' contributions

SI: Conceived idea. SI, SA, MK: Designed research methodology. SA, MK: Literature search. SI: Data collection, literature review. SA: Data interpretation. SI, SA: Manuscript writing. All authors have read and approved the manuscript.

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Not applicable.

Consent for publication

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Competing interest

The authors declare that they have no competing interests.

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