

## Case Report

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# Lymphoepithelial cyst of submandibular gland, a clinical dilemma- case report

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## ABSTRACT

Swellings of the head and neck region present great difficulty in accurate diagnosis as well as proper management. Swelling associated with the maxilla and mandible has a tendency to be provisionally diagnosed as orofacial space infections, which is the most common cause for acute swelling in this region. Fine needle aspiration cytology (FNAC), ultrasonogram and computed tomography can be used in supporting diagnosis. It usually occurs due to the process of lymphocyte-induced cystic ductular dilatation and the confirmatory diagnosis is always made postoperatively by histopathological examination. The mainstay in the treatment of a lymphoepithelial cyst remains the surgical approach, which includes complete enucleation of the cyst along with total excision of the involved salivary gland. Here we present an interesting case of swelling in the right submandibular region which was provisionally diagnosed as submandibular space infection, but later proven histopathologically as a lymphoepithelial cyst of the submandibular gland.

**Keywords:** Lymphoepithelial cyst, Submandibular gland swelling

## INTRODUCTION

Swellings of the head and neck region present great difficulty in accurate diagnosis as well as proper management. Neck swellings can occur as a result of accumulation of fluid, lymph, inflammatory or tumour cells in the area under skin or in deep planes. Trauma and infection are the other common cause of neck swellings.<sup>1</sup>

Swelling associated with the maxilla and mandible has a tendency to be provisionally diagnosed as orofacial space infections, which is the most common cause for acute swelling in this region. Thorough examination of orofacial region should be done to rule out foci of infection before making any treatment plan. Other possible differential diagnosis of the neck swellings are sebaceous cyst, brachial cyst, dermoid cyst, lipoma,

enlarged submandibular lymph nodes or gland.<sup>2</sup> Here we present an interesting case of swelling in the right submandibular region which was provisionally diagnosed as submandibular space infection as supported by clinical and radiographic evidence. Eventually excision of submandibular gland was done and histopathology was reported as lymphoepithelial cyst of the submandibular gland.<sup>3</sup>

## CASE REPORT

A 39 year old female patient reported to the department of oral and maxillofacial surgery with a chief complaint of swelling in the right side of the neck just below the mandible since one month. Patient had pain in the right lower posterior tooth region after which she noticed the swelling which started as a small swelling and gradually

increased to the present size. She had consulted a general dentist and took some antibiotics and analgesics and got relief from the symptoms. A few days later she experienced severe lancinating pain along with the recurrence of the swelling which did not subside with further medication. Her medical, dental and personal history was non-contributory.

On inspection, a diffuse ovoid swelling is seen in the submandibular region extending from the middle of right body of mandible to the angle of the jaw anteroposteriorly and of about 5cm in size and 3cm extensions inferiorly below the border of the mandible with ill-defined margins blending with adjacent skin (Figure 1).



**Figure 1: Diffuse ovoid swelling in the submandibular region.**

The swelling was firm on palpation, with no local elevation of temperature and the swelling was non reducible, non compressible and fixed to the underlying tissues. Intra oral examination revealed grossly decayed right mandibular molar tooth. No intra oral swelling was evident in relation to the decayed tooth and no vestibular tenderness was present.



**Figure 2: OPG revealing radiolucent lesion in the apical region of the grossly decayed right mandibular molar tooth.**

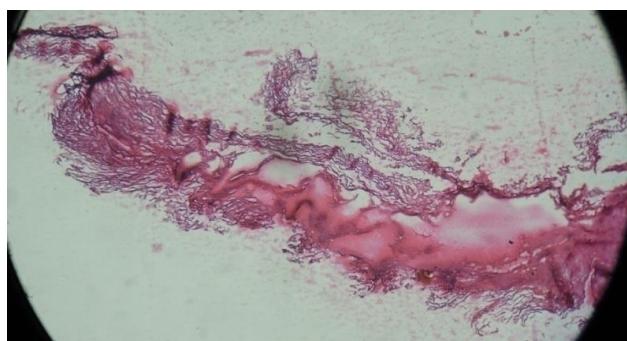
A clinical provisional diagnosis of submandibular space infection with abscess was made and radiographic investigations were sought. IOPA and OPG were taken and both revealed a radiolucent lesion in the apical region of the grossly decayed mandibular molar tooth (Figure 2). From the above findings, a diagnosis of submandibular

abscess with an abscess was made. Taking these findings in to consideration we proceeded with extracting the offending tooth and put her on antibiotic coverage and she was reviewed after 3 days. Although she was relieved from pain, swelling was still present. So a USG neck was advised and the radiologist commented as a non-vascular oval well defined hypoechoic mass in the submandibular compartment extending posteriorly up to the anterior border of the sternocleidomastoid muscle which is in close approximation with the submandibular salivary gland and he concluded it as a chronic abscess.



**Figure 3: White cheesy material obtained on aspiration of lesion.**

Since the provisional diagnosis of an abscess is supported by USG, we decided to go for an aspiration followed by incision and drainage. Aspiration was performed which yielded a white cheesy material (Figure 3) which was not suggestive of an abscess. The content was sent for histopathological examination and aspirate was reported to contain flakes of keratin (Figure 4) and a provisional diagnosis of sebaceous cyst was made.

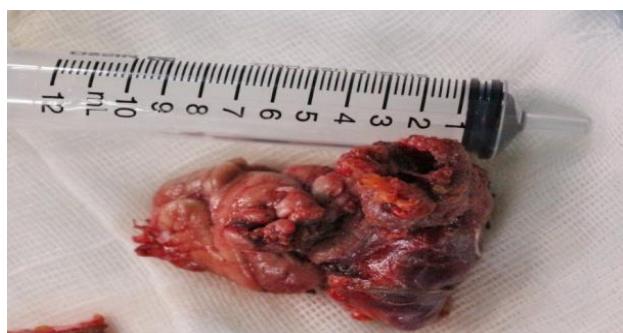


**Figure 4: Histopathological examination of aspirate reported to contain flakes of keratin.**

Patient reviewed after a week and she gave a history of pus drainage extra orally from the site of aspiration. She was taken to the operation theatre for excision of the infected sebaceous cyst under local anesthesia. But intra operatively it was found that the lesion was a cyst like structure with plenty of thick cheesy material with firm adherence to right submandibular gland. Preservation of the gland was difficult and hence submandibular gland excision along with cystic lesion was proceeded under general anesthesia. (Figures 5 and 6).

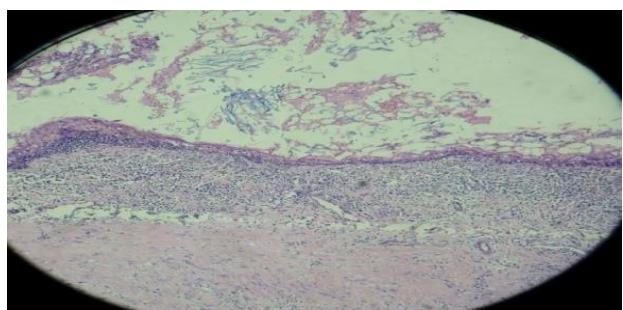


**Figure 5: Pus drainage extra orally from the site of aspiration.**



**Figure 6: Submandibular gland along with cystic lesion post excision.**

The excised specimen was sent for biopsy and the report came as lymphoepithelial cyst of submandibular gland. On microscopic examination the biopsy tissue showed an orthokeratinised stratified squamous surface epithelium. The underlying connective tissue is fibrovascular with aggregate of lymphoid tissue (Figure 7).



**Figure 7: Orthokeratinised stratified squamous surface epithelium, fibrovascular connective tissue with aggregate of lymphoid tissue.**

## DISCUSSION

Lymphoepithelial cyst are usually found in the lateral neck region of adults and hence called as lateral cervical cyst and sometimes called as brachial cyst because it is believed to be originating from epithelial remnants of brachial cleft. This lesion is usually unilateral with reported bilateral incidence of 2%. There is no sex predilection for the lesion and are not usually associated

with any malformations in the head and neck region. Majority of the lesion occur in the left side for unknown reason.

Cervical LEC are soft fluctuant mass and may reach up to 10cm in size, presenting in second or third decades of life. It is mostly diagnosed when the lesion become complicated with dentoalveolar or upper respiratory tract infections or trauma.<sup>1</sup> Cervical LECs are usually associated with progressive symptoms like swelling, pain, infection and pressure sensation or sometimes respiratory obstruction. Four major theories have been put forth and explained by Maran and Buchnan in 1974 which are, 1) brachial apparatus theory, 2) cervical sinus theory, 3) thymopharyngeal theory and 4) inclusion theory.<sup>2</sup>

Clinically the possible differential diagnosis is dermoid cyst, cystic hygroma, cervical ranula, inflammatory or infectious lymphadenopathy etc. A good amount of information is obtained by doing an aspiration, USG or a CT. Aspiration is especially good in identification of the cystic content. In LEC usually a clear watery fluid is obtained but pus can also be there if the cyst is infected.<sup>3</sup> Cytology and culture can give us a valuable information about the type of cells and bacteria in the content and we can plan the antibiotic coverage as indicated for the patient.

Squamous cell carcinoma occurring in the LEC or a, malignant LEC or Brachiogenic carcinoma is a rare possibility. Martin proposed four criteria for the provisional diagnosis of such a clinical lesion. They are 1) the tumour should occur in the line extending from anterior to the tragus to the anterior border of sternocleidomastoid muscle, to the clavicle. 2) The histologic appearance of the lesion should be consistent with an origin from tissue present in the brachial vestigial. 3) The patient must survive at least five years without development of other tumour than can be regarded as primary. 4) It must be demonstrated that cancer developed in the wall of an epithelial lined cyst situated in the lateral aspect of the neck.<sup>4</sup>

Histologically the cyst is lined by stratified squamous epithelium or respiratory epithelium or sometimes both. Recurrent infection changes the lining epithelium in to a fibrous lining or a granulomatous lining. When we consider the lining epithelium, those having the columnar epithelium are clinically dormant while those having squamous epithelium with purulent content are usually symptomatic.<sup>5</sup>

Hildebrand described a case of LEC in salivary gland in 1895. Bhasker et al suggested a hypothesis regarding the development of LEC in the salivary glands that the cystic transformation of the salivary gland epithelium entrapped in lymph node within the gland result in such a cyst. Ricket suggested that the majority of LEC occur in the neck and originates from the remnants of brachial apparatus.<sup>6</sup>

Knapp in 1970 proposed a theory for the origin of intra oral LEC. He proposed that small nodular aggregates of lymphoid tissue are scattered whole throughout the oral cavity and within the lymphoid tissue small epithelial lined lymphoid crypts are observed. A small number of these crypts undergo obstruction, with dilatation and eventual formation of the cystic cavity.<sup>7</sup>

Khelemsky and Mandal reported a case of LEC of floor of mouth in 2010 and they typically describe the lesion as a freely mobile, dome shaped, submucosal nodule, with a smooth non ulcerated surface with yellowish to white hue and cheesy consistency.<sup>8</sup>

If the cyst is diagnosed after the age of 50 years, the clinician should be careful in diagnosing because the chance for metastatic squamous cell carcinoma in a cervical lymph node with cystic degeneration may mimic LEC of neck. Now a day's LEC are increasingly diagnosed in AIDS patients and in AIDS risk group. The pathogenesis of aids related LEC is not yet understood. It is suggested that if the submandibular gland is affected by lymphadenopathy, the ductal wall become obstructed by keratin and metaplasia, leading to the development of a cyst.<sup>9</sup>

Treatment of LEC is pure surgical excision. Intraoperative findings suggest the occurrence of four types cervical LEC. They are: 1) Those lie superficially beneath the platysma and cervical facia along the anterior border of the sternocleidomastoid muscle. 2) Lies on the great vessels. 3) Extends through the carotid bifurcation to the lateral wall of pharynx, occasionally exhibiting a prolongation to the skull base. 4) Cyst usually lined by columnar epithelium and lying against the pharyngeal wall.<sup>5</sup>

Rapid increase in the size of LEC is a usual complication following upper respiratory tract infection. This is mainly due to the reactive hyperplasia of the lymphoid components associated with necrosis, liquefaction and suppuration. Ingoldby suggested that the acute or chronic infectious and inflammatory conditions of the cyst will complicate its proper diagnosis and thus the management. Fine needle aspiration followed by a USG greatly facilitate in the accurate diagnosis of the swelling, but not always. Drainage of the cyst under antibiotic coverage is the initial management of infected LEC, but there is always a risk of developing persistent discharge from the operated site. Surgery should be delayed until the acute episodes of infection are cleared.<sup>2</sup> Hence all infected LEC should be through inspected for any orofacial foci and managed accordingly. In case of infected cyst, definitive surgical treatment can be delayed and should be treated with antibiotics and drainage. Surgery in infants should

be deferred up to the age of 4 years. Recurrence is extremely rare if complete surgical excision is obtained.

## CONCLUSION

Lymphoepithelial cyst (LEC) is a rare cause of swelling in the neck. Since in present case the presentation was atypical and they also manifest in major salivary glands especially in the parotids. Presentation in the submandibular glands is rare and can prove as a clinical dilemma. It should be kept in mind that the lymphoepithelial cysts are benign in nature and should be treated as early as possible as it can transform itself into a malignant lesion such as malignant lymphoma, adenocarcinoma, mucoepidermoid carcinoma and surgery remains the main-stay in the treatment of all lymphoepithelial and lateral cervical cyst.<sup>10</sup>

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