

Case Report

Bilateral Multiple Lymphoepithelial Cysts of Palatine Tonsils

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ABSTRACTS

Lymphoepithelial cyst of oral cavity is a rare cystic lesion that presents as an asymptomatic submucosal mass and is usually discovered during routine dental examinations. The site most commonly affected is floor of the mouth but tonsillar involvement seems to be very rare. Multiple lymphoepithelial cysts have been reported in parotid of HIV positive patients however in oral cavity they usually present as solitary lesions. We report a case of multiple bilateral lymphoepithelial cysts of the palatine tonsils in a 72 years old male that presents with fever, pain and obstructive sleep apnea. We did not find any bilateral or multiple tonsillar lymphoepithelial cysts in the literature and the present paper seems to be the first report. We suggest although lymphoepithelial cysts rarely occur in the tonsils but they should be considered in differential diagnosis of adenotonsillar enlargement and related obstructive sleep apnea.

Keywords: Palatine Tonsil, lymphoepithelial Cyst

Introductions

Oral lymphoepithelial cyst (LEC) is a rare lesion that develops within oral and pharyngeal lymphoid tissues and is microscopically identical to branchial cleft cyst but smaller in size (1). Histologically, it presents as a cystic lesion lined by stratified squamous epithelium with desquamated epithelial cells seen filling the cyst lumen. Surrounding the cyst lining there are variable amounts of lymphoid tissue with occasional germinal center formation

(2). Etiologically they are thought to arise from entrapment of epithelium within lymph node or lymphoid tissue and subsequent epithelial proliferation results in clinically evident mass. It is also possible that they may develop from surface mucosal epithelium that becomes enclaved in lymphoid tissue during embryogenesis (1). Multiple LECs have been reported in parotid of HIV positive patients (3-7); however in the oral cavity they present as a solitary asymptomatic submucosal mass. The most commonly affected sites are floor of the mouth followed by ventral

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and posteriolateral portions of the tongue but tonsillar involvement is very rare. We could only find one case of solitary LEC of the tonsil in literature (8). To the best of our knowledge this is the first report of bilateral multiple tonsillar LECs presented with pain, fever and obstructive sleep apnea (OSA).

Case report

A 72 year old Iranian male was admitted to Ayatollah Taleghani University Hospital in Tehran, Iran with pain, snoring, sleep apnea and fever since 12 months. On systematic physical examination no abnormality was detected except for the bilateral tonsillar hyperplasia and his medical history was unremarkable. In examination right tonsil was +1 in tonsillar hypertrophy grading scale with a yellowish surface containing a cystic lesion, and left tonsil was +2. Tonsillectomy was performed under general anesthesia.

On gross examination in the right tonsil a cystic space with 1.2 cm in diameter containing a creamy material was observed. After tissue fixation in formaldehyde (formalin) solution, the specimen was dehydrated by using a series of ethanol solutions of increasing concentration until pure, water-free alcohol was reached. Then clearing process was established by using xylene. At this point the tissue was infiltrated by histological wax and embedded in paraffin blocks. Finally the specimens were cut in 5 μ sections and prepared by hematoxylin and eosin (H&E) staining.

Microscopically in the left tonsil two large cystic spaces (Fig. 1A) and in the right tonsil multiple smaller cysts were seen (Fig. 1B). The cysts were lined by parakeratinized stratified squamous epithelium without rete ridges (Fig. 1C). The cysts lumen was filled by desquamated epithelial cells. A dense lymphocytic infiltration was observed surrounding the cyst epithelium with formation of germinal center in some areas. The diagnosis of lymphoepithelial cyst was established histologically.

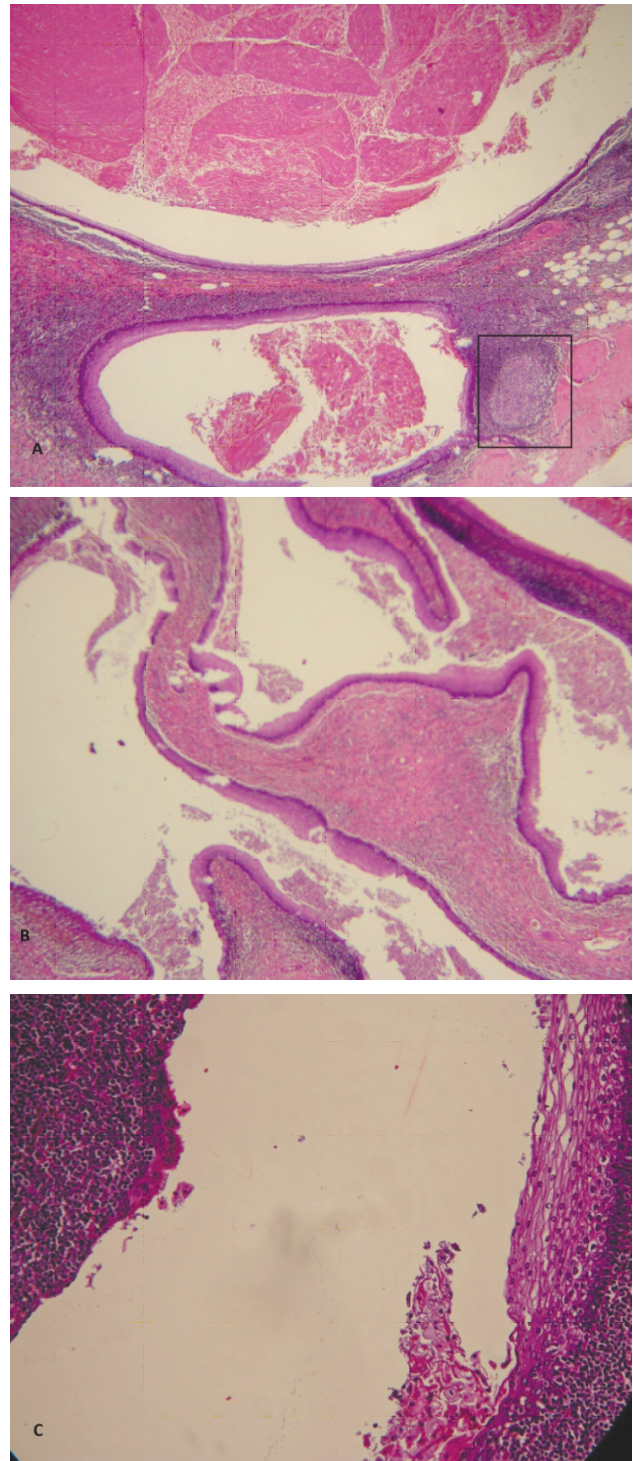


Fig .1- Histological aspect in hematoxylin-eosin stain. (A) Left tonsil with 2 large cystic spaces. The cyst wall contains lymphoid tissue with occasional germinal center formation (highlighted area) ($\times 100$); (B) Right tonsil with multiple cystic spaces ($\times 100$); (C) cyst lining is parakeratinized stratified squamous epithelium and is supported by a dense lymphocytic infiltration ($\times 400$).

Discussion

Bhaskar and Bernier first recommended the term LEC in 1959 (9, 10). Gold and Lewittown described the first oral LEC in 1962 (11, 12). A lymphoepithelial cyst may occur in virtually any organ. After the neck and oral cavity the most frequent sites reported in the literature are pancreas and thyroid gland (12).

Oral lesions are thought to arise from entrapment of epithelium within lymph node or lymphoid tissues and subsequent epithelial proliferation results in clinically evident mass (13). It is also possible that they may develop from surface mucosal epithelium that becomes enclaved in lymphoid tissue during embryogenesis (1,14, 15).

Knapp believes the lymphoepithelial cysts found in the oral tonsils were not true cysts but pseudocysts. He proposed that pseudocysts develop when the tonsillar crypt opening becomes plugged, which results in enlargement of the tonsillar tissue secondary to an accumulation of purulent materials or desquamated cells and keratins (16). Pancreatic lesions are believed to be epithelial inclusions embedded within the pancreas during embryogenesis (17) and cervical LEC develop from remnants of the branchial clefts (5, 18) as they are known as Branchial Cleft Cysts. Viral infections like EBV have been suggested to play a role in pathogenesis of parotid LECs in HIV positive patients (19).

Histologically LEC demonstrates a cystic cavity that is lined by parakeratinized stratified squamous epithelium without rete ridges. In rare instances in the oral LEC the epithelial lining also may contain mucous cells and some cervical LECs demonstrate respiratory epithelium. The most striking feature is the presence of lymphoid tissue in the cyst wall with occasional germinal

center formation (20).

Clinically they present as a small submucosal mass, usually less than 1 cm in diameter ranging from 3 to 15 mm (12, 21). They are asymptomatic and often detected during dental practice (22). The site most commonly affected is floor of the mouth where approximately 50% to 60% of the cases are found. The ventral and posteriolateral portion of the tongue constitute an additional 40% of cases (10, 13, 21).

In the present case the largest cystic space was 1.2 cm in diameter on gross examination and the patient reveals symptoms including pain fever and obstructive sleep apnea (OSA) that might be related to the location of the lesion that caused tonsillar enlargement. Except for one case of unilateral involvement of left palatine tonsil reported by Choi (8) we could not find any bilateral or multiple LECs of palatin tonsils in the English literature and the present case seems to be the first report.

The ages of the patients ranged from 4 to 81 years with a mean age of 32-34 (23, 24). In the oral cavity they present as solitary lesions but Friedrich reported an unusual case of bilateral intraosseous LEC of the mandible (25). Multiple benign lymphoepithelial cysts of parotid occur in patients with HIV infection generally as one of the signs of this infection (3-7).

Choi *et al.* reported bilateral LECs in thyroid gland (26). Scarce reports mention the coexistence of oral LECs with entities such as the epidermoid cyst (27), geographic tongue (28) and epithelial inclusion cyst (1). Costa reported simultaneous occurrence of a lymphoepithelial cyst and squamous cell carcinoma in the oral cavity (29). Table 1 shows cases of oral LECs reported in the literature since the first report by Gold in 1962 (11).

Table 1: Oral lymphoepithelial cyst cases reported in the literature

Number of the cases	Location	Gender	Age (yr)	Reference
1	Floor of the mouth	M	42	2
1	Palatine Tonsil	F	30	8
1	Tongue	M	30	10
1	Floor of the mouth	M	32	11
1	Tongue	F	28	11
2	Floor of the mouth (n=1), Tongue (n=1)	F=2	56 (55-57)	12
24	Floor of the mouth (n=15), tongue (n=8), palate (n=1)	M=17 F=7	40 (15-65)	14
1	Floor of the mouth	M	30	15
24	Floor of the mouth	M=9 F=15	43 (12-74)	20
1	Tongue	F	19	22
21	Floor of the mouth (n=17), palate (n=2), Buccal mucosa (n=1), Retromolar area (n=1)	M=9 F=12	36 (7- 65)	23
13	Floor of the mouth (n=7), tongue (n=2), palate (n=4)	M=12 F=1	32 (17- 48)	24
9	Floor of the mouth	M=9	33 (20- 46)	24
2	Bilateral Intramandibular (n=1)	F	29	25
1	Tongue	F	46	28
1	Floor of the mouth	M	40	30
38	Floor of the mouth (n=19), Tongue (n=14), palate (n=4), Retromolar area (n=1)	M=23 F=15	43 (14-81)	31
6	Floor of the mouth (n=2), Tongue (n=3), palate (n=1)	M=3 F=3	43 (25-60)	32
1	Floor of the mouth	M	34	33
1	Floor of the mouth	M	5	34
1	Floor of the mouth	M	29	35
1	Tongue	M	4	36
1	Tongue	M	72	37
1	Tongue	F	28	38
1	Floor of the mouth	M	34	39
9	Floor of the mouth (n=2), Tongue(n=4), Palate (n=1), Oropharynx (n=2)	M=2 F=8	38 (16-60)	40

F: Female; M: Male

Adenotonsillar enlargement is the main cause of obstructive sleep apnea (OSA) in the paediatric population. However, this prevalent syndrome is more complicated in adults (41). The morbidity of OSA includes hypertension, arrhythmia, heart disease, erythrocytosis, and hyperlipidemia (42). Obstructive sleep apnea has also been described in cases of benign lymphoid hyperplasia, plasmacytoma, amyloidosis, pharyngeal tumours and diseases that involve the nasopharyngeal structures (43). Obstructive sleep apnea is a common disorder effecting overweight/obese adults. Non-obese adults with obstructive sleep apnea should be carefully assessed for evidence of an anatomic limitation of the upper airway lumen that might result from various craniofacial anomalies, macroglossia and adenoid tonsillar hypertrophy (42). Tonsillar hypertrophy is a common remedial risk factor for Obstructive sleep apnea in the pediatric population, but may also play a role in the pathophysiology of sleep-related upper airway obstruction in adults.

In anterior floor of the mouth salivary lesions like sialolith may be similar in appearance to a lymphoepithelial cyst. However a history of pain and swelling of the associated salivary gland would be expected with a salivary duct stone (12). Developmental anomalies such as teratomas or dermoid cyst, benign mesenchymal neoplasms and salivary gland tumors might also be considered in differential diagnosis for a floor of the mouth soft tissue mass (13). When it involves the parotid gland, a lymphoepithelial cyst must be distinguished from salivary lymphoma, Warthin's tumor and low grade mucoepidermoid carcinoma. Conservative excisional biopsy is generally used for diagnosis as well as for treatment. Recurrence is not expected (12). We suggest that although lymphoepithelial cysts rarely occur in the tonsils, they should be considered in differential diagnosis of adenotonsillar enlargement and

related obstructive sleep apnea.

Acknowledgment

The authors declare that there is no conflict of interests.

References

1. Ahn SK, Won JH, Lee SH, Choi EH, Choi S. Lymphoepithelial cyst associated with epithelial inclusion cyst. *Am J Dermatopathol* 1996; 18(4):424–6.
2. Young WG, Claman SM. A lymphoepithelial cyst of the oral cavity. *Oral Surg Oral Med Oral Pathol* 1967; 23(1):62-70.
3. Finfer MD, Schinella RA, Rothstein SG. Cystic parotid lesions in patients at risk for AIDS. *Arch Otolaryngol Head Neck Surg* 1988; 114(11):1290-4.
4. Kumar VV. Parotid Lymphoepithelial Cysts as an Indicator of HIV Infection. *J Can Dent Assoc* 2011; 77:28.
5. Vargas PA, Villalba H, Passos AP, Saldiva PH, Mauad T, Caiaffa Filho HH, *et al.* Simultaneous occurrence of lymphoepithelial cysts, cytomegalovirus and mycobacterial infections in the intraparotid lymph nodes of a patient with AIDS. *J Oral Pathol Med* 2001; 30(8):507-9.
6. Favia G, Capodiferro S, Scivetti M, Lacaita MG, Filosa A, Lo Muzio L. Multiple parotid lymphoepithelial cysts in patients with HIV-infection: report of two cases. *Oral Dis* 2004; 10(3):151-4.
7. Ferreira Alves CA, Junior OR, Borba AM, Cantanhede S, de Souza OM, da Garcia Naclerio-Homem M. Parotid lymphoepithelial cyst in non-HIV patient. *Clin Exp Dent* 2011; 3 Suppl 1:400-3.
8. Choi J, Kim K, Kim B. Lymphoepithelial cyst of the palatine tonsil. *Ear, Nose Throat J* 2010; 89:584-5.
9. Bhaskar SN, Bernier JL. Histogenesis of branchial cysts: A report of 468 cases. *Am J Pathol* 1959; 35(2):407–43.
10. Suzuki H, Baba S, Hashimoto K. Lymphoepithelial cyst in the sublingual region: Report of a case and review of literature. *Oral Med Pathol* 2000; 5(2):105–8.

11. Gold C, Lewittown NJ. Branchial cleft cyst located in the floor of the mouth. *Oral Surg Oral Med Oral Pathol* 1962; 15(9):1118-20.
12. Stramandinoli-Zanicotti RT, de Castro Ávila LF, de Azevedo Izidoro AC, Izidoro FA, Schussel JL. Lymphoepithelial cysts of oral mucosa: two cases in different regions. *Bull Tokyo Dent Coll* 2012; 53(1):17-22.
13. Hong SS, Ogawa Y, Yagi T, Wakasa T, Sakurai M, Sato M, *et al.* Benign lymphoepithelial lesions with large cysts. *Oral Surg Oral Med Oral Pathol* 1990; 19(6):226-70.
14. Bhaskar SN, Colonel L. Lymphoepithelial cysts of oral cavity; report of twenty four cases. *Oral Surg* 1966; 21(1):120-8.
15. Vickers RA, Von Der Muhll O. An investigation concerning inducibility of Lymphoepithelial cysts in Hamsters by autogeneous epithelial transplantation. *J Dent Res* 1966; 45(4):1029-32.
16. Knapp MJ. Pathology of oral tonsils. *Oral Surg Oral Med Oral Pathol* 1970; 29(2):295-304.
17. Younus S, Bleibel W, Bleibel H, Hernady N. Lymphoepithelial cyst of the pancreas. *Dig Dis Sci* 2007;52(11):3136-9.
18. Little JW, Rickles NH. The histogenesis of the branchial cyst. *AM J Pathol* 1967; 50(5): 533-47.
19. Yen TL, Murr AH, Rabin J, Mhatre AN, Lalwani AK. Role of cytomegalovirus, Epstein-Barr virus, and human herpes virus-8 in benign lymphoepithelial cysts of the parotid gland. *Laryngoscope* 2004; 114(8):1500-5.
20. Chaudhry AP, Yamane GM, Scharlock SE, Sunder R, M, Jain R. A clinico-pathological study of intraoral lymphoepithelial cysts. *J Oral Med* 1984; 39(2):79-84.
21. Merchant NE. Lymphoepithelial cyst of the floor of the mouth: a case report. *Br Dent J* 1972; 132(7):271-2.
22. Sakoda S, Kodama Y, Shiba R. Lymphoepithelial cyst of Oral cavity. *Int J Oral Surg* 1983; 12(2):127-31.
23. Guinta J, Gataldo E. Lymphoepithelial cysts of oral mucosa. *Oral Surg Oral Med Oral Pathol* 1973; 35(1):77-84.
24. Acevedo A, Nelson JF. Lymphoepithelial cyst of Oral cavity: report of nine cases. *Oral Surg Oral Med Oral Pathol* 1971; 31(5):632-6.
25. Friedrich RE, Scheuer HA, Fuhrmann A. Bilateral intraosseous Lymphoepithelial cyst of the mandible. *Invivo* 2011; 25(3):451-4.
26. Choi CJ, Choi SW, Cho JG, Woo JS. Bilateral lymphoepithelial cysts of the thyroid gland. *Thyroid* 2010; 20(1):111-13.
27. Epivatianos A, Zaraboukas T, Antoniadis D. Coexistence of lymphoepithelial and epidermoid cysts on the floor of the mouth: report of a case. *Oral Dis* 2005; 11(5): 330-3.
28. Pereira KM, Nonaka CF, Santos PP, Medeiros AM, Galvão HC. Unusual coexistence of oral lymphoepithelial cyst and benign migratory glossitis. *Braz J Otorhinolaryngol* 2009; 75(2):318.
29. Costa FW, Pereira KM, Viana TS, Cavalcante RB, Nogueira AS. Simultaneous occurrence of a rare lymphoepithelial cyst and squamous cell carcinoma in the oral cavity. *Braz J Otorhinolaryngol* 2011; 77(2):270.
30. Calman HI. Sublingual branchiogenic cyst-report of a case. *J Oral Surg* 1963; 16(3):333-8.
31. Buchner A, Hansen LS. Lymphoepithelial cysts of the oral cavity. *Oral Surg* 1980; 50(5):441-9.
32. Toto PD, Wortel JP, Joseph G. Lymphoepithelial cysts and associated immunoglobulins. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1982; 54(1):59-65.
33. Iwase T, Teratani K, Saito A, Funatsu K, Sato M, Kiuchi K, Umemura S. Immunohistochemical and lectin histochemical studies on the lymphoepithelial cyst of the oral cavity and neck. *J Nihon Univ Sch Dent* 1985; 27(1):28-34.
34. McDonnell D. Spontaneous regression of a yellow sublingual swelling: a case report. *Pediatr Dent* 1990; 12(6):388-9.
35. Kumara GR, Gillgrass TJ, Bridgman JB. A lymphoepithelial cyst (branchial cyst) in the floor of the mouth. *N Z Dent J* 1995; 91(403):14-5.
36. Flaitz CM. Oral lymphoepithelial cyst in a young child. *Pediatr Dent* 2000; 22(5):422-3.
37. Flaitz CM, Davis SE. Oral and maxillofacial

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- pathology case of the mouth: oral lymphoepithelial cyst. *Tex Dent J* 2004; 121(7):624–31.
38. López-Jornet P. Oral lymphoepithelial cyst. *Ann Dermatol Venereol* 2007;134(6-7):588.
39. Khelemsky R, Mandel L. Lymphoepithelial cyst of mouth floor. *J Oral Maxillofac Surg* 2010; 68(12):3055–7.
40. Nonaka CF, Henriques AC, de Matos FR, de Souza LB, Pinto LP. Nonodontogenic cysts of the oral and maxillofacial region: demographic profile in a Brazilian population over a 40-year period. *Eur Arch Otorhinolaryngol* 2011;268(6):917–22.
41. Arens R, Mc Donough JM, Costarino AT, Mahboubi S, Tayag-Kier CE, Maislin G, Schwab RJ, Pack AI. Magnetic resonance imaging of the upper airway structure of children with obstructive sleep apnea syndrome. *Am J Respir Crit Care Med* 2001; 164(4):698-703.
42. Tsou YA, Cheng YK, Lin CD, Chang WC, Tsai MH. Small B cell lymphocytic lymphoma presenting as obstructive sleep apnea. *World J Surg Oncol* 2004; 2(1):26.
43. Fidan H, Fidan F, Unlu N, Ela Y, Ibis A, Tetik L. Prevalence of sleep apnea in patients undergoing operation. *Sleep and Breathing* 2006; 10 (3):161-5.