CASE REPORT

Lymphangioma Circumscriptum: Case Reports with Review of Literature

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Abstract

Lymphangiomas are congenital hyperplastic malformations of lymphatic vessels, which are benign in nature. Age wise, lymphangiomas have predilection for early childhood. Most common location is head and neck region, majority being superficial in location, although some may extend into deeper planes. Oral cavity is comparatively less common site, out of which anterior two third of tongue is most favored site. Macroscopic appearance together with histopathological examination of biopsied sample helps in diagnosis. Most common modality of treatment is surgical excision, although recurrence is common. Prognosis is usually good in majority of the cases, except where tumors invade vital structures or large tumors obstructing airway. Here we report two cases of intraoral lymphangiomas with review of literature.

Keywords: *Lymphangioma, Lymphatic Vessel.*

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Introduction

Lymphangiomas are benign hamartomatous malformations of sequestered lymphatic vessels in which superficial lymphatics do not communicate with deeper lymphatics. Most common sites are lips, tongue, palate and cheeks. Usually they are superficial in location resembling frog eggs appearance containing clear fluid. [1] Patients tend to have speech disturbances, poor oral hygiene and bleeding from tongue associated with oral trauma.[2] Treatment is usually surgical excision. Here we report two cases of oral lymphangiomas with review of literature.

Case Report -1

A 22 year old female patient came with lesion over anterior one third of tongue since 5 years of age. The lesion gradually increased in size and number over the period of time. There was history of oozing clear fluid with no history of macroglossia, bleeding and halitosis. There was no history of sudden increase in size of the lesions. Family history was absent. Systemic and general examination was within normal limits. Histopathological examination of biopsied lesion showed copious lymphatic vessels disseminated inside the stroma (Fig.1,2).

Case Report -2

A 5 year old female came to OPD with chief complaint of asymptomatic fluid filled lesions over anterior part of tip of tongue and over ventral surface since last 3 years. The lesion gradually increased in size and number over period of time. There was no history of slurred speech, macroglossia, halitosis associated with lesions. Cervical lymphadenopathy was absent. In biopsy specimen, multiple lymphatic vessels lined by endothelium with rare extravagated red blood cells observed. (Fig.3, 4).

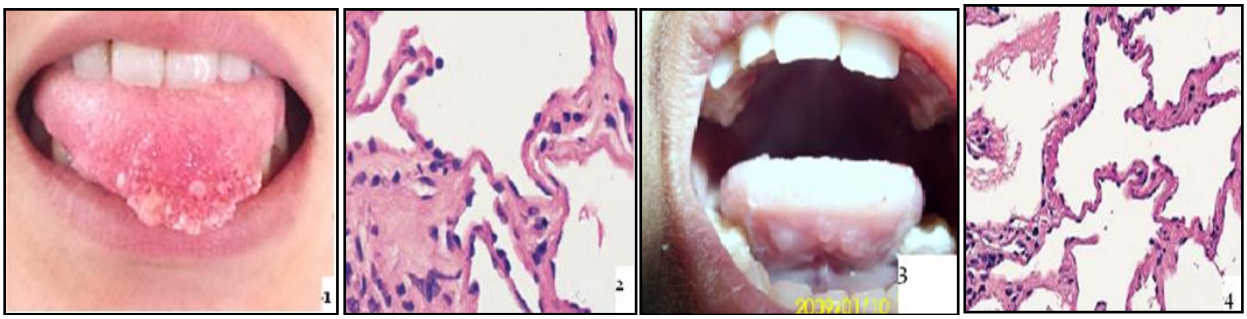

***Fig 1**: Ill-defined lobulated swelling consisting of tense vesicles with macerated walls*
***Fig 2**: Enlarged endothelial lined lymphatic vessels filled with lymph [H and E stain]*
***Fig 3**: Vesicles resembling frog eggs appearance.*
***Fig 4**: Enlarged lymphatic vessels lined with endothelium and filled with lymph*

Discussion

Lymphangiomas are the lymphatic analogue of the hemangioma of the blood vessels, which is classified into three types:

1. lymphangioma simplex/capillary lymphangioma which consist of small capillary sized vessels,
2. cavernous lymphangiomas composed of larger dilated lymphatic vessels,
3. cystic lymphangiomas/cystic hygroma, which exhibits large macroscopic cystic spaces.

All above mentioned sizes of vessels can be found within the same lesion and its size may depend upon the nature of neighboring tissues. Lymphangiomas are sequestered portions of lymphatic system that hold the knack of producing lymph and increase in volume of lymph inside it leads to larger growth in the contiguous tissue.[3,4] Most common location of cystic lymphangioma is in neck and axilla, cavernous lymphangioma in mouth. Rarely, however cystic variety can be found in the trunk within the internal organs or the connective tissue in and around the abdominal or thoracic cavities and cavernous type can be found rarely in the retroperitoneum. Amongst the cervical lymphangiomas, posterior triangle is most favored site and this lesions are stereotypically soft and fluctuant.[5,6] They occur less frequently in the anterior triangle, but more likely to result in respiratory difficulties or dysphagia if they grow large. Rarely cervical Lymphangiomas extending into the mediastinum or oral cavity may measure up to or greater than 15 cm.[4] De Seress has proposed classification of head and neck lymphangiomas according to supra and infrahyoid spread and uni/bilaterality of lesions.[7] It has been found in various studies that relapse was more common in suprahyoid as compared to infrahyoid lesions.[8,9] About half of all lesions are noted at birth, and around 90% develop fully by 2 years of age. [4] Lymphangiomas are of rare occurrence in the oral cavity, out of which anterior two thirds of tongue is most common site, followed by palate, gingiva, oral mucosa, lips and alveolar ridge of the mandible. [10] In the anterior location of tongue, it may cause macroglossia.[11,12] Brisk tumor enlargement may occur due to upper respiratory infections owing to:

1. Increased Lymph Production
2. Blocked Lymphatic Drainage
3. Secondary Infection of the tumor.

They are mostly superficial in location, where it shows collection of translucent vesicles giving facade of frog eggs or tapioca pudding.[6] Occasionally purple discoloration may occur in the vesicles indicating secondary hemorrhage into the lymphatic spaces. Deeper tumors are soft, ill-defined.[4] Histopathological examination of biopsied lesions show lymphatic vessels with marked dilation (cavernous lymphangioma) or macroscopic cystoids (cystic hygroma). Most often, walls of adjacent soft tissue show lymphoid aggregates owing to its infiltration by lymphatic vessels and lining endothelium of these vessels are classically thin with occasional lymphocytes and protienaceous fluid.[4,13] The finding of red blood cells in some vessels raise a doubt, whether they are blood vessel or lymphatic vessel. Also purple discoloration may be misleading as to, whether

it is secondary hemorrhage into the lymphangioma or mixed lesion of lymphangioma and hemangioma. In case of oral lesions, subepithelial location and replacement of connective tissue papillae are characteristic features.[5] Usual modality of treatment is surgical excision, although recurrence is the main issue, particularly common in cavernous type and least common in cystic lymphangioma. Total excision is not always possible as in case of large tumors and tumors involving vital structures. Due to above observations, most of the surgeons do not opt for excision of non-enlarging lymphangiomas of tongue. Spontaneous regression is very rare. Regrettably, lymphangiomas do not respond to sclerosing agents as do hemangiomas. Although recently OK-432, a sclerosing agent showed some success in unresectable tumors. OK-432 is a lyophilized incubation mixture of streptococcus pyogenes of low virulence, which has lost its capacity to produce streptolysin-O with penicillin G potassium. Prognosis is good in majority of the patients, except in small fraction of the patients with large tumors in neck or tongue which may result in airway obstruction.[4] Mortality rates for cystic hygroma patients is 2-5%. A few cases of squamous cell carcinoma arising from lymphangioma circumscriptum have been reported.[14]

Conclusion

Albeit its erratic incidence in the oral cavity, lymphangiomas need to be diagnosed early so that it can be treated at a stage, where almost complete removal can be possible without posing much threat of complications.

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References

1. Irvine AD, Sweeney L, Corbett JR. Lymphangioma circumscriptum associated with paravesicle cystic retro peritoneal lymphangioma. Br J Dermatol 1996;134:1135-37. [CrossRef] [PubMed]
2. Whimster I. Pathology Lymphangioma Circumscriptum.Br J Dermatol 1976;94:476-86. [CrossRef]
3. Courtney M, Tocoushed J. Sabiston's Textbook of Surgery. In editors: Iraklis I, Pipinos B, Timothy Baxter. The Lymphatics, Elsevier 18th ed vol 2 ;2007:2026-27.
4. Neville BW, Damm DD, Allen CM. Oral and Maxillofacial Pathology, Saunders Elsevier, 3rd ed;2009:547-49.
5. Rajendran R, Sivapathasundaram. Shafer's Textbook of Oral Pathology, Elsevier, 6th ed;2009:146-8.
6. Greenberg, Glick, Ship. Burket's Oral Medicine, BC Decker, 11th ed;2008:139-40.
7. De Serres LM, Sie KC, Richardson MA. Lymphatic malformation of the head and neck: a proposal for staging, Arch Otolaryngol Head Neck Surg 195;121:577-582. [CrossRef] [PubMed]
8. Ricciardelli LJ, Richardson MA. Cervicofacial cystic hygroma patterns of recurrence and management of the difficult case. Arch Otolaryngol Head Neck Surg 1991;117:546-553. [CrossRef] [PubMed]
9. Hartl DM, Roger G, Denoyelle F. Extensive lymphangioma presenting with upper airway obstruction. Arch Otolaryngol Head Neck Surg 2000; 126:1378-82. [CrossRef] [PubMed]
10. Jamaroon A, Pongsiriwet S, Srisuwan S, Krisanaprakornkit S. Lymphangiomas of the tongue. Int J Pediatr Dent, 2003;13:62-63. [CrossRef] [PubMed]
11. Guelmann M, Katz J. Macroglossia combined with lymphangioma a case report. J Cli Pediatr Dent 2003;27:167-70. [CrossRef] [PubMed]
12. Karmody CS, Fortson JK, Calcaterra VE. Lymphangiomas of head and neck in adult. Otolaryngol Head Neck Surg 1982;90(3.1):283-88. [PubMed]
13. Jeeva Rathan J, Harshavardhan BG, Muthu MS, Venkatachalapathy et al. Oral lymphangioma: A case report. J Indian Soc Pedod Prev Dent 2005;23:185-9. [CrossRef] [PubMed]
14. Wilson GR, Cox NH, Mclean NR, Scott D. Squamous cell carcinoma arising within congenital lymphangioma circumscriptum, Br J Dermatol, 1993;129(3): 337-339. [CrossRef] [PubMed]