Case Report

DOI: 10.7860/JCDR/2023/59451.17646

Lymphangioma Circumscripta- A Diagnostic Dilemma

Dentistry Section

PRIYANKA RASTOGI[1], SACHIN KUMAR[2], DIPEN MAJUMDER[3], RUDRA BHARDWAJ[4], DEVESHI NIGAM[5]

ABSTRACT

Lymphangiomas are developmental malformations that present as benign hamartomas of lymphatic channels. They present at birth or before two years of age, with 50-70% predilection for the head and neck region and less frequently reported in the oral cavity. However, affected sites in the oral cavity include the tongue, palate, gingiva, buccal mucosa, lips and alveolar ridge. Lymphangioma of the buccal mucosa is rare, only 27 cases of lymphangioma of buccal mucosa have been reported till date, out of which seven were in children. The prognosis is good for most patients, but recurrence has also been reported in some cases. This present case is of Lymphangioma Circumscriptum in a 13-year-old female patient involving right buccal mucosa and lip since birth. This lesion was surgically removed under general anaesthesia and there was a recurrence of the lesion after one year of follow-up. In the literature there was no reported case of Lymphangioma Circumscriptum involving the lip of a child, so this present case was the first reported case of Lymphangioma Circumscripta involving the lip in a child with recurrence.

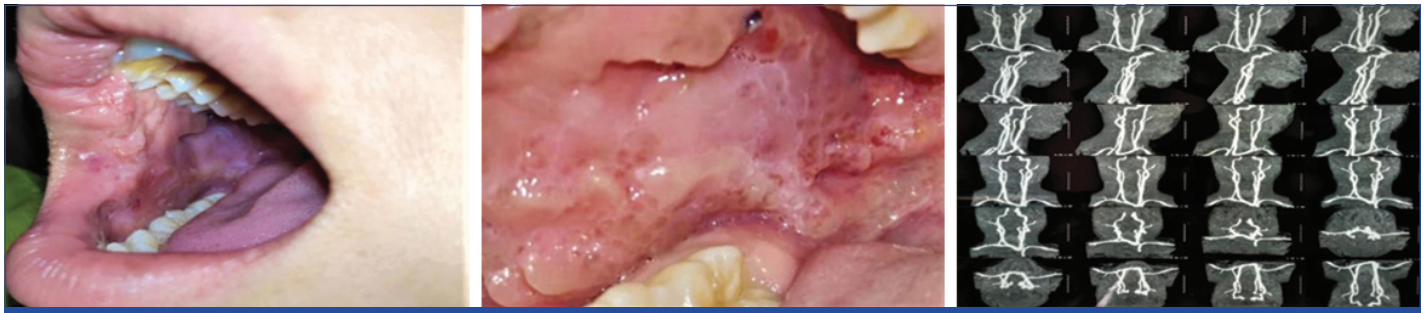
Keywords: Amyloidosis, Hamartoma, Hecks disease, Malformation

CASE REPORT

A 13-year-old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of multiple growth on the right-side of the inner cheek and upper lip region since birth. These growths gradually increased in size and reached the present size. The patient underwent surgery, for the excision of growths in Bareilly, five years back but recurrence was observed after 1-2 years of surgery and she visited to the institution for the same. Past medical history and natal/neonatal history was non significant.

Intraoral examination of right buccal mucosa and lip showed multiple papular appearances, with colour ranging from translucent to yellow-reddish and soft in consistency [Table/Fig-1,2]. Measuring about >2 cm extending antero-posteriorly 1 mm away from the vermillion border of lip to retromolar area and upper alveolar sulcus to lower alveolar sulcus superio-inferiorly on the right-side of buccal mucosa. CT neck Angiography was done which showed asymmetric thickening of buccal mucosa on right-side, measuring approx. 7.4×34 mm, with mild stranding of overlying subcutaneous fat seen with no bone involvement, no calcification and no involvement of adjacent muscles of mastication [Table/Fig-3]. On correlating the history, clinical and radiographical features provisional diagnosis of Focal Epithelial Hyperplasia or 'Hecks Disease' was given. Amyloidosis was also considered as a differential diagnosis. A written informed consent was taken from the patient.

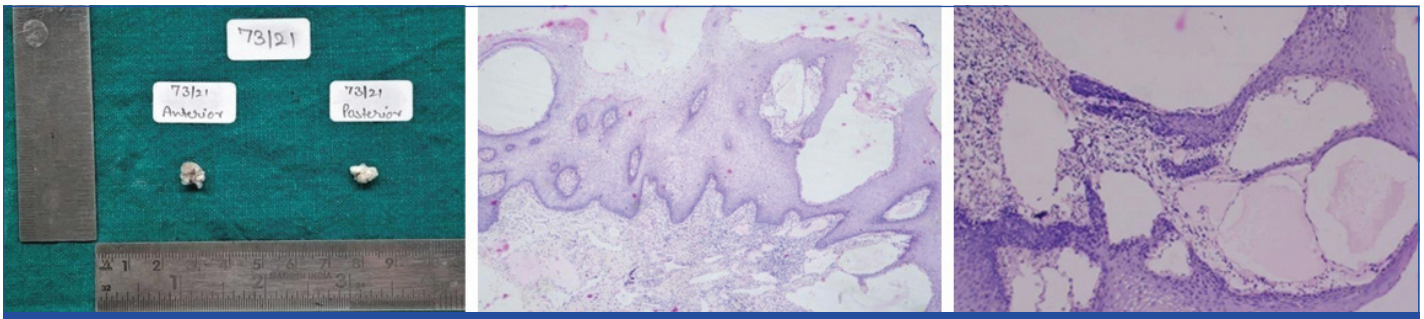
Routine haematological examination and exfoliative cytology were performed prior to incisional biopsy. The haematological findings were within normal range. Exfoliative cytosmear showed Class II cytosmear with mild cellular atypia. An incisional biopsy was performed under local anaesthesia from two sites, one from anterior portion of right buccal mucosa and another from retromolar area. Two bits of soft tissue specimens were fixed in 10% buffered formalin which were creamy white in colour, firm to leathery in consistency measuring in length×breadth×height as follows- Anterior soft tissue bit- 8×7×6 mm, Posterior soft tissue bit- 9×6×5 mm [Table/Fig-4]. The light microscopic view of Haematoxylin and Eosin (H&E) stained soft tissue sections showed hyperplastic parakeratinised stratified squamous epithelium overlying a fibrocellular connective tissue stroma. The epithelium showed numerous intraepidermal dilated endothelial lined lymphatic vessels. The connective tissue stroma showed diffusely distributed dilated endothelial lined lymphatic channels containing eosinophilic lymph, adjacent to overlying epithelium. The connective tissue showed diffuse infiltration of chronic inflammatory cell predominantly consisting of lymphocytes. Deeper connective tissue stroma showed the presence of muscle bundles and minor salivary glands [Table/Fig-5,6]. On the basis of Histopathological findings, final diagnosis of "Lymphangioma Circumscripta" was made. So, complete surgical excision was planned and performed under general anaesthesia followed by reconstruction of buccal fat pad and collagen membrane [Table/Fig-7,8]. Patient was prescribed Intravenous (i.v.) injection of Amoxicillin 1000 mg with Clavulanic acid 200 mg, Metronidazole 100 mL infusion, Pantoprazole 40 mg and Diclofenac Sodium AQIM 75 mg for three days, followed by oral medication of same drugs for four more days. Postoperative healing was uneventful, on six months of follow-up there was no sign of recurrence but after



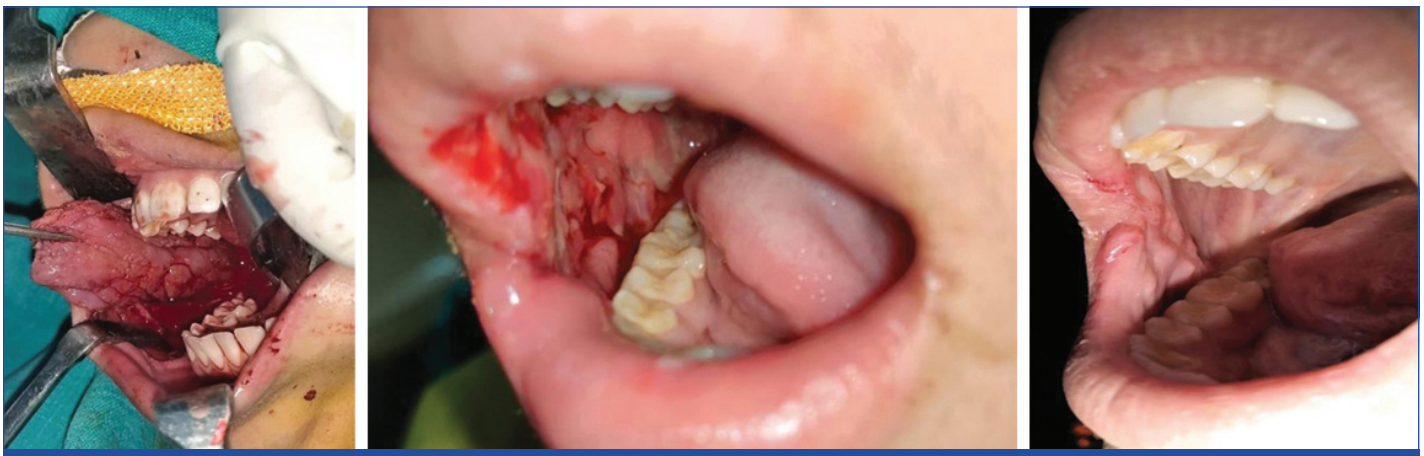
[Table/Fig-1]: Clinical photograph showing papular growth on right buccal mucosa involving upper lip.
[Table/Fig-2]: Clinical photograph showing frog egg or tapioca-pudding appearance.
[Table/Fig-3]: CT neck Angiography showing no bone involvement, no calcification and no involvement of adjacent muscles of mastication. (Images from left to right)

one year of follow-up the lesion showed recurrence on right buccal mucosa [Table/Fig-9]. Patient is under observation and the surgery is planned but the patient is deferring the surgery.

[1,4-20]. There is no sign of racial predominance and equal gender predilection reported in most of the studies [2]. Regarding the development of Lymphangioma, it is hypothesised that it occurs



[Table/Fig-4]: Incised tissue specimens from anterior and posterior areas of the lesion.
[Table/Fig-5]: Photomicrograph of the section showing dilated endothelial lined lymphatic vessels within the epithelium (H&E stain, x40).
[Table/Fig-6]: Photomicrograph of the section showing dilated endothelial lined lymphatic vessels within the connective tissue (H&E stain, x100). (Images from left to right)



[Table/Fig-7]: Complete surgical removal was done under general anaesthesia.
[Table/Fig-8]: Reconstruction of buccal fat pad and collagen membrane.
[Table/Fig-9]: Follow-up after one year of surgery with recurrence. (Images from left to right)

DISCUSSION

Lymphangiomas are benign developmental malformations, present as hamartomas of lymphatic vessels [1]. Lymphangiomas are rare, they account for 4% of all the vascular tumours and 25% of all benign vascular tumours in children [2]. Lymphangiomas, first described by Redenbacher in 1828 and the incidence ranges from 1.2 to 2.8/1000 newborns. Approximately, 50% are present at birth and 90% are diagnosed before the age of 2 years, with (50%-70%) predilection for head and neck. Most common site for Lymphangioma is neck and axillae. Involvement of oral cavity is rare, which commonly involves the tongue followed by palate, buccal mucosa, gingiva, floor of mouth and lip [3]. Only 27 cases of lymphangioma of buccal mucosa have been reported till date, out of which seven were in children as per availability of data [Table/Fig-10] due to proliferation of congenitally obstructed or remnants of the primitive lymphatic cells, commonly capable of accumulating fluids, which leads to a cystic presentation. These entrapped cells do not merge with any large lymphatic vessels. Hence they are responsible for accumulation of lymph at an abnormal site [13].

Clinically, oral lymphangioma are present at superficial surface which appear to be pebbly, vesicles-like lesion ranging from reddish or reddish-purple in colour, which are called "frog-egg" or "tapioca-pudding" appearance. On the basis of clinical presentation lymphangioma can be classified as macrocystic (site >2 cm^3), microcystic (size <2 cm^3) and mixed variants (combination of these two types) [1]. The differential diagnosis of oral lymphangioma includes haemangioma, teratoma, dermoid cyst, thyroglossal cyst, amyloidosis, neurofibromatosis, granular cell tumour, and pseudoepitheliomatous hyperplasia [2,21].

S. No.	Author's name and year	Place	Age and gender	Clinical presentation	Final diagnosis	Treatment and follow-up
1	Pereira JC et al., (1979) [4]	NA	NA/NA	Papular lesion on buccal mucosa	Lymphangioma of oral mucosa	NA
2	Tasar F et al., (1995) [5]	Turkey	10 years/Female	Swelling on buccal mucosa	Lymphangioma of buccal mucosa	Nd-YAG laser surgery/No recurrence
3	Brennan TD et al., (1997) [6]	Philadelphia	NA	NA	Lymphangioma of buccalmucosa (8 cases)	Surgical excision/recurrence after treatment
4	Harashima T et al., (2001)[7]	Japan	9 years/Female	Purple to red coloured vesicle-like lesion on buccal mucosa since 1 year	Lymphangioma	Nd-YAG laser surgery/No recurrence
5	Bozkaya S et al., (2006) [8]	Turkey	21 years/Male	Discolaration on right buccal mucosa with papillary projection since birth	Lymphangioma of right buccal mucosa	Radiofrequency tissue ablation/no recurrence
6	Domingo ST et al., (2008) [9]	Spain	NA (2 cases)	Blue, soft, irregular, smooth, elastic and sessile growth on buccal mucosa	Lymphangioma	NA
7	Dogan N (2010) [10]	Turkey	35 years/Female	Bluish-purple coloured fibrotic lesion on left inside cheek region since 18 months	Lymphangioma of left buccal mucosa	Surgical excision/recurrence after 2 months
8	Coskunses F et al., (2012) [11]	Turkey	47 years/Male	NA	Lymphangioma of buccal mucosa	NA

9	Haranal S et al., (2013) [12]	India	28 years/Female	Reddish discolouration with smooth surface since birth	Lymphangioma	Surgical excision
10	Yoganna SS et al., (2014) [13]	India	14 years/Male	Recurrence of pebble like swelling with dilated veins on left cheek region since 5 years	Lymphangioma	Surgical excision/No recurrence after second surgery
11	Kaur M et al., (2015) [14]	India	17 years/Male	Swelling on the right-side of the buccal mucosa with multiple small translucent papules since 13 years	Cavernous lymphangioma	Incisional biopsy/NA
12	Babu DBG et al., (2015) [1]	India	60 years/Male	Solitary well defined swelling in the left cheek region since 6 months	Lymphangioma Circumscripta of the left buccal mucosa	Surgical excision/No recurrence after 1 year of follow-up
13	Pammar C et al., (2015) [15]	India	45 years/Female	Single lobulated bluish red translucent lesion on the left buccal mucosa since 2 years	Lymphangioma on buccal mucosa	Surgical excision/No recurrence after 1 year of follow-up
14	Devi A et al., (2016) [16]	India	17 years/Females and 6 years/ Female	Erythematous transparent vesicles were present on left buccal mucosa involving the retromolar region	Lymphangioma	NA
15	Kolay SK et al., (2018) [17]	India	32 years/Female	Erythematous area and transparent vesicles are present on left buccal mucosa	Cavernous Lymphangioma	NA
16	Yalcin M (2019) [18]	Turkey	15 years/Female	Discolourisation was seen in left buccal mucosa, lesion was exophytic with colour ranging from reddish purple to yellow	Lymphangioma	Excised by electrocautery/ No recurrence after 1 year of follow-up
17	Tayefeh A (2020) [19]	Iran	50 years/Female	Nodule with pebbly surface on the left buccal mucosa extending to the retromolar region	Lymphangioma	NA
18	Senthilnathan N et al., (2021) [20]	India	52 years/Female	Multiple violaceous discoloured swelling present on left buccal mucosa	Lymphangioma	NA
19	Present Case	India	13 years/Female	multiple papular appearances of right buccal mucosa and lip, with colour ranging from translucent to yellow-reddish and soft in consistency	Lymphangioma Circumscripta	Surgical excision. Recurrence was seen after 1 year

[Table/Fig-10]: Details of total number of lymphangioma of buccal mucosa cases reported in the literature from 1979 to 2021 [1,4-20].

In this present case, the similar clinical presentation could have been easily misdiagnosed as an oral amyloidosis or Heck's disease and might have leads to diagnostic dilemma [22,23]. The oral manifestations of amyloidosis includes nodules, papules, plaques and enlargement of tongue, the colour of the lesion may vary from yellow, orange, red, blue and purple. Dissanayaka DWVN et al., reported a case of oral amyloidosis representing multiple ulcerative lesions on tongue and buccal mucosa [22]. Focal epithelial hyperplasia or Heck's disease commonly present in children and involve oral mucosa, lips, tongue, gingival and palate [23]. Ozden B et al., reported a case of Heck's disease on a seven-year-old girl representing soft, sessile papules and nodules on labial and buccal mucosa [24]. Devi A et al., reported similar lesion on buccal mucosa and retromolar region [16]. This was the first documented case reported of congenital Lymphangioma Circumscriptum of a 13-year-old child on the buccal mucosa involving the upper lip.

The microscopic features of lymphangioma circumscripta are dilated, single endothelial cells lined cystic spaces in the epithelium and connective tissue, which often contains red blood cells and eosinophlic lymph [8,10]. The immune-histochemical profile shows expression for CD31, D2-40 and endothelial cells are positive for Factor VIII immune-histochemical staining [13,17]. Study conducted by Brennan TD et al., showed type IV collagen were not completely encircled on basement membrane in lymphangioma cases but vascular channels in haemangioma were completely encircled [6]. In this present case, histological features showed dilated endothelial lined lymph vessels filled with eosinophelic lymph involving epithelium and connective tissue. Histopathologically, Lymphangioma can be classified into four main types: cavernous lymphangioma, cystic hygroma, lymphangioma circumscriptum and benign lymphangio-endothelioma [2].

Several contemporary treatment choices are available for lymphatic malformation such as: aspiration or drainage, radiotherapy, radiofrequency ablation, sclerotherapy, Laser surgery (Nd-YAG, CO_2 12), Bleomycin and pingyangmycin, cryotherapy, cryotherapy and surgical excision [2,19]. Aspiration or drainage are temporary treatment techniques, a wide-bore needle can be used to remove lesional fluid. These techniques might be used for reducing the size of the lesion but it is not a permanent treatment choice [8]. Now-a-days radiotherapy is not an acceptable treatment method because Nagata M et al., and Berry JA et al., have reported malignant transformation of Lymphangioma after radiotherapy [25,26]. Radiofrequency ablation can be used for localised superficial lesions to avoid deeper tissue fibrosis. Various sclerosing agents have been used for Lymphangioma such as hypertonic saline, 25% dextrose, sodium morrhuate, tetracycline, doxycycline, ethanol, bleomycin, cyclophosphamide and OK-432. Due to limited effect of sclerotherapy on macrocystic or mixed lymphangiomas, doxycycline seems to be an alternative agent. Widely used standard surgical method for Lymphangioma is Nd-YAG laser surgery due to less bleeding. Surgical removal of the lesion is best treatment option with inclusion of a surrounding normal tissue border without damaging the vital structure. Cryotherapy is performed by cryoprobe under extreme cold temperature. The best part of this technique is its good postoperative aesthetic results with less fibrous tissue [19]. Plasma knife surgery is a new technique for superficially located lymphangioma. The probe tip is used for thermal damage to the adjacent surrounding tissue without being heated [20]. Recurrence rate is about 15-53% due to its infiltrative nature. Cases reported by Brennan TD et al., Dogan N and Yoganna SS et al., showed recurrence of lymphangioma [6,10,13]. Tongue and pharynx are the most common sight for recurrent [10]. In this case report, there was sign of recurrence after a year of follow-up.

CONCLUSION(S)

Clinical diagnosis of Lymphangioma is quite challenging due to its variable clinical appearance. Based on the above case report it is concluded that there is difficulty in clinical diagnosis. So one should always prefer biopsy for histopathological evaluation to reach to the final diagnosis, in order to initiate the fast and appropriate treatment for better prognosis. Since, complete surgical excision

may not guarantee complete cure of the disease, as these may show recurrences, so long term follow-up is must after surgery.

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PARTICULARS OF CONTRIBUTORS:
1. Professor and Head, Department of Oral and Maxillofacial Pathology and Microbiology, Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India.
2. Professor, Department of Oral and Maxillofacial Pathology and Microbiology, Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India.
3. Postgraduate Student, Department of Oral and Maxillofacial Pathology and Microbiology, Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India.
4. Senior Lecturer, Department of Oral and Maxillofacial Pathology, Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India.
5. Postgraduate Student, Department of Paediatric and Preventive Dentistry, Kothiwal Dental College and Research Centre, Moradabad, Uttar Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Dipen Majumder,
Postgraduate Student, Department of Oral and Maxillofacial Pathology and Microbiology, Kothiwal Dental College and Research Centre,
Moradabad-244001, Uttar Pradesh, India.
E-mail: dipenmajumder482@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Aug 04, 2022
- Manual Googling: Aug 30, 2022
- iThenticate Software: Oct 19, 2022 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Aug 02, 2022**
Date of Peer Review: **Aug 31, 2022**
Date of Acceptance: **Oct 21, 2022**
Date of Publishing: **Mar 01, 2023**