Intraoral Lymphangioma in an Elderly Woman: A Case Report with Review
Priya Sahni, Reena Chaudhary, Avani Patel

Abstract
Lymphangioma is a benign proliferation of lymphatic vessels that occur as a focal superficial lesion within the oral cavity and as massive diffuse lesion of the neck (cystic hygroma). Most of the lymphangioma spontaneously involute during puberty. Those that fail to involute usually demonstrate a cessation of the growth and may be left untreated. Commonly located at head and neck, they rarely occur in the buccal mucosa. Intraoral lymphangioma occur more frequently on the dorsum of tongue. Here, we present an unusual case of a 55-year-old female patient with lymphangioma of the left buccal mucosa. The clinical, radiological, and treatment modalities of this case are discussed.

Keywords: Buccal mucosa; Cystic hygroma; Lymphangioma; Oral cavity.

Introduction
Lymphangiomatous lesions are rare congenital malformations of the lymphatic system that occur throughout the body with greater frequency in the cervico-facial area. It has also been called a malformation, a hamartoma, and an embryonic tumour. About 90% of these develop before the age of 2 years; and 50% of the lesions are noted at birth, although, there have been reports of its occurrence in adults. This was described first by Redenbacher in 1828 as a ranula congenita. In 1843 Werner described it as ‘cystic hygroma’, in greek hygroma means ‘hydro’= moist and ‘oma’= tumour. In 1855 Rokitansky said that a cystic hygroma was simply a collection of serious fluid held in place by surrounding tissue. Virchow in 1854 was the first to give accurate description of lymphangioma. In 1872 Krester hypothesized that hygroma was derived from lymphatic tissue.

Lymphangiomas are believed to arise from lymph sac sequestration and enlarge due to inadequate drainage, from lack of communication with the central lymphatic channels or excessive secretion of lining cells. The most common location in the mouth is the dorsum of tongue, involvement of which leads to macroglossia, followed by lips, buccal mucosa, soft palate, floor of the mouth. Although cases in gingiva, lingual mucosa and mandibular alveolar ridge have also been described. It affects both sexes equally. For lymphangiomas slow progressive enlargement is the rule, but rapid enlargement infrequently occurs secondary to infection or trauma. They are also known to be associated with Turner’s syndrome, Noonan’s syndrome, trisomies syndrome, cardiac anomalies, and fetal alcohol syndrome.

Three theories hypothesis the origin of this abnormality.
1. The blockage of normal growth of the primitive lymph channels occur during embryogenesis.
2. The primitive lymphatic sac does not reach the venous system and
3. During embryogenesis, lymphatic tissue was in a wrong location.

Recently, Norgall et al. suggested that Immunohistological Characterization of lymphatic endothelial cells suggests an involvement of VEGFR-3 and -2 in the aetiology of the lymphangioma. Upregulation of VEGFR-3 and -2 signalling are the major cause for the aberrant lymph vessel formation. Podoplanin, lymphatic vessels endothelial HA receptor-1 (LYVE-1), Prox 1 and D2-40 are the lymphatic endothelial cell markers for distinguishing blood vessels from lymphatic vessels.

The clinical course of this pathology varies from a regressing cyst to an aggressive invasion lesion. Spontaneous or traumatic haemorrhage are the most common complication encountered. Computed tomography, ultra sound and magnetic resonance imaging can be used as diagnostic aids to delineate the relationship of the lesion with the surrounding structures and to plan...
surgery. The present case is of an unusual presentation of Lymphangioma in buccal mucosa of an elderly female patient.

Case Report
A 55-year-old female patient reported with a chief complaint of pain in lower right back tooth region since five days. Pain was dull and continuous in nature and relieved by medication. There was no relevant family and medical history. On general physical examination, patient was moderately built and nourished with vital signs within normal limit. On extra oral examination there was no gross facial asymmetry and the mouth opening was within normal limit. On clinical examination, deep caries was noticed i.r.t. 46. Also a 1x2 cm oval shaped lesion was noticed on left buccal mucosa. The mucosal lesion was greyish purple in colour, with smooth surface. (Figure 1a) On palpation the lesion was nodular, firm to soft in consistency, non-tender, non-compressible and without any evidence of bleeding. The patient reported that she was unaware of the lesion as there were no symptoms associated with it. Panoramic radiograph depicted phlebolith and periapical abscess in relation to tooth #46. No evidence of adjacent bone involvement was noticed and on the basis of clinical appearance of the lesion on the buccal mucosa, a provisional diagnosis of vascular malformation was made.

A Complete blood examination was done, The lesion was surgically excised with clear borders and was sent for histopathological evaluation. (Figure 1b) The received tissue was cystic, greyish white in color, soft in consistency measuring about 0.8 to 0.6 cm in diameter. (Figure 1c & d) Histopathological examination of hematoxylin & eosin stained section show surface lining of parakeratinized stratified squamous epithelium. Sub epithelial connective tissue stroma shows enlarged vascular spaces. Vascular spaces were lined by flattened thin endothelial cells and comprised of RBCs and eosinophilic proteinases material and occasional lymphocytes. The deeper connective tissue stroma showed longitudinal and cross section of skeletal muscles along with adipose tissues. On the basis of histopathology, diagnosis of lymphangioma was established. (Figure 1e & f)

The patient was followed up for one year without any recurrence.

Discussion
Lymphangiomas are excessive vascular proliferation of lymphatic system. Most lymphatic vessel proliferation arise during childhood. There is no sex predilection. Two major types lymphangioms occur in the head and neck region. First being self-limiting tumours usually found in the oral mucosa and the second being cystic hygroma usually massive soft and fluctuant tumours found in the lateral neck region. Oral mucosal lymphangioma are most commonly found in the tongue of children, where they involute at puberty to attain a given size and persist without further growth. The lesions present on the dorsum of the tongue are often racemose with grape like clusters, reddish to yellowish in appearance and soft on palpation. The other areas involved may be lips, alveolar ridge of the mandible, palate, gingival and buccal mucosa. Cervical lymphatic malformations are common in the posterior triangle and soft and fluctuant in consistency. If present in the anterior triangle the lesion may result in respiratory difficulties or dysphagia if they grow large. Occasionally, the cervical lesions may extend in to the mediastinum or upward in to the oral cavity. Rapid enlargement may occur secondary to an upper respiratory tract infection because of increased lymph production, blocked lymphatic drainage or secondary infection of the tumour. Such lesions pose great difficulties in surgical removal.

According to Kennedy et al, Lymphangioma are classified based on their extent as:

1) Superficial cutaneous lymphangioma
   Lymphangioma simplex
   Lymphangioma circumscriptum

2) Cavernous lymphangioma
   Loose: Mucous membrane of lips, cheek, and floor of mouth
   Compact: Tongue, abdomen, or flank

3) Cystic hygroma—cystic lymphangioma

4) Diffuse systemic lymphangioma
   Lymphangioma—hemangioma

Lesions designated as acquired lymphangiectasia may develop as a result of infection or surgery that interferes with regional lymphatic drainage.

A classification of the lymphangioma of head and neck on the base of anatomical involvement had been proposed by De Serres LM (lymphatic malformation):
Stage/class I - infrathyroid unilateral lesions
Stage/class II - suprathyroid bilateral lesions
Stage/class III - suprathyroid or infrathyroid unilateral lesions
Stage/class IV - suprathyroid bilateral lesions
Stage/class V - suprathyroid or infrathyroid bilateral lesions
Stage/class IV - infrathyroid bilateral lesions

Lymphangioma do not show racial and sexual predilection and are often diagnosed in paediatric patients. Occurrence in adolescence and adulthood is reported but is frequently uncommon.

Figure 1: The clinical photograph of lesion on buccal mucosa (a). The surgically excised specimen (b) upon gross macroscopic examination showing a single bit of cystic tissue, greyish white in colour, soft in consistency measuring about 0.8 to 0.6 cm in diameter (c) was cut in to two halves showing blood entrapped in large cystic spaces (d). The photomicrograph of hematoxylin and eosin stained section under low (e) and high power view (f) shows large vascular spaces lined by endothelium just beneath surface parakeratinized stratified squamous epithelium.

The present case was from the buccal mucosa which is a rare site of occurrence. Lymphangiomas are present as either microcystic, macrocystic & mixed varieties. The presented case was a microcystic variant. Brennal et al.\(^9\) in their review on oral lymphangiomas reported 22.7 years as the mean age of occurrence and both sexes equally affected. Clinically oral lymphangiomas present as superficial lesion may demonstrate pebbly surface. The surface appears like frog eggs or tapioca pudding. Deeper lesions are present as soft ill-defined masses. Trauma may be one of the cause for superficial lymphangiomas.\(^{10}\)

Although benign, due to its infiltrating nature, indefinite demarcation and involvement of vital structures, these lesions may pose complications. Clinically, the lesions may appear like many other oral lesions such as haemangioma, teratoma, lingual thyroid nodules and granular cell tumour. Adult lymphangiomas are very infrequent, however the discussed case is of a 55-year-old
woman. Neither malignant transformation nor familial tendency of the lesions associated with age has been reported. Histopathologic features consist of lymphatic vessels with marked dilatation. The vessels often diffusely infiltrate the adjacent soft tissue and demonstrate lymphoid aggregates in their walls. The capillary lymphangioma consists of small capillary-sized vessels, cavernous lymphangioma consist larger dilated lymph channels, and cystic lymphangioma exhibits large macroscopic cystic spaces. More often, all sizes of vessels may be found within the same lesion. Lymphatic vessels are located just beneath the epithelium, often replacing connective tissue papillae. The literature review has shown 15 reported cases of lymphangioma till date to which the present case can be added. Of 15 reported cases, Brennan et al reported eight cases from buccal mucosa in their review on lymphangiomas on oral cavity. Two cases were reported by Domingo et al in their study of benign tumour of oral mucosa from the archives of general university hospital of Valencia. Dogan et al, Coskunses et al, Haranal et al, Pammar et al reported one case each. Various methods of treatment of lymphangiomas have been attempted including surgery, radiation, laser therapy, cryotherapy, electrocautery, sclerotherapy, and ligation. Large lesion of the tongue may be difficult to excise completely and may have high recurrence rate (39%).

<table>
<thead>
<tr>
<th>Year</th>
<th>Authors</th>
<th>Number of cases</th>
<th>Treatment done</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1997</td>
<td>Brennan et al.</td>
<td>8</td>
<td>Surgical excision</td>
<td>NA</td>
</tr>
<tr>
<td>2006</td>
<td>Bozkaya et al.</td>
<td>1</td>
<td>Radiofrequency tissue ablation technique</td>
<td>Nil</td>
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<tr>
<td>2008</td>
<td>Torres-Doming et al.</td>
<td>2</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>2010</td>
<td>Dogan et al.</td>
<td>1</td>
<td>Surgical excision later Cryotherapy follow up 1 year</td>
<td>2 Months</td>
</tr>
<tr>
<td>2012</td>
<td>Coskunses et al.</td>
<td>1</td>
<td>Surgical excision follow up 3\textsuperscript{1/4} years</td>
<td>NR</td>
</tr>
<tr>
<td>2013</td>
<td>Haranal et al.</td>
<td>1</td>
<td>Surgical excision</td>
<td>NA</td>
</tr>
<tr>
<td>2015</td>
<td>Pammar C et al.</td>
<td>1</td>
<td>Surgical excision</td>
<td>NA</td>
</tr>
<tr>
<td>2016</td>
<td>Present Case</td>
<td>1</td>
<td>Surgical excision</td>
<td>NA</td>
</tr>
</tbody>
</table>

Table 1: Cases of lymphangioma of buccal mucosa reported in the literature

The complications may affect the patient broadly in four dimension such as aesthetic, occlusal, functional and psychosocial. Complication related to infection of lymphangioma at base of tongue may result with Ludwig’s angina. Seroma formation, infections, minor bleeding, recurrent cellulitis and lymph fluid leakage are some of the few postoperative complication of oral and cervical lymphangiomas. Conclusion

Lymphangioma is primarily a childhood disease. It has to be identified and diagnosed early for uneventful treatment, reducing functional, psychological disturbances and cosmetic disfigurement. Oral lesions may be encountered in adults, where they have to be completely excised with clear borders to prevent recurrences.

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References

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