CASE REPORT

Cavernous lymphangioma of lower lip

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Abstract
Lymphangiomas are benign tumours of lymphatic vessels showing marked predilection for head and neck region. They are extremely rare in the oral cavity. The common site of occurrence for lymphangioma in the oral cavity is the anterior dorsum and lateral border of tongue. Rarely has it been reported in other parts of oral cavity such as the palate, cheeks, floor of the mouth, gingiva and lips. The most accepted treatment option for lymphangioma is complete surgical excision. There is only a minimal chance of recurrence. We report a case of cavernous lymphangioma of the lower lip which was excised and had satisfactory post-operative healing.

Introduction
Lymphangiomas are congenital malformations of lymphatic system that involve skin and subcutaneous tissue.\(^1,2\) This hamartomatous lesion was first described by Redenbacher in 1828.\(^3\) They are believed to arise as a result of lymph sac sequestration which fails to establish communication with larger lymphatic channels. They enlarge due to improper drainage or excessive secretion of lining cells.\(^4,5,6,7\) It has been reported that approximately 75\% of cases of lymphangiomas occur in the head and neck region.\(^8\) The most frequent site of involvement in the oral cavity is the tongue.\(^7\)
Less frequent sites of involvement include areas like palate, buccal mucosa, gingiva, floor of the mouth and lips. Current treatment methods of lymphangioma include surgery, sclerotherapy, laser therapy or a combination of these. The present report focuses on a rare case of lymphangioma of the lower lip in a male child with a review of the scientific literature.

Case report
A 13 year old boy reported to the Department of Oral Medicine and Radiology with a chief complaint of a growth on the lower lip for the past six months. The patient noticed a rapid enlargement of the growth following orthodontic treatment. He gave a history of a similar growth on the same site about one year back which was removed by surgical excision. On clinical examination, a painless growth was seen on the right lower lip in relation to 43, 44 region. The lesion appeared as multiple reddish pink transparent vesicles grouped together, measuring approximately 1.5 × 1 cm in dimension with an irregular surface and borders (Figure 1). A differential diagnosis of lymphangioma and mucocele were considered based on the clinical features. Wide surgical excision of the lesion was carried out and the biopsy specimen sent for histopathological examination. Post operative wound healing was satisfactory without any complications.

Figure 1. Lymphangioma of right lower lip of a thirteen year old boy (10x)
Histopathological examination showed large dilated cavitary spaces lined by flattened endothelial cells containing eosinophilic fluid located just beneath the surface epithelium (Figure 2). Numerous smaller and dilated lymphatic channels, few blood vessels and inflammatory cells were also noticed in the deeper connective tissue (Figure 3). The surface was covered by an atrophic keratinized stratified squamous epithelium. Based on the histopathological features, a final diagnosis of lymphangioma was made. Follow up of the patient was recommended for at least two years since this was a recurrent lesion.
Figure 2. Photomicrograph of the lip lesion (H&E x10) Enlarged lymphatic vessels containing lymph are seen in the superficial connective tissue

Figure 3. Photomicrograph of the lower lip lesion (H&E x10). Smaller lymphatic vessels containing lymph are seen in the deeper connective tissue
Discussion
Lymphangiomas are hamartomatous lesions of lymphatic vessels that show predilection for head and neck region. They occur as a result of structural defects in the lymphatic pathway, which usually develop during the sixth week of gestation. Approximately about 50% of the cases are noticed at the time of birth and around 90% develop by 2 years of age. The age of diagnosis can range from nineteen weeks of gestation to the second decade of life. According to a retrospective study of lymphangioma by Alqahtani et al, the average age at diagnosis was 3.3 years. No definite sex predilection is noticed. Determining the true incidence of these lesions is difficult due to lack of uniformity in classification and nomenclature. Lymphangiomas show a marked predilection for the neck and head region mostly the cervical area (posterior triangle, anterior triangle, submandibular and parotid region in descending order). Other common sites, outside the head and neck, include the axilla, shoulder, chest wall, mediastinum, abdominal wall, and thigh.

Lymphangiomas of lateral and posterior neck region are frequently associated with chromosomal anomalies like Turner’s syndrome, Noonan’s syndrome and foetal hydrops. Primary intra-osseous lymphangioma is a very rare entity that occurs as a result of gradual extension of a peripheral soft tissue lymphangioma to the periosteum of adjacent bone and into the marrow spaces.

Lymphangioma rarely involve the oral cavity. Most commonly involved site is the dorsum of tongue. Lower lip is an uncommon site comprising of approximately 6% of all oral lymphangiomas. Characteristic clinical appearance of oral lymphangioma is a superficial pebbly vesicular lesion resembling “frog egg / tapioca pudding”. Deep seated lesions appear as soft ill defined submucosal masses that markedly distort the local anatomy. Half of the cases may be diagnosed with the help of clinical manifestations alone. Early diagnosis may be established with the help of ultrasonography, MRI, arterogram and CT angiography.

In the oral cavity, lesions like haemangioma, lingual thyroid, thyroglossal duct cyst, granular cell tumour, cheilitis granulomatosa, mucocele/ ranula and cellulitis may be considered for differential diagnosis.

The lesions are capable of rapid growth with infection or trauma. This may result in disfigurement as well as severe impairment of respiration, deglutition and speech especially lymphangioma of the tongue. Even though they are benign lesions, spontaneous regression is rarely seen (1.6–16.0%). With adequate follow-up, it is seen that a regression is usually followed by a recurrence.
The case discussed here occurred on the lower lip which is a rare site for lymphangioma with a rapid growth rate in response to trauma. It was a recurrent case of lymphangioma within a period of one year, following surgical excision.

Histopathologically, lymphangioma may be classified into three types according to the size of the lymphatic vessels- lymphangioma simplex (capillary lymphangioma) which consists of small capillary sized vessels, cavernous lymphangioma composed of larger dilated lymphatic vessels and cystic lymphangioma (cystic hygroma) exhibiting large macroscopic cystic spaces. The lymphatic channels are usually lined by a single layer of endothelial cells. Often all three histological types may be found in the same lesion. Among these the most common intraoral type is cavernous lymphangioma.

The structure of blood vessels and lymphatic vessels show certain similarities and differences. The supporting pericytes and smooth muscle cells of blood vessels produce a large amount of extracellular matrix, whereas supporting cells are sparse in the case of lymphatic channels and hence lack a continuous basement membrane. Lymphatic vessels also have fenestrations in their vessel walls. These factors attribute to the increased permeability of lymphatic channels. D2-40 is an excellent immunohistochemical marker for lymphatic vessels and helps in differentiation lymphatic endothelial cells from endothelial cells lining the blood vessels.

Though lymphangiomas are benign lesions, the involvement of vital structures or aesthetic /functional requirements may necessitate treatment of these lesions. The most accepted treatment modality for lymphangioma consists of complete surgical excision with minimal chances of recurrence. Complete excision is possible only for small and localized lesions. The management of head and neck lymphangioma is challenging because of its close association with adjacent vital structures and poor demarcation of the lesion. Hence, complete excision is difficult to achieve in some cases and recurrence can occur following incomplete removal. Multiple treatment modalities have been recommended for deep seated and large lesions. They include cryotherapy, electrocauterization, sclerotherapy (OK-432), administration of steroids, embolization and laser therapy. According to Gilberth, CO₂ laser technique was more convenient with less chance for recurrence than any other methods. The various treatment options for lymphatic malformations have their advantages and disadvantages and the method of treatment should depend on the status of the patient, available technology and expertise.

Conclusion
Lymphangioma of the head and neck are benign lymphatic malformations with a marked predilection for head and neck. In the oral and maxillofacial region diagnosis of lymphatic malformations is easy but management become difficult due to involvement of vital structures and chances for disfigurement. Though rarely seen in the oral cavity lymphangiomas are an eventuality to take into consideration by the oral surgeon. Early recognition is of utmost importance to initiation of proper treatment and avoidance of complications.

References
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